Indian Journal of Plastic Surgery 1999;32 54-68

INDIAN JOURNAL OF PLASTIC SURGERY

Invited Article

TRENDS IN CRANIOFACIAL AND CLEFT SURGERY FOR CONGENITAL DEFORMITIES - PERSONAL EXPERIENCE

Kenneth E Salyer, Rajiv Agarwal
International Craniofacial Institute, Dallas Texas, USA

INTRODUCTION

Modern craniofacial surgery owes its existence to the classic works and pioneering efforts of Paul Tessier, a Frenchman who started performing these difficult operations involving facial bone and cranial vault movements over thirty five years ago.[1-3] He first performed mock surgery on cadaver skulls before translating his technique to live patients with monstrous craniofacial deformities. Those were the times when there was no intracranial approach to the face, precision cutting saws and drills, controlled hypotensive hemodilutive anaesthesia, resorbable plates and screws, bone substitutes and last but not the least a support team of dedicated individuals. With his untiring efforts, scientific presentations and relentless pursuit in the treatment of complex facial deformities which were not even completely understood at that time he gave birth to a new era of craniofacial surgery which has developed fast into one of the most challenging and gratifying subspecialities of plastic surgery. It has helped thousands of children with congenital and acquired deformities to lead a socially and psychologically acceptable life.

The emergence of craniofacial surgery attracted a group of devoted followers who through their personal efforts widened the scope of this subspeciality. Many new craniofacial training centres and training programs developed in the United States and abroad. In 1969 one of the first craniofacial centre in the United States was beginning to evolve by the senior author (KES) which was established in 1971. In 1986 in Dallas a dedicated centre was established called the International Craniofacial Institute, where more than 9,000 craniofacial patients have been treated from every state in the United States and sixty countries. With the rapid advances in technology, anaesthesiology and surgical techniques and also with the close co-operation of pediatric neurosurgeons the overall results from craniofacial surgery have improved with the complication rate less than 5%, infection rate of 1.9% and mortality rate is less than 0.1% [4]. Over these years of continuous work and experience we have developed our philosophy, approach and protocol for the treatment of craniofacial, cleft lip-palate and related problems, developing many innovative procedures and techniques[5,6].

THE CRANIOFACIAL TEAM

Craniofacial surgery requires an experienced and dedicated team. Our team includes the craniofacial surgeon, pediatric neurosurgeon, pediatric intensivists, pediatric anaesthesiologist, orthodontist, pedodontist, geneticist, speech pathologist, psychologist, physical anthropologist, otolaryngologist, ophthalmologist, neuroradiologist, photographer, medical illustrator, craniofacial nurse co-ordinator and a social worker. As the team represents many disciplines, the surgeon is called upon to weigh many points of view for decision making. Team conferences concentrate on the most dramatic or challenging cases. In the operating room it may be important to have two teams working simultaneously, one in the area of the head and the other for example at the patient's side obtaining bone or cartilage grafts.

ANAESTHESIA FOR MAJOR CRANIOFACIAL OPERATIONS

Major craniofacial surgery is performed under controlled hypotension with the mean arterial pressure in the range of 50-55 mm Hg, normovolemic hemodilution with the hematocrit in the range of 24-27% and hyperventilation (p_aCo_2 22-28 mm Hg) for achieving brain shrinkage. Controlled hypotension is achieved by the use of oral clonidine as part of the

preoperative preparation with the addition of IV labetolol as needed. Normovolemic hemodilution is performed after the airway has been secured and all intravenous and arterial lines are in place. Replacement fluids are 2-3 ml RL: 1 ml whole blood for half the collection and at a 1:1 ratio of 5% plasma protein fraction for the other half of the collection. For patients heavier than 50 kg, the entire collection. may be replaced with RL at 2-3 ml of whole blood collected. Management of difficult airways can be achieved today without the use of tracheostomy which has been essentially eliminated.

UNILATERAL CLEFT LIP NOSE

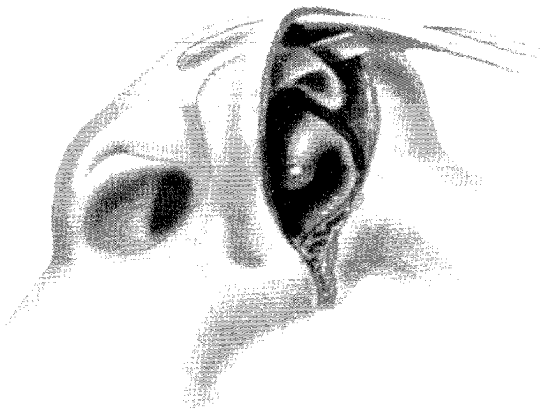
In the last two decades many advances have been made in cleft repair, including a more focussed approach towards primary or early secondary correction of the cleft lip nose.[7] Multiple factors cause the nasal deformity which include displacement of the lower lateral cartilage with flattening and false lengthening on the cleft side with shifts of the columella, septum and the underlying skeletal base. Most complete clefts of the maxillary segments have varying degrees of maxillofacial skeletal dysplasia resulting in variable growth abnormalities. For many years it was thought that correction of nasal deformity interferes with the growth of the nose and that the repair may harm or cause increased deformity. This was based on "bad" surgery which caused severe scarring and contributed to the deformity instead of correcting it. Our experience with all types of congenital deformities makes us more convinced that early well executed surgery releases a potential for more normal growth and development.[8]

The goal of primary cleft lip nose repair is to create a balanced, symmetrical lip and a normal looking nose. Our personal approach to this problem creates the nasal floor, repositions the base of the ala into a symmetrical position with the noncleft side, widely dissects the lower lateral cartilages and repositions them to obtain optimal symmetry of the alae, nostrils and nasal tip. Before starting cleft lip nose correction passive presurgical orthopedic treatment is used, a palatal plate is molded and applied to the upper arch which covers the palatal cleft and also simultaneously helps in repositioning the maxillary segments after primary cleft lip closure. We believe that active presurgical orthodontic treatment is unnecessary and probably is detrimental to growth.

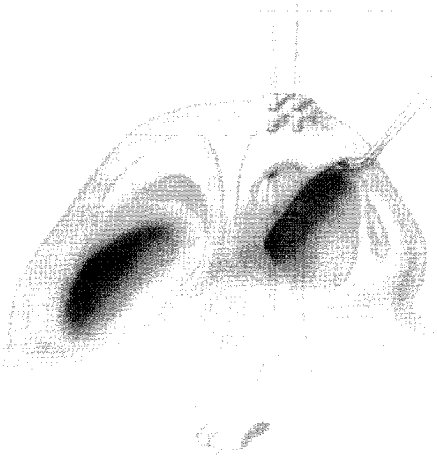
A modified Millard rotation-advancement approach is used for cleft lip nose repair (Fig.1). The modification does not include a back-cut as a general approach, however the incision that creates the C-flap is used as the approach to the lower lateral cartilages. Through this incision, the medial crura are dissected apart continuing over the alar dome so that the skin is undermined over both lower lateral cartilages. Lateral access to the lower lateral cartilage is at the alar base, which allows separation of the nasal ala from the displaced lesser maxillary segment. Releasing the soft tissue from the skeletal base on the cleft side just above the turbinate allows for marked improvement of the position of the nasal structures. All abnormal attachments of the lower lateral cartilage are released except at the level of the genu which allows for shifting the lower lateral cartilage without totally dissecting it from the skin and the nasal lining. Following this dissection all the elements are placed in their new positions without any tethering or distortion. The key orbicularis oris catgut suture approximates the lip creating a structural base for the final lip and nose reconstruction. Any excess tissue in the nasal floor is not removed at this time to avoid the most undesirable complication of a small nostril. Therefore closure of the floor of the nose may err on leaving a slightly larger nostril on the cleft side. Stent sutures are placed through and through the lining, lower lateral cartilages and the skin starting at the peak of the dome of the cleft side and they are very crucial for readjusting the lower lateral cartilage and redraping the skin envelope to obtain optimal symmetry. More recently intra-nasal stents are used postoperatively for 3 months to maintain shape and control scarring.

Reconstruction of the nasal sill is very important and requires specific attention because inability to equalise the sill on both sides frequently leads to an undesirable result inspite of good cartilage work. In children presurgical orthopedic treatment when combined with adequate intraoperative muscle approximation allows acceptable reconstruction of the cleft nasal sill. For secondary cases and for those who present late, especially adult incomplete clefts the use of turn over orbicularis oris muscle flap for augmenting the nasal sill gives satisfactory results.[9]

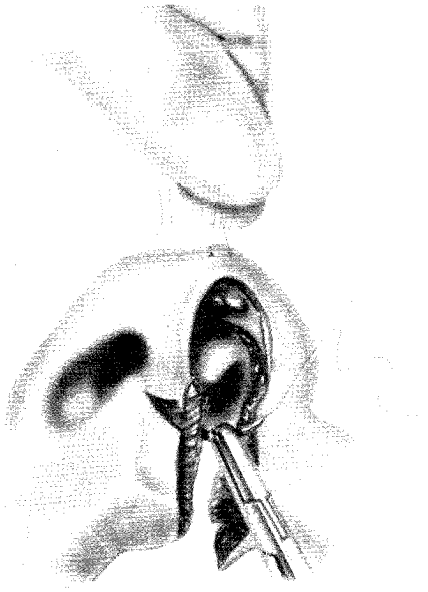
There may be remaining minimal nasal deformities of the ala, nostril asymmetry, or inadequate tip projection on the cleft side. The primary surgical technique used for unilateral cleft nose deformity eliminates major deformity of the nose converting the secondary correction to more minor procedures of aesthetic



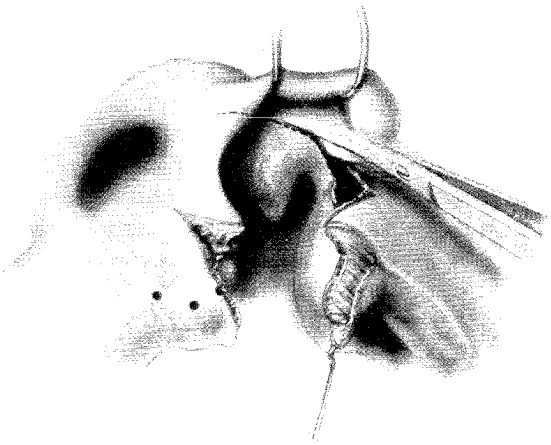
(Fig-1a) The complete mobilization of all the displaced nasal and lip elements is the key to ideal reconstruction



(Fig-1b) Repositioning of the cartilages with readaption of the skin, cartilage and lining is achieved by using through and through stent sutures



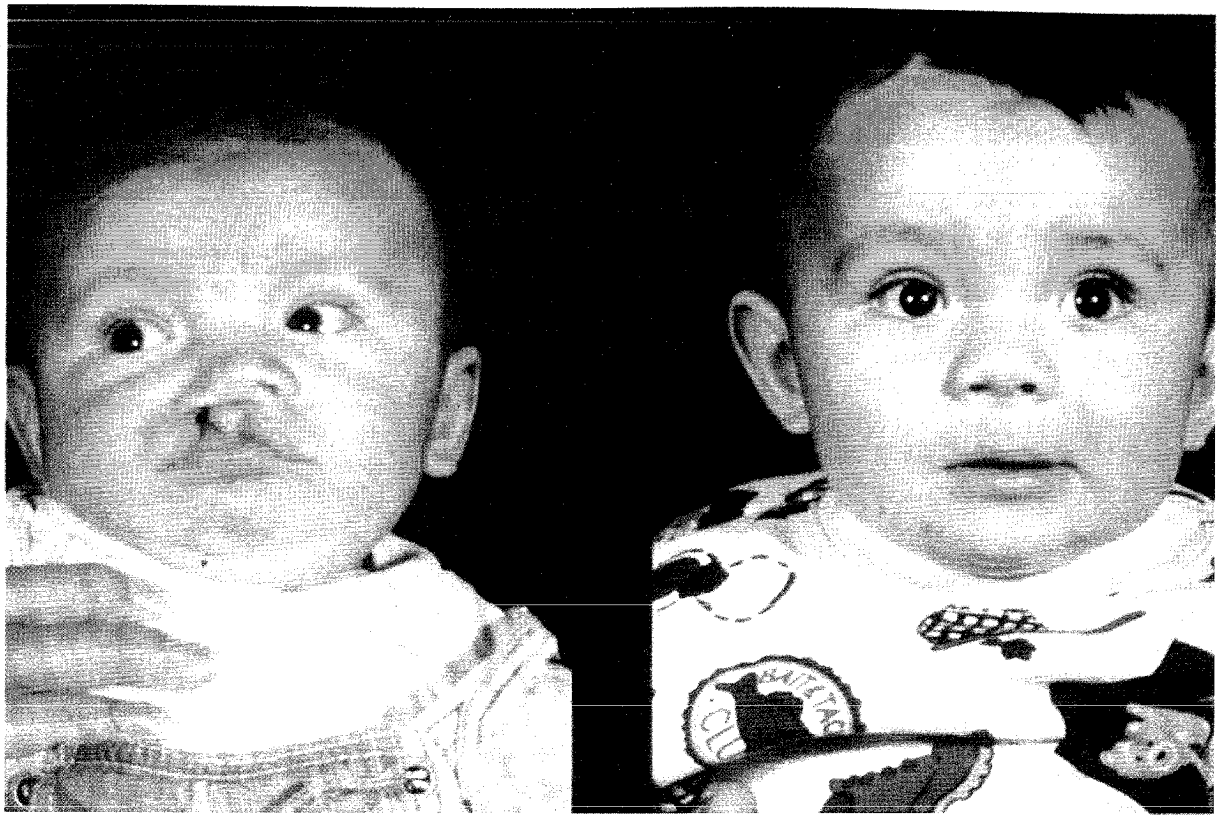
(Fig-1c) Placement of the key muscle suture in the lip sets up the foundation for a symmetric nose and lip



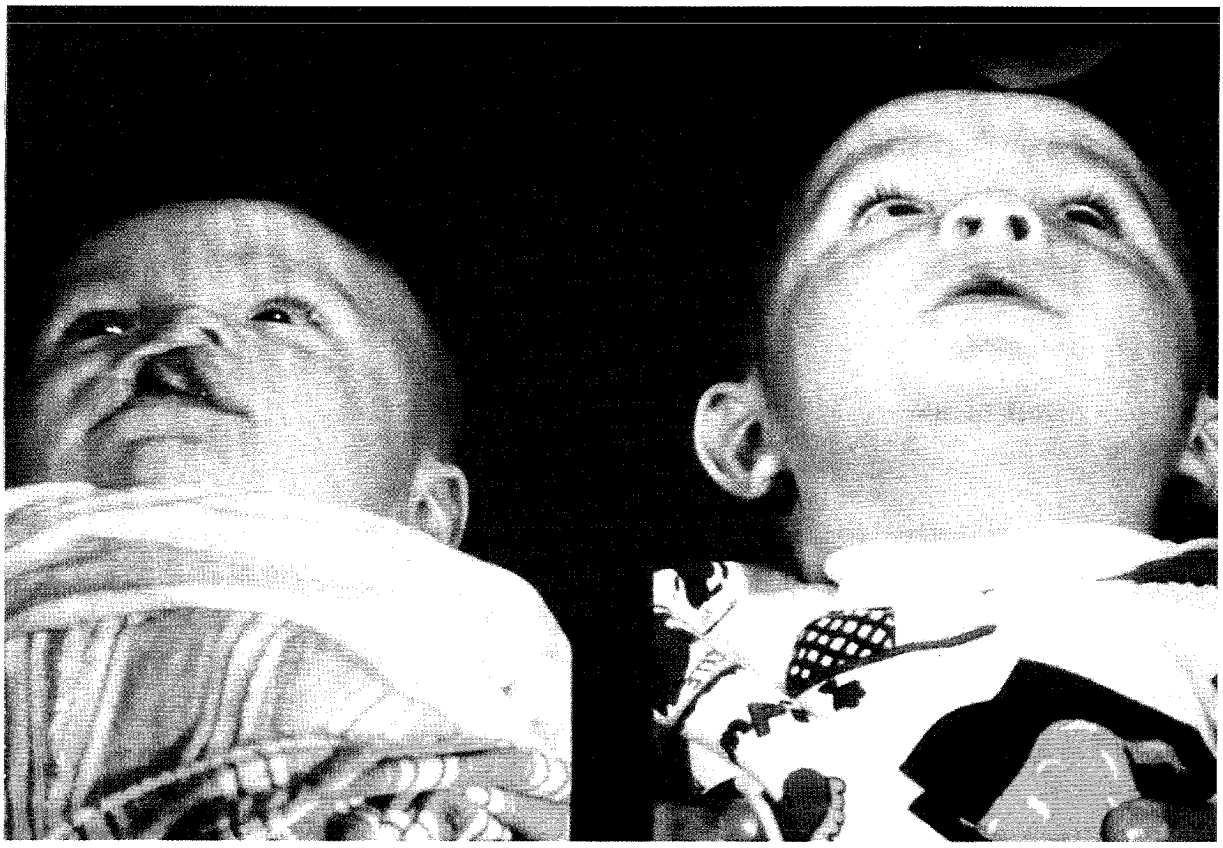
(Fig-1d) Incision along the inferior turbinate facilitates alignment of the lower lateral cartilage

enhancement. In the last 30 years the major contributions in achieving surgically improved results in addition to primary cleft nasal repair are:

1. Passive orthodontic treatment in the infant.
2. Surgical orthodontic speech oriented ongoing care.
3. Palatal expansion with cancellous bone grafting at the time of development of the canine tooth eruption in the cleft space, usually at age 8 or 9 years.
4. Secondary nasal surgery when necessary including septoplasty, turbinectomy, cartilage grafting and nasal osteotomies.
5. Orthognathic surgery provides the ability to change significantly the facial skeleton by performing a Le Fort I osteotomy of the maxilla frequently accompanied by bilateral sagittal split and sliding genioplasty of the mandible. The lamellar split of the malar bone or demineralized bone grafting for improved facial convexity is frequently preferred at our centre. Approximately 20% of our primary cases require orthognathic surgery.



(Fig-1e) Before and after result in a case with right unilateral complete cleft lip and palate in frontal view



(Fig-1f) Before and after result of the same patient in basal view

MAXILLARY OSTEOTOMY

Maxillary and mandibular osteotomies allow the plastic surgeon the opportunity to create aesthetic results in addition to restoring the occlusal relationship (Fig.2). The most important procedure in cleft cases is the Le Fort I maxillary osteotomy which is performed in more than 75% of the patients undergoing orthognathic surgery either by itself or in combination with mandibular or chin procedures (Fig.3). Three dimensional movements of the maxilla allows vertical, anterior and posterior changes safely and predictably under direct vision. Maxillary osteotomies are closely correlated with presurgical orthodontic preparation and planning. A horizontal maxillary osteotomy is preferable which brings the maxilla forward into a stable position which is then fixed by miniplate fixation.

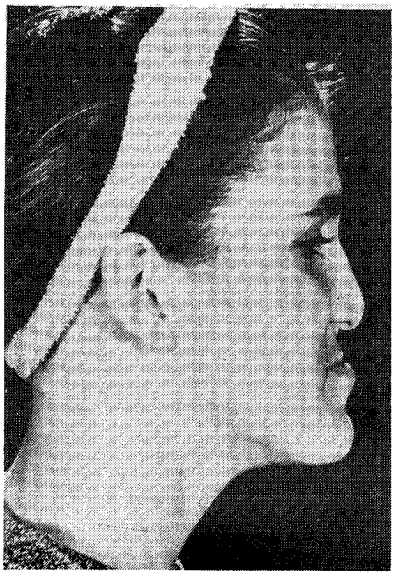
Vertical shortening of the maxilla is required when the central incisors are exposed 3 to 4 mm below the inferiormost portion of the upper lip in repose. Vertical lengthening is indicated when there is inadequate show of the central incisors. When there is considerable vertical shortening of more than 4 to 5 mm it may be necessary to open the nose and remove the leading edge of the septum to prevent buckling or it may be necessary to simultaneously perform a turbinectomy to prevent nasal airway obstruction. For vertical lengthening of the maxilla demineralized bone, hydroxyapatite, or the cranial bone are used as interpositional grafts depending upon the particular case and other associated procedures and exposure.

Segmental maxillary osteotomies may be performed with the Le Fort I osteotomy which has been downfractured. The index finger is placed on the mucosa and an anterior maxillary osteotomy is performed dividing the cleft maxilla into two or three segments. We prefer presurgical orthodontic expansion. This improves post-operative stability and soft tissue contracture in the cleft patient which is usually soft tissue deficient.

MANDIBULAR OSTEOTOMY

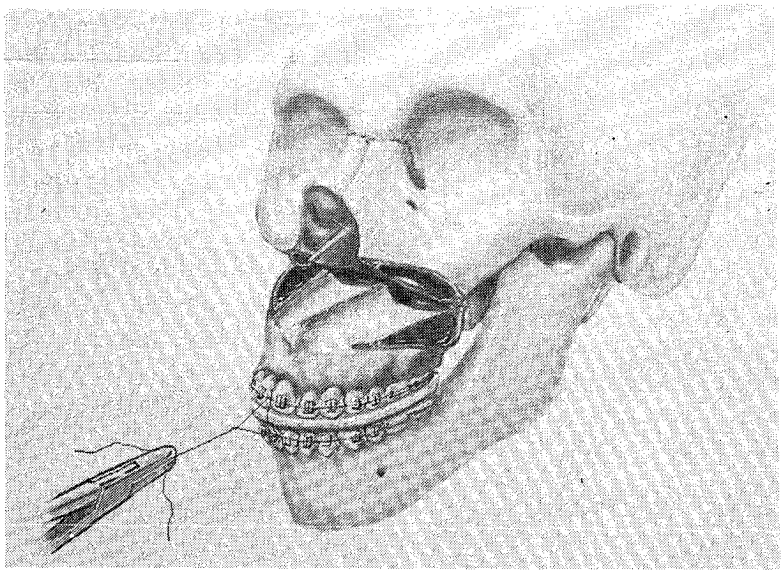
Surgery of the mandible is routinely performed to correct a variety of dentofacial deformities in the cleft patient and frequently in combination with maxillary surgery and genioplasty. The modified sagittal split osteotomy is the ramus procedure used in 85% of our cases. On occasion anterior and posterior subapical mandibular segmental surgery, body osteotomies, or other ramus procedures may be indicated. Our first priority in planning orthognathic surgery is to produce an attractive face, normal occlusion is considered secondarily. Most patients requiring mandibular osteotomy for dentoalveolar problems also require orthodontic treatment for optimum results. Performing simultaneous maxillary Le Fort I and mandibular osteotomy allows improved facial balance and stable functional results using internal rigid fixation.

The sagittal split osteotomy first described by Obwegeser[10] and modified by DuPont[11] and refined by Dautrey[12] has diminished the incidence of inferior alveolar nerve damage. Further



(Fig-2a) Preoperative right lateral view showing marked maxillary-malar-paranasal deficiency with midfacial retrusion

(Fig-2b) Postoperative right lateral view 3 years after Lefort I correction, split cranial-bone grafting for maxilla and malar deficiency, and secondary nasal surgery.



(Fig-3) A prefabricated dental splint is wired into the position of the upper and lower teeth following Lefort I osteotomy to stabilise the maxilla into its new position

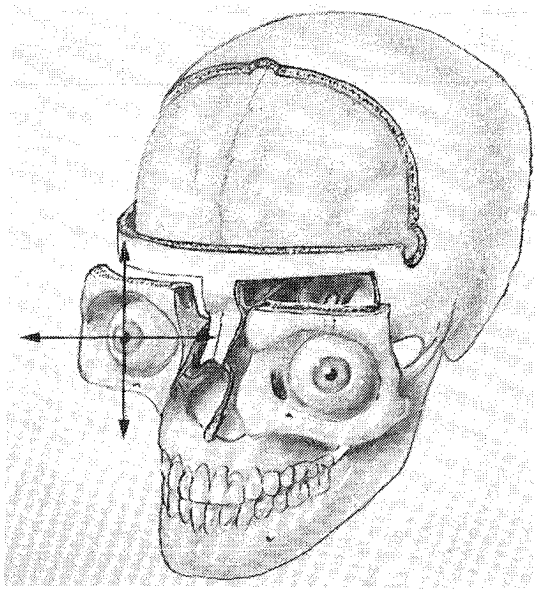
improvements and refinements by the senior author (KES) have facilitated safer and more rapid performance of the procedure. Using our modified technique most cases now average only 30 minutes to perform the osteotomies bilaterally. From initially using extensive dissection we now limit the planes of dissection, thus decreasing blood loss, simplifying the surgery, and increasing vascularity to the osteotomized segments. This decreases the vascular ischaemia of the mandibular ramus that has been implicated in aseptic necrosis, sequestration, and loss of bone that can be associated with this procedure.

The osteotomy starts with a horizontal medial osteotomy placed above the level of the mandibular nerve, the saw blade is then reinserted at the anterior edge of this cut to extend through the mandible staying above the angle of the mandible posteriorly but through the lower border of the mandible anteriorly. Once the osteotomy has been performed, the soft tissues are modified to allow easy occlusal intermaxillary fixation making sure that the condyle is pushed into the glenoid fossa. Fixation is accomplished by either screws, plate or wires. Screw placement is greatly facilitated by a microoscillating side drilling motor driven hand piece which avoids percutaneous incisions and scarring.

Mandibular setback is performed using the same technique. The proximal segment is remodeled by cutting the bone using a tongue in groove technique which fits the distal segment. The segments are secured with interosseous wires, multiple screws or plates which fix the proximal and distal segments.

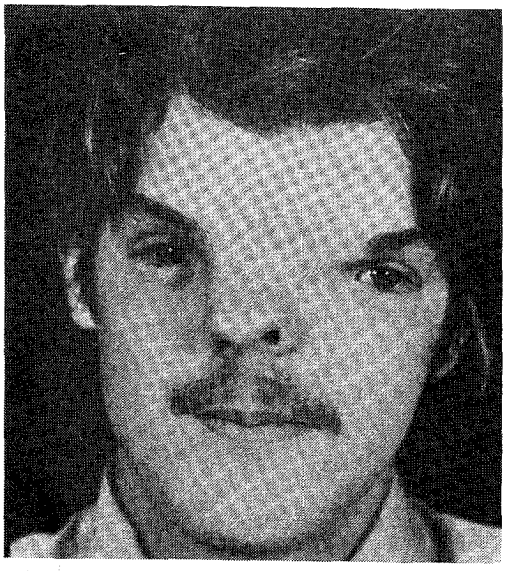
HYPERTELORISM

Correction of hypertelorism challenges the most experienced craniofacial surgeon because individual artistry plus complex movements of the nose and orbits are required to achieve good results (Fig.4,5). Before the advent of intracranial surgery, subcranial approach was used by Converse[13] to correct minor to moderate deformity. This approach however requires an accurate assessment of the anatomy, to prevent inadvertent entrance into the intracranial space which can lead to major complications. Generally an intracranial total orbital osteotomy is the technique of choice for major cases of hypertelorism in fully grown patients. Preoperative soft tissue planning is important to achieve an aesthetic result of the nasal correction. Displacement of the orbits may be symmetrical or asymmetrical in vertical or horizontal dimensions. Also the deformity of the orbital contour may involve separate segments of the orbit, the rims and four walls may be affected in different combinations depending on which walls are most distorted or misplaced. Hypertelorism is frequently associated with excessive pneumatization of the ethmoid sinuses which presents a technical problem at the time of surgical correction. We have shown that complete anterior ethmoidectomy is necessary to achieve

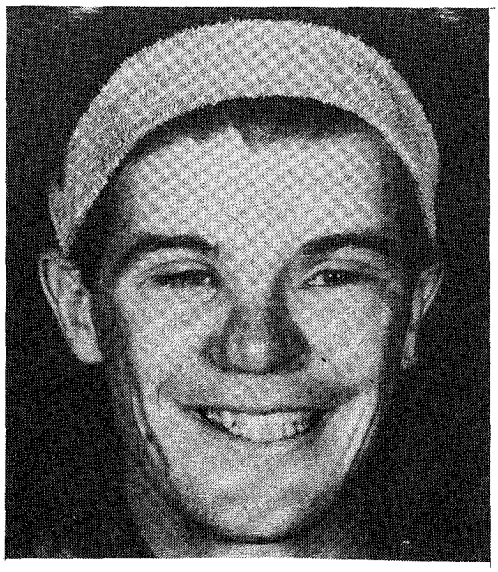


(Fig-4) The globe and orbit may be translocated in any direction. The appropriate orientation of the orbit, and especially the globe must be maintained to avoid postoperative complications

correction of the hypertelorism. The position of the globe is of great importance in planning and performing the correction of hypertelorism. Exophthalmos or enophthalmos, unilateral or bilateral may be present creating additional problems in repositioning of the orbits. The opening of the palpebral fissure on one or both sides may be decreased or increased. If the changes are marked, additional surgical intervention may be required. Also it is possible for the nasolacrimal system to be displaced, distorted, hypoplastic, obstructed or even absent on one or both sides. Another marked characteristic feature of hypertelorism is the nasal deformity evident in the majority of cases. The nose and the columella are short and wide. The lower lateral cartilages are usually deformed, flattened and elongated. The nasal tip is obtuse and in the majority of cases duplication of the septal cartilages creates an excessively thick nasal septum and an excessively wide nasal bridge. The nasal bones are usually hypoplastic and most of the nasal framework is cartilaginous. Besides these, hypertelorism may be associated with a median cleft of the nose and or lip, unilateral or bilateral cleft of the nasal ala, lack of formation of the nasal tip and a V-shaped hairline extension into the forehead over the area of cranium bifidum. It is important to realize that all of these features may be present in various combinations and severity and therefore careful clinical examination, combined with a detailed analysis of the X-rays and CT scan allow the surgeon to make a precise diagnosis and to formulate a detailed plan of the surgical procedure. The timing of correction depends on the patient's age, the severity of the deformity and the psychosocial effects of the existing deformity on the patient and his or her family. We do surgery between 2-4 years of age when the interorbital distance increases rapidly exceeding 30-40mm, resulting



(Fig-5a) Preoperative frontal view of a 21-year-old patient with hypertelorism, with an interorbital distance of 50mm, orbital dystopia, and a short nose



(Fig-5b) Postoperative frontal view 5 years after correction of hypertelorism and nasal reconstruction by using a costochondral graft and del Campo medial canthopexy and forehead remodeling

in both wide bizygomatic and hemifacial distances. We correct hypertelorism and craniosynostosis in the same operation prior to one year of age, performing both forehead advancement and frontocranial remodelling at the same time in order to take advantage of the bone formation potential of the dura.[12]

The hypertelorism correction is performed through intra- or extracranial exposure. Intracranial exposure is obtained by a bifrontal craniotomy using the bicoronal skin flap approach.[14] No scalp hair is ever shaved. It is only necessary to have clean hair and scalp preoperatively. The skull and soft tissues are undermined in the subperiosteal plane, supraorbital nerve is turned down with the globe in order to preserve its sensation and the entire globe is mobilized in subperiosteal plane. The classic four-wall intracranial correction of hypertelorism is not performed in the growing child's face because of tooth damage. Certain modifications have been developed including the inverted U and the variations of medial orbital wall osteotomy.

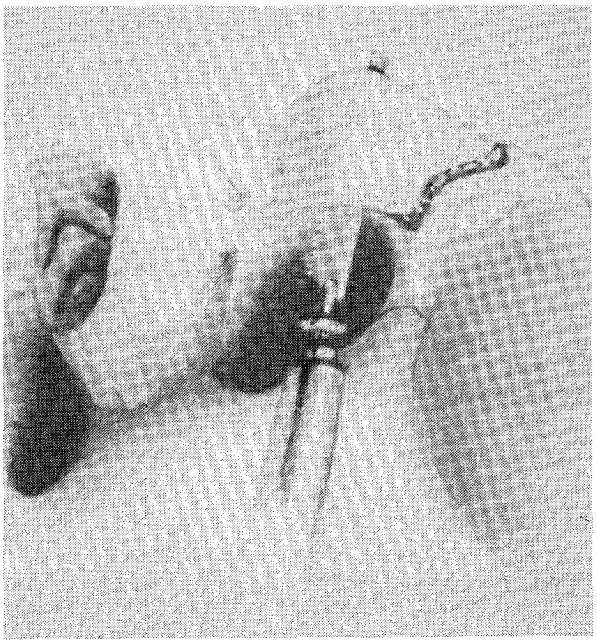
Preservation of the olfactory nerves at the time of intracranial approach can be ensured through careful dissection, modification of the osteotomies and preservation of the nasal mucosa attached to the olfactory fibres. Paramedial osteotomies are performed around the cribriform plate, or if the plate is very wide or deformed, it may be completely removed to facilitate medial translocation of the orbits. In cases where the superior medial orbital walls appear to be wide, a segmental osteotomy of the orbits with removal of the excessive bone results in optimal correction. Prior to medial orbital wall osteotomy, the nasal lacrimal apparatus is identified and complete unroofing of the lacrimal duct system is performed to prevent damage. Removing the bone over the nasal lacrimal duct allows for the translocation of the orbit without kinking the duct by bony segments.

LAMELLAR SPLIT OSTEOTOMY

Lamellar split osteotomy is a new concept in craniofacial surgery which was conceived and originated by Salyer et al in 1990.[15] This involves splitting insitu of the frontal, orbital, zygomatic and or nasal bone into an internal and external lamella. The outer lamella can be maneuvred by bending, sliding or rotation to produce a three-dimensional contour change of the external shape of the skull bone being operated. The internal lamella stays in its native position and thus preserves the reference points important in establishing the bony topography and facial symmetry. A basic advantage with this technique is the ability to change the contour without using additional implants. The outer table is easily fixed in its new position with plates and screws making the procedure safe and efficient. Vascularity of the outer table is preserved by leaving attached periosteum and muscle attachments to the segments (Fig.6).

Prior to evolution of this technique, the classic bilamellar bony osteotomies containing the entire thickness of the dysmorphic bone left a gap or a retruded area on the bony skeleton which made planning by three dimensional CT or cephalometrics very difficult along with making the positioning of the osteotomized segment a decision based on recall and judgement rather than on precise repositioning with respect to references.

In establishing the indications for use of lamellar split osteotomy, the key determinants are the dysmorphology of the anomaly and the maturity of bone. Patients with mild to moderate frontonasal dysplasia, hypertelorism, hypotelorism, malar-maxillary deficiency or orbital dystopia are good candidates. For those having severe hypertelorism, hypotelorism or orbital dystopia where translocation of the globe is necessary, classic osteotomies are indicated.



(Fig-6) Lamellar split of the malar bone can be accomplished by using either the coronal approach or the intraoral approach. The split is carried over to the level of the inferior orbital nerve so that the entire zygomatic arch can be changed in shape, projection, and width

In establishing the maturity of the craniofacial skeleton, the following factors must be considered. First, prior to 2 years of age, the bony consistency of the orbital facial bones is soft with minimal cancellous structure. Secondly, 75 percent of the natural orbital volume is not obtained until 2 years of age.[3] Finally in pediatric patients under 3 years of age, primarily in craniosynostosis, concerns of increasing cranial volume by advancing or altering the anterior or middle fossa are more important than orthomorphic restructuring. In light of these concerns, patients under the age of 2 years are not considered as candidates for lamellar split osteotomy. Thus this versatile procedure can be used to alter the shape and contour of the frontal, orbital, malar, maxillary and mandibular bones in patients 2 to 3 years of age and older.

The operation is performed under general hypotensive endotracheal anaesthesia. This procedure can be performed extracranially or intracranially depending upon the extent of the osteotomies. In the intracranial approach the cranium, orbits, and malar areas are exposed in the subperiosteal plane using the coronal incision. After removal of the bifrontal cranial bone flap the internal lamella is split from the external lamella with a reciprocating saw.

Malar augmentation using the lamellar split osteotomy results in improved facial aesthetics and is especially useful for treating malar bone hypoplasia. This deformity may result from congenital conditions, trauma or other various etiologies. Hypoplastic malar-zygomatic bony complex is one of the main characteristics of both Treacher-Collins syndrome and unilateral or bilateral hemifacial microsomia. A coronal incision is used for access and subperiosteal dissection is performed to expose the lateral orbital wall down to the zygomatic arch and then to the anterior wall of the maxilla, identifying the infraorbital nerve. The inferior orbital rim is cut lateral to the infraorbital nerve and osteotomy is extended down through the outer table in front of the maxillary sinus. Extending this osteotomy laterally allows the malar bone to be split at its attachment to the maxilla and to the zygomatic arch. Separation of the outer table allows for its advancement and counterchange, increasing the lateral and anterior projection of the malar-zygomatic complex and simultaneously maintaining the muscle attachments. To maintain this new position of the bony segment, a bone graft is placed between the outer and inner table and fixed using a single screw or a small plate. Thus this procedure allows not only tranposition of the malar outer table laterally and anteriorly but also allows change in the direction and shape of the malar outer table.[16]

CRANIAL VAULT REMODELING

Cranial vault remodeling provides expansion or reduction of the cranial volume. Cranial vault expansion is indicated when increased intracranial pressure produces clinical symptoms. Craniosynostosis and hydrocephalus are the most frequent causes of increased intracranial pressure. Various tumors as well as fibrous dysplasia may have the same effect.

Cranial vault reconstruction involves reshaping the proportions of the deformed cranium to achieve normal shape and a better balance between the cranium and the face. The shape and size of the anterior, middle and posterior cranial fossa can also be changed, improving the shape and balance of the head and face.

The key to total cranial vault reconstruction is the use of the forehead bandeau, a concept that we have applied to other areas of the cranial vault. The bandeau allows the angle of the forehead, orbital, and frontonasal angle disproportions to be improved, and anterior and middle cranial-base abnormalities to be expanded or changed. The midcranial vault bandeau is helpful in adjusting the vertical height of the cranium. The occipital bandeau changes the height and posterior projection in the occipital region and corrects deformities of the posterior cranial vault and fossa.[5] Applying these techniques for total cranial vault expansion in syndromal craniosynostosis improves results in the most difficult cases. The application of the same techniques to cranial vault reduction has opened new possibilities for treating children with hydrocephalus. Head reductions have allowed these patients to hold up their heads and walk.

Craniosynostosis leads to an abnormally shaped head and face which is disfiguring and causes social and psychological embarassment. Frontocranial remodeling is successful in infants with craniosynostosis and is best performed at six months of age, but patients who have not had this surgery or who had inadequate remodeling previously, are candidates for forehead remodeling at any age. Patients with simple suture synostosis make up the majority of our patients with cranial vault abnormality. Since 1973, the "tongue in groove" method has been used by the senior author (KES) with good results which allows for accurate, stable placement of the deformed parts in these cases. Another technique

that is effective for the retruded, flat relatively high forehead seen in oxycephaly is the transposition of bone flaps with temporal Z-plasty which allows the forehead to be rocked forward as described by Marchac.[17]

It is always important to make a differential diagnosis between a craniosynostosis and a positional deformation. The latter generally corrects itself in a few months or is corrected by nonsurgical moulding of the head. True synostosis does not correct itself and the abnormality remains the same or worsens over time. To avoid complications, surgery should be performed early enough to allow the brain, the skull and the face to grow normally. The primary diagnosis can be made on the basis of skull radiographs and the most important differential diagnosis which determines the outcome of surgery is whether there is a simple suture synostosis or syndromal synostosis. Our results in simple suture synostosis where one or more sutures are involved are excellent. Few cases require secondary surgery, almost all syndromal craniosynostosis cases require repeated procedures because they are programmed to grow abnormally.

A number of patients with anatomic abnormalities or syndromes, such as Apert's syndrome have a frontal or supraorbital prominence. Pneumatization of the frontal sinuses in nonsyndromic patients may also cause a similar excessive supraorbital prominence which can be easily corrected by remodeling. In Apert's, recurrence of this severe deformity occurs over time. Frontometaphyseal dysplasia also produces hypertrophied, thickened and distorted supraorbital ridges. An intracranial approach may be required for complete reconstruction of the frontocranial-orbital bone.

At our centre cranial vault remodeling is performed at six months of age when the bone is mature enough for stabilization of the bony fragments. Total cranial vault remodeling evolved from frontocranial remodeling, which involves access to the anterior, middle and posterior fossae allowing expansion of these abnormal portions of the cranium. The midportion of the cranial vault in the temporo-parietal region is removed as a large U-shaped segment of bone which can be used to lower the vertical dimension of the head. All syndromal deformities require repeated cranial and facial procedures to achieve optimal results. Total cranial vault remodeling results in improved appearance in these patients.

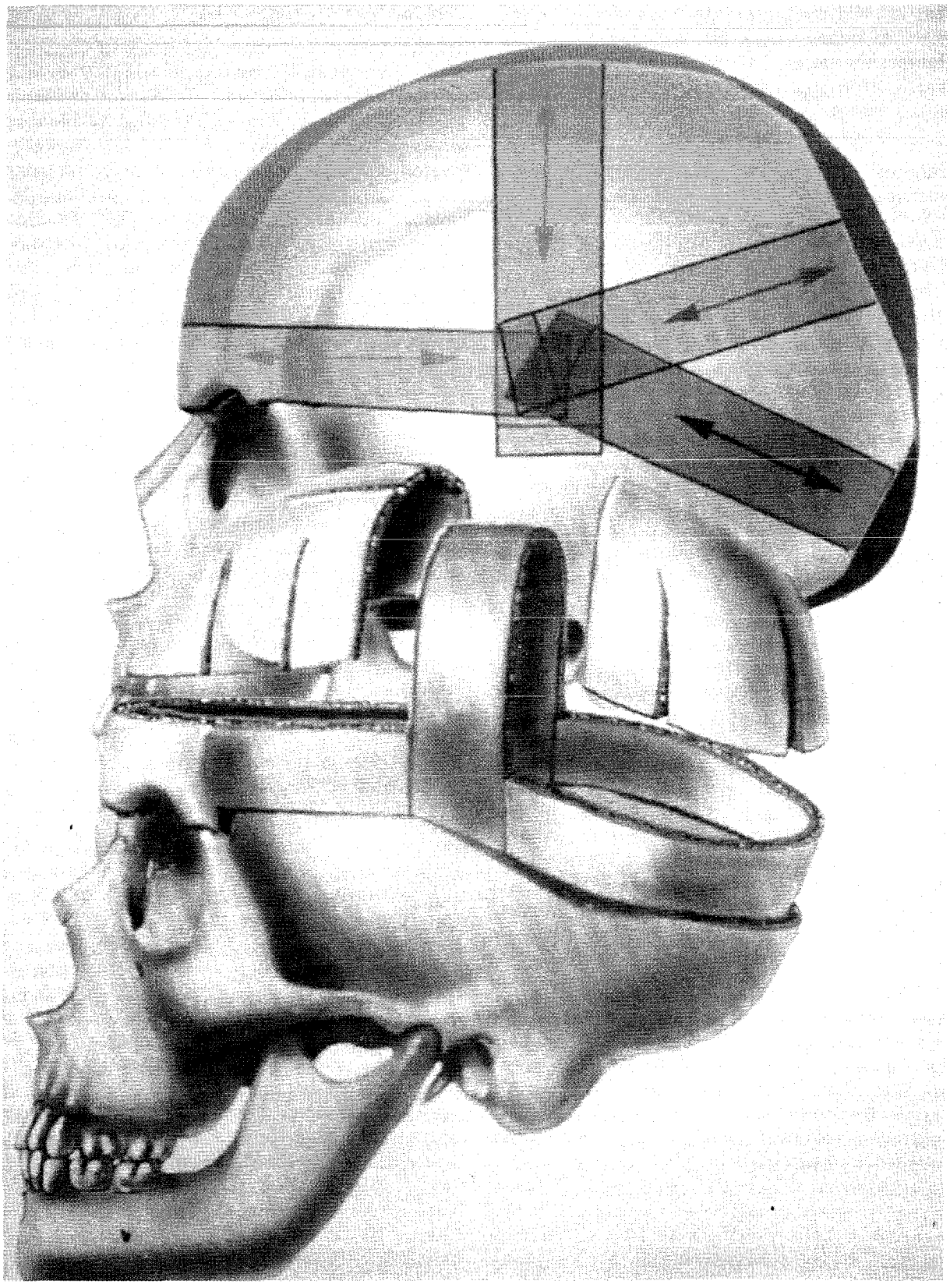
THE BANDEAU CONCEPT

Reconstructing the proper balance and harmony of the face begins with the manipulation and positioning of the forehead by use of a bandeau. The bandeau is the key architectural component of craniofacial reconstruction and is frequently treated as a separate unit in frontocranial reconstruction.[18] In frontocranial remodeling, the bandeau includes the supraorbital rim, glabellar region, and the nasofrontal angle which together form the lower portion of the forehead. The normal supraorbital bar is obliquely and anteriorly directed, forming an angle of 90 to 120 degrees with the nose. The forehead begins to curve approximately 1 cm above the orbital rim and passes vertically and gently backwards towards the vertex.

In young patients before the development of the frontal sinus, the supraorbital bandeau can be removed en bloc and manipulated in various ways to achieve the desired correction. It can be advanced, recessed or tilted depending upon the deformity. At times the bandeau is so distorted that it is not possible to change its shape sufficiently to achieve a good aesthetic result. In such cases the original bandeau is discarded and a new one is harvested from an appropriately shaped portion of the cranium. New variations for cutting and advancing of the bandeau are used occasionally to decrease the exposure to the nasopharynx in combined forehead and midfacial movements, thereby decreasing the theoretic risk of infection.

In addition to improving the appearance of the frontocranial region, advancement of the supraorbital bar and forehead can also result in functional improvements. Intracranial hypertension is frequently seen as an accompaniment of various craniofacial malformations, being found not only in complex but also in simple craniosynostosis. Intracranial hypertension has been held responsible for optic nerve damage with loss of vision leading to blindness, and has been implicated as a factor in mental retardation. Advancement of the forehead and anterior cranial base relieves intracranial hypertension by increasing intracranial volume, thus possibly averting these problems. The orbital advancement expands the orbital volume and aids in the correction of exophthalmos, such as found in Crouzon's and Apert's syndromes.

The key to total cranial-vault reconstruction lies in the use of the forehead bandeau, midcranial-vault bandeau and the occipital bandeau (Fig.7). The midcranial-vault bandeau is helpful in



(Fig-7) The bandeaus are designed and cut while the intracranial and orbital structures are protected. These are then removed, remodeled, and replaced providing structural stability and symmetry and guiding the changes in cranial-vault proportions and volume. The adjacent bones below and above the bandeau are cut and expanded to meet the new shape and position of the three bandeaus

(Fig-8a) Preoperative frontal view of a 2½ - year-old patient with scaphocephaly

(Fig-8b) Postoperative frontal view 6 months after correction using a modification of this technique

adjusting the vertical height of the cranium. The occipital bandeau changes the height and posterior projection in the occipital region and corrects deformities of the posterior cranial vault and fossa. Applying these techniques for total cranial vault expansion or reduction improves results in most difficult cases (Fig.8).

BONE GRAFTING IN CRANIOFACIAL SURGERY

The craniofacial bones are a mosaic of structurally and aesthetically significant building blocks which contribute to the protection, support and contour of the face and cranium. Changes in the shape of these bones occur rapidly until the completion of puberty and continue to occur throughout the adult life albeit at a slower rate as a result of interaction between the overlying periosteal-muscle-soft tissue matrix and the underlying skeletal framework. Therefore craniofacial surgery is concerned with reconstruction of the hard tissue contour which in turn significantly influences the overlying soft tissue mask.[19,20]

Porous Hydroxyapatite

Autogenous bone grafts when particularly used as an onlay undergo unpredictable resorption and remodeling which leads to disappointment and frustration about the overall aesthetic contour. This has led to an ongoing search for suitable biological and non-biological substitutes which could circumvent these problems. Porous hydroxyapatite has been found to be an useful alternative to autogenous bone as an onlay and inlay bone graft substitute on some occasions.[21,22] The senior author (KES) has studied bone substitution materials since the early 1970s when he was Director of the Biomaterials Maxillofacial Laboratory at the Veterans Hospital of the University of Texas Health Science Center. The Interpore 200 porous hydroxyapatite is a biocompatible implant material derived from specific marine coral. The intraoperative care of the hydroxyapatite requires shaping with a diamond tripped burr attached to a high speed water-cooled drill. After shaping, the sculpted implant must be cleansed with saline irrigation and brushed to evacuate the micropores. The biggest use of this material today is in vertical lengthening of the maxilla as an interpositional graft because its resorption rate is less than 1% per year. We have recently used hydroxyapatite paste (CRS) for cranial vault contour defect filling in the fully grown skeleton. It is contraindicated in the growing skull.

Demineralized bone

Much research in experimental animals and patients has now been conducted using demineralized bone and bone morphogeneic protein. Our search for a good bone substitute led us to the P.B.B. Our research and observation of the material since 1990 has demonstrated that it can be osteoinductive as well as osteoconductive. In experimental studies it has shown to induce new bone formation. In clinical practice when we have re-operated on patients upto 3 years later, calcification plus remodeling of new bone has been seen. Secondary contour defects in craniofacial surgery occur as a result of bone remodeling, resorption and changes with growth.

Demineralized bone has in the recent few years been increasingly used for resurfacing cranial defects, cranial and facial bony augmentation or replacement, and as a filler material in cranial vault remodeling at our centre.[23,24,25] The demineralized perforated bone matrix (DPBM) is prepared by the Pacific Coast Tissue Bank in Los Angeles, California according to a detailed protocol that simultaneously allows a high degree of safety and excellent biological compatibility. Prior to harvesting tissue, potential donors are screened for infectious and malignant diseases including HIV. Tibial bone after purification is cut into triangular prisms and then is subjected to microperforation using 500μm drill bit. Next the bone is treated with ethanol to remove lipids and then is demineralized by suspension in 0.6N hydrochloric acid at 6° C with continuous mechanical agitation. Demineralized bone is extensively used at our centre for coverage of bony defects and augmentation of cranial vault, forehead, temporal/malar, nasal/paranasal/, and orbital/maxillary complex areas.

Evaluation of results was possible using postoperative biopsy which was done in these cases while performing secondary procedures. Biopsy demonstrated some areas of new bone formation whereas other implants remained in the same condition as they were when initially inserted. Our experience with DPBM indicates that it presents a positive alternative in patients in whom there is need for extra bone implants which cannot be obtained from the patient. More than 547 patients have received 1,333 implants in various type of craniofacial deformities. The material is easy to work with; it is pliable yet also stable enough to adequately protect the brain.

DISTRACTION OSTEOGENESIS

Distraction osteogenesis is a process by which new bone is generated across the gap of a low energy osteotomy which is separated in a progressive and gradual fashion without disrupting the vascular supply. Introduced clinically for craniofacial surgery in 1992 by McCarthy,[26] this technique allows a controlled method of simultaneous new bone formation and soft tissue expansion, which is more stable and superior to performing classic osteotomy and bone grafting.

Distraction osteogenesis has been performed on the mandible since 1993 at our centre. Over 60 cases have undergone distraction procedures on hemimandibles and midfaces. Mandibular distraction was used on patients with craniofacial microsomia, Treacher Collins syndrome, traumatic mandibular deformities and Romberg's disease using multi-bidirectional and unidirectional devices. The average mandibular distraction distance is 21.3mm. Recently midfacial distraction has been performed successfully on six patients with severe midface hypoplasia secondary to syndromal craniofacial anomalies. Distraction osteogenesis will someday become a major treatment modality. It awaits development of better three dimensional distraction devices used internally or externally.

INTERNAL RIGID FIXATION IN CRANIOFACIAL SURGERY

Fixation has greatly been facilitated with the introduction of miniplates and the technique used for miniplate fixation varies from surgeon to surgeon. Our preferred method at this time is the Medicon craniomaxillofacial titanium miniset, which increases the adaptability of the plates during surgery. Our complication rate for miniplate fixation is low, occasionally palpation of these plates makes it necessary to remove them at a secondary setting. These plates should not be left in infants or young children permanently because they may impede growth or migrate. This has led to the development of resorbable fixation.

RESORBABLE FIXATION IN CRANIOFACIAL SURGERY

Resorbable plates and screw system has added a further dimension and refinement in the practice of modern craniofacial surgery. The implants are malleable and designed to secure bone fragments and facilitate healing. They are made from a special polylactic acid copolymer and are available in a variety of appropriate sizes. Heating the material to 55 degree C in the warming basin makes it completely pliable and conformable to any craniofacial bony structure. At body temperature the implants regain total rigidity, moreover cutting of the plate to any required size is easy using a battery operated cautery tip. Unlike titanium implants, these implants completely resorb in one to three years, obviating the need for removal procedures while avoiding long term palpability and temperature sensitivity. These materials are non-toxic, non-irritating and amorphous avoiding growth restriction in pediatric patients.

Non-resorbable plates in young children have to be removed because of the risk of migration into the dura or brain. Morever plate exposure and infection are other risks with the conventional system which may necessitate their removal.

To conclude modern craniofacial surgery has come a long way from where it originated over thirty years back through the pioneering work of Tessier. With advancements in anaesthesia and drugs, controlled hypotensive anaesthesia allows major craniofacial reconstructions with minimal blood loss and without the need of blood transfusions. The modern fixation systems including the resorbable systems allow rigid fixation of the osteotomized skeleton in desirable position. The development of these new techniques has opened numerous and unlimited possibilities of treating the complex and disfiguring craniofacial deformities which were previously considered inoperable.

With the start of a new century, we are now looking towards improved international availability of new technology, team approach and establishment of a few centres of excellence dedicated to treatment of craniofacial deformities. We have proven in Dallas at our institute over the past 12 years, that a dedicated team with dedicated effort to craniofacial surgery yields improved results and reduced number of operations, morbidity and mortality. All children and adults with congenital and other craniofacial deformities should have access to the excellence of care in every part of the world and each country with a significant population of 20 million or more.

Our goal is the global treatment of all children with craniofacial deformities, through the internet, teleconferencing and with documentation of improved treatment, we can achieve excellence. An international network of centres of excellence dedicated to craniofacial care is our goal.

References

1. **Tessier P. Dysostosis craniofaciales.** Ann Chir Plast 1967;12:774-783.
2. **Tessier P. Orbital hypertelorism:** I. Successive surgical attempts, material and methods, causes and mechanism. Plast Reconstr Surg 1974;53:1.
3. **Tessier P.** The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis, Crouzon's and Apert's diseases. Plast Reconstr Surg 1971;48:419-442.
4. **Whitaker L, Munro I, Jackson I, Salyer K.** Problems in cranio-facial surgery. J Maxillofac Surg 1976;4:131-136.
5. **Salyer and Bardach's Atlas of Craniofacial and Cleft Surgery.** Vol I and II. Lippincott Raven publishers, Philadelphia, 1999.
6. **Bardach JB, Salyer KE.** Surgical Techniques in Cleft lip and palate. Chicago: Mosby Year book Publishers, II ed, 1991.
7. **Salyer KE.** Early and late treatment of unilateral cleft nasal deformity. Cleft Palate Craniofacial J 1992;296:556-569.
8. **Salyer KE.** Primary correction of the unilateral cleft lip nose: a 15-year experience. Plast Reconstr Surg 1986;77: 558-566.
9. **Agarwal R, Bhatnagar SK, Pandey SD, Singh AK, Chandra R.** Nasal sill augmentation in adult incomplete cleft lip nose deformity using superiorly based turn over orbicularis oris muscle flap. An anatomical approach. Plast Reconstr Surg 1998;102(5):1350-1359.
10. **Obwegeser H.** Indications for surgical correction of mandibular deformity by sagittal splitting technique. Br J Oral Surg 1976;14:150-155.
11. **Dal Pont G.** Retromolar osteotomy for the correction of prognathism. J Oral Surg 1961;19:42-47.
12. **Salyer KE.** Techniques in aesthetic craniofacial surgery. New York: Gower Medical Publishing, J.B Lipincott, 1989.
13. **Converse J.** Ocular hypertelorism and pseudo-hypotelorism: Advances in surgical treatment. Plast Reconstr Surg 1970;45:1.
14. **Jackson I, Munro I, Salyer K, Whitaker LA.** Atlas of craniofacial surgery. St Louis:Mosby,1982.
15. **Salyer KE, Hall CD, Joganic EF.** Lamellar split osteotomy: a new craniofacial technique. Plast Reconstr Surg 1990;86: 845-853.
16. **Salyer KE.** Malar augmentation using lamellar split osteotomy. J Craniofac Surg 1994;5(2):116-123.
17. **Anderl H, Muhlbauer W, Twerdy K, Marchac C.** Frontofacial advancement with bony separation in craniofacial dysostosis. Plast Reconstr Surg 1983;71:303-7.
18. **Salyer KE, Hall JD.** Bandeau - the focal point of frontocranial remodeling. J Craniofac Surg 1990;1:18-31.
19. **Salyer KE, Taylor DP.** Bone grafts in craniofacial surgery. Clin Plast Surg 1987;14(1):27-35.
20. **Salyer KE, Taylor DP.** Bone grafting in cleft deformity: A craniofacial approach. Aust N Z J Surg 1987;57:67-76.
21. **Salyer KE, Hall CD.** Porous hydroxyapatite as an onlay bone graft substitute for maxillofacial surgery. Plast Reconstr Surg 1989;84(2):236-244.
22. **Salyer KE.** Synthetic biomaterials for bone. Current Therapy in Plast Reconstr Surg, 1989.
23. **Salyer KE, Gendler E, Menendez JL, Simon TR, Kelly KM, Bardach J.** Demineralized perforated bone implants in craniofacial surgery. J Craniofac Surg 1992;3(2):55-62.
24. **Salyer KE, Gendler E, Squier CA.** Long term outcome of extensive skull reconstruction using demineralized perforated bone in Siamese twins joined at the skull vertex. Plast Reconstr Surg 1997;99(6):1721-1726.
25. **Salyer KE, Bardach J, Squier CA, Gendler E, Kelly KM.** Cranioplasty in the growing canine skull using demineralized perforated bone. Plast Reconstr Surg 1995: 96(4): 770-779.
26. **McCarthy J, Schreiber J, Karp N, Thorne C, Grayson B.** Lengthening the human mandible by gradual distraction. Plast Reconstr Surg 1992;89:1-8.

Authors

Kenneth E. Salyer MD, FACS, FAAP, FICS, Director

Rajiv Agarwal, MS, DNB (Surg), MCh, DNB (Plast Surg), MNAMS
Craniofacial Fellow

International Craniofacial Institute, Dallas, Texas, USA

Requests for reprints to Dr. Kenneth E. Salyer, International Craniofacial Institute, 7777 Forest Lane, Suite C-717, Dallas, Texas 75230, USA.