Craniofacial Surgery:

Present and Future

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The possibilities for radical craniofacial restructuring have increased dramatically in the past 6 years with the development of craniofacial surgery. The field developed from a background of patients with major craniofacial birth defects allowing orderly planning and expansion to correction of a multitude of other craniofacial structural problems. The procedures concentrate upon changing the skeletal structures using extensive subperiostial dissection of soft tissue, and adding bone to fill in areas of deficiency. There are three grades of complexity in craniofacial procedures. After extensive soft tissue sub-periostial stripping about the orbits and upper face, the simplest form consists of onlay bone grafts. The next most complicated involves osteotomies to shift the face into a more normal position. In its most complicated form, abnormal proportions of bone are removed and the orbits or cranium are shifted into a new or normal position. We have had experience with 69 patients since September, 1972. Thirty-six have had intracranial procedures. Infection has been the most serious problem, and there have been no instances of death or blindness. A number of lesser problems occur. Future applications of craniofacial surgery are appearing with great frequency as more experience is gained with its uses. It has particular application in acute and late reconstruction of patients with traumatic defects about the face. Preventive osteotomies are an area with great potential, by releasing stenotic areas of bone and allowing the developing brain to mold the upper face and orbits. There is also applicability in surgery of tumors about the craniofacial structure and in cosmetic surgery.

THE POSSIBILITIES for the radical alteration of the craniofacial structure have increased dramatically in the past 5 to 6 years. Following the pioneering efforts of Dr. Paul Tessier,[10,11] several centers in North America have developed expertise, and large teams for doing this type of surgery.

As experience accumulates, the ramifications of the surgery, and the possibilities engendered by it are increasing. Almost all parts of the craniofacial structure can now be radically altered in one way or another. The main force leading to the development of the field has been the possibilities for radical reconstruction of birth defects. These problems have provided a backlog of cases with identifiable and rather consistent defects, allowing planning to proceed on an orderly and sequential basis. It has, however, been evident from the beginning that the number of individuals with birth defects to be reconstructed by these techniques represents a rather small number. Indeed, it has been estimated that there are perhaps 1200 identifiable infants born with these deformities in the United States each year.[7] In spite of the relatively small number of patients with birth defects to be reconstructed, the techniques are, however, widely applicable.

Craniofacial skeletal structural changes are the foundation of the surgery, with soft tissue corrections following and emphasized secondarily. The unique features of the surgery include: 1) wide exposure of the craniofacial skeleton through a coronal incision and sub-periosteal dissection; 2) freeing the entire orbital contents back to the apex; 3) dissecting the soft tissues of the face off their

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TABLE 1. *Reasons for Surgery*

Craniofacial dysostoses (Apert, Crouzon, plagiocephaly)	22
Hypertelorism	15
Mandibulofacial dysostosis & hemifacial microsomia	12
Other (tumors, trauma)	20

TABLE 2. *Surgical Approaches*

Intracranial	36
Extracranial	
with facial advancement	11
without advancement	22

skeletal attachments except along the inferior edge of the maxilla, the mucosal surface of the nose, and the attachments of the lacrimal apparatus and supraorbital, infraorbital, optic and sometimes olfactory nerves. The simplest craniofacial surgical procedures then consist only of adding bone where it is deficient. Large amounts of rib or iliac bone have been consistently successfully added to deficient areas, for correction of such deficiencies.

The next most complicated form of craniofacial surgery, involves detachment of hypoplastic areas of bone and movement into a more normal position. This category includes major mid-face advancements for mid-face hypoplasia, forehead retrusion, or deficiencies in other areas of the skull. The repositioned segments are then held in their new position by bone grafts, or simply wired.

Probably the most complicated of the craniofacial surgical procedures involves removal of abnormal or excessive bone or soft tissue sometimes with replacement in other areas. This has particular application in patients with hypertelorism, with excess bone and soft tissue between the orbits and lateral displacement of the orbits. The bone and soft tissue between the orbits is excised and the orbits moved toward each other in the midline. The defects left bilaterally are then filled with bone. The bony nose usually is completely reconstructed with bone grafts.

These extensive procedures have been described in detail by Tessier[10–13] and others.[1–3,6,8] Other congenital structural anomalies about the face are corrected using some of the principles of the major craniofacial procedures. Of particular importance in this regard because of its frequency is hemifacial microsomia. It has been estimated that 1 patient in 5600 live births has hemifacial microsomia.[7] These corrections generally involve work about the lower half of the face, concentrating particularly on the jaws.

Our Experience

Our experience has been with 69 patients from September, 1972, through February, 1976. The age range has been 2 weeks to 51 years, but with only three patients over the age of 30 years. Eight have been less than two years of age, and the bulk of the remainder were in the 3–15 year age group. We are doing increasingly early correction of those individuals with deformities involving the orbits or skull, as are others.[5] The benefits of early correction seem numerous. When, however, movement of the jaws is a necessary component of the correction, there may be good reason for delay until the mixed dentition stage in most cases. Whether or not the jaws continue to grow normally, and relapses are strictly technical in nature, remains unproven. It seems certain that the technical difficulties of fixing jaws in a new position are greater in the age group before mixed dentition starts. Therefore, except in unusually severe deformities, jaw movement at present is probably best delayed until the 7 to 9 year age group or later.

The length of hospital stay has ranged from 6 to 44 days, the usual period of stay being 8–12 days. If there are no complications, virtually all patients are discharged by 9–10 days postoperatively. The diagnostic categories treated by us are listed in Table 1. Patients included in the series had dissections requiring elevation of large amounts of soft tissue subperiosteally, and with a common denominator of at least unilateral total freeing of orbital contents except at the apex and lacrimal apparatus region. In most instances a coronal incision was used, though in some instances of hemifacial microsomia and trauma, scars or modifications of the coronal incision were used. The approaches used are indicated in Table 2.

Quality of end result is not yet measurable. The great majority of patients report that they are "not stared at as often," are "much happier," "feel more outgoing," and "are pleased with the result." These are highly subjective measurements, though they are reflections of probably the most important goal of the surgery. The attempt at altering a severely deformed face to one that is normal or near normal is so important psychosocially in our society that this is certainly the number one objective.[14] Two patients have had measured improvement in vision, and several patients have had simultaneous release of intracranial pressure when a craniofacial procedure was done at the time of the craniectomy. Jaw relations have been

TABLE 3. *Problems*

Bleeding (>1/2 blood volume)	9
Sensory nerve loss (supraorbital or infraorbital)	3
Sixth nerve palsy	2
Seventh nerve palsy (frontal branch)	2
Pneumothorax	11
Soft tissue necrosis	4
Strabismus	Many
Partial bone graft loss	?
Prolonged hospitalization	6

TABLE 4. *Complications*

Death	0
Blindness	0
Infection	
Minor	3
Serious	6
CSF leaks	3
None	43

TABLE 5. *Future Applications*

Cosmetic
Tumors
Ressection
Reconstruction
Trauma
Acute
Late
Preventive Osteotomies

improved consistently when the jaws have been moved, though small degrees of relapse are common.

Problems associated with this surgery have been frequent, and some seem at this point to be inseparable from the surgery.[9,15] Soft tissue defects and scarring associated with tissue shifts and removal of excess tissue are a major unsolved problem. Spreading in the medial canthal region so that postoperative telecanthus occurs is frequent following correction of hypertelorism and the method of preventing this is not yet known, in spite of adequate construction of a bony structure. Nasolacrimal apparatus obstruction is a problem that occurs postoperatively on occasion, and may require dacryocystorhinostomy for correction. This results from dissecting out the nasolacrimal apparatus from the lacrimal groove, on occasion undoubtedly with damage to that structure during orbital or mid-face advancement. We had a 12% incidence of nasolacrimal apparatus obstruction present preoperatively and an additional 6% postoperatively. Temporary sensory nerve loss occurs, though we have had no instances of permanent major sensory nerve losses. Nerves involved have been the infraorbital and supraorbital divisions of the Trigeminal. Strabismus following dissection about the orbits is a frequent problem, and minor degrees probably occur in every patient. Most have been self-correcting, and there have been only three patients requiring extra ocular muscle surgery following a craniofacial procedure. Bleeding can be extremely difficult to control, and occurs often with osteotomies into areas that are not accessible to the usual means of hemorrhage control. In spite of this we have had only two patients who have bled more than one volume of blood. Bleeding is generally controlled with the aid of hypotensive anesthesia. Partial bone graft loss through absorption probably occurs in many patients, and perhaps to some extent in all. At this point it is impossible to meas-

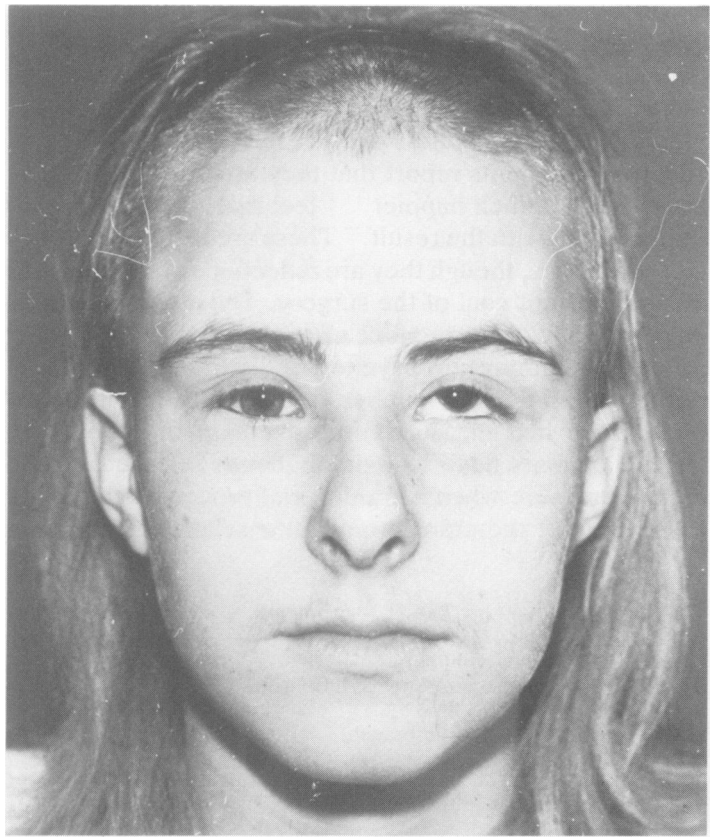

FIG. 1a. J.A., two months after injury. Note depressed forehead segment and left eye abnormality associated with severe orbital fracture.

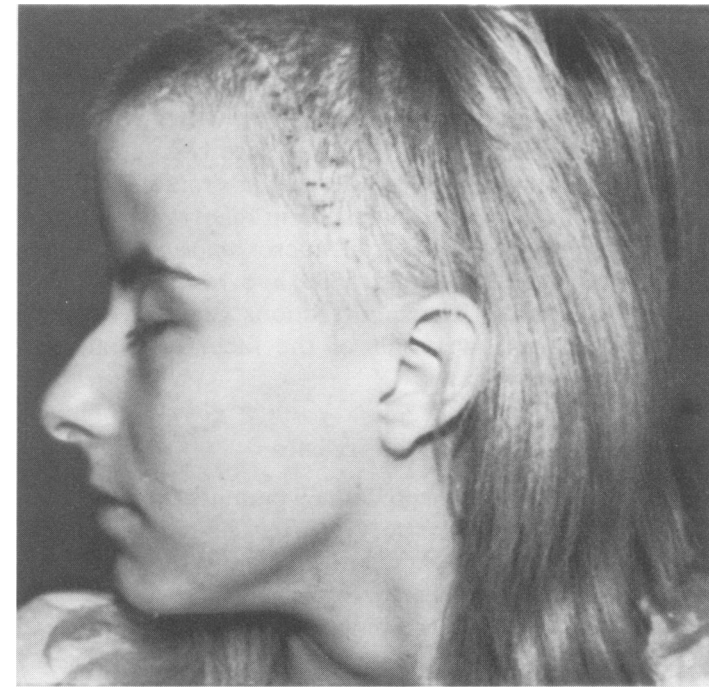

FIG. 1b. J.A., two months after injury—depressed forehead-frontal bone and nasal bone.

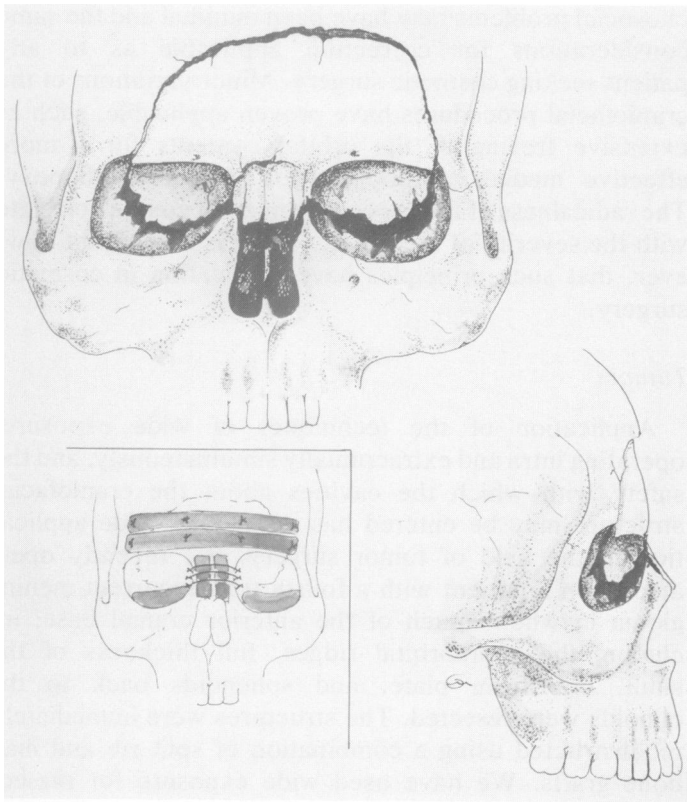

FIG. 1c. J.A., bony defects. Dark areas are onlay rib grafts through coronal incision.

ure. Facial growth following these corrections in children is an unknown quantity. However, increasing evidence indicates that it is probably normal. Certainly in our experience, growth about the upper half of the face including that about the orbits is benefited by early surgery as documented by several patients with isolated craniostenosis, who had surgical procedures done at a very early age. At this point, growth of the lower half of the face is uncertain following these surgical procedures. Problems encountered are listed in Table 3.

Our complications are listed in Table 4. Other major centers have reported a mortality rate of 1–2%,[3] and a number of instances of blindness have been reported, following craniofacial procedures. We have not encountered either. Our biggest problem has been infection. We have had 6 serious infections, prolonging the patients' hospitalization and requiring surgical drainage, and in 4 instances removal of part or all of the bone grafts. One patient developed a brain abscess, and eventually had removal of the entire frontal bone flap, and superior halves of the orbital grafts. Death seemed imminent at the time, but she is now 1½ years following surgery, and is finally almost free of any evidence of infection.

Three other patients have had osteomyelitis of the frontal bone flap or in portions about the orbit, smouldering along for up to one year, before finally resolving. Each of these patients will require another reconstructive procedure.

There were three instances of minor soft tissue infections. In these patients there was no prolongation of hospitalization, and they were treated with antibiotics with no alteration of the end result. Three patients had cerebrospinal fluid leaks, requiring reoperation. One was the patient who developed the brain abscess and extensive osteomyelitis. The other two were reoperated and the end result does not seem to have been altered by the extra surgical procedure.

Most deaths that have been reported have been associated with intracranial procedures and brain swelling, airway problems, or bleeding. These should all be potentially preventable, and at present the only unpreventable serious complication would seem to be that of infection. The infection rate is high (about 7% serious) but still seems low when one considers the large amounts of free bone grafts and frequent entry into the nasal and sinus cavities as well as the oral cavities while often working within the cranial cavity. We have used high doses of

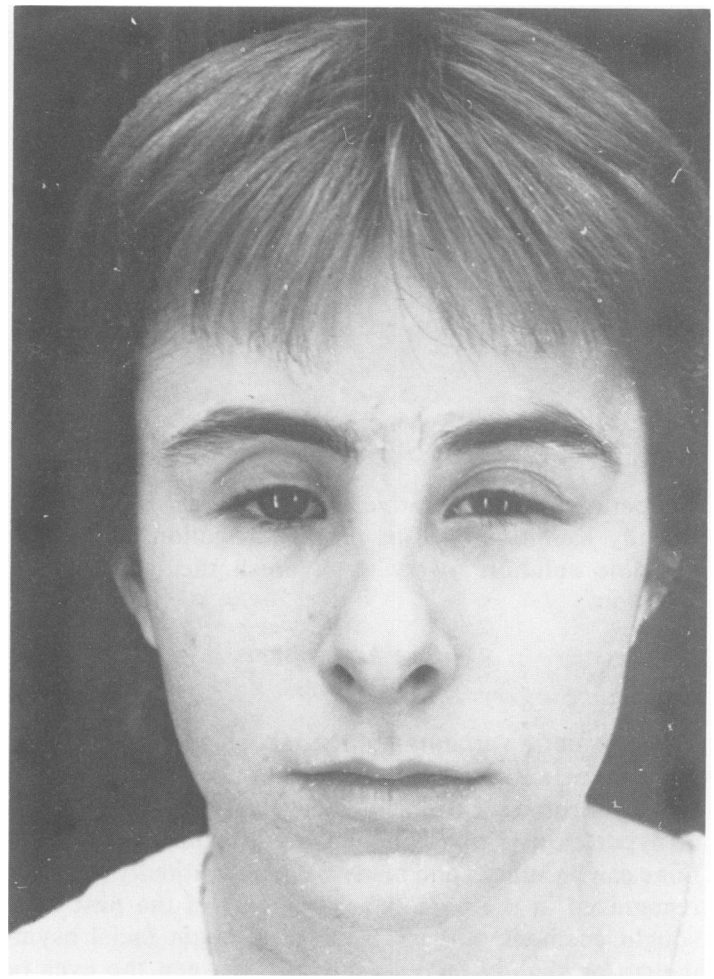

FIG. 1d. J.A., 1 year postoperative.

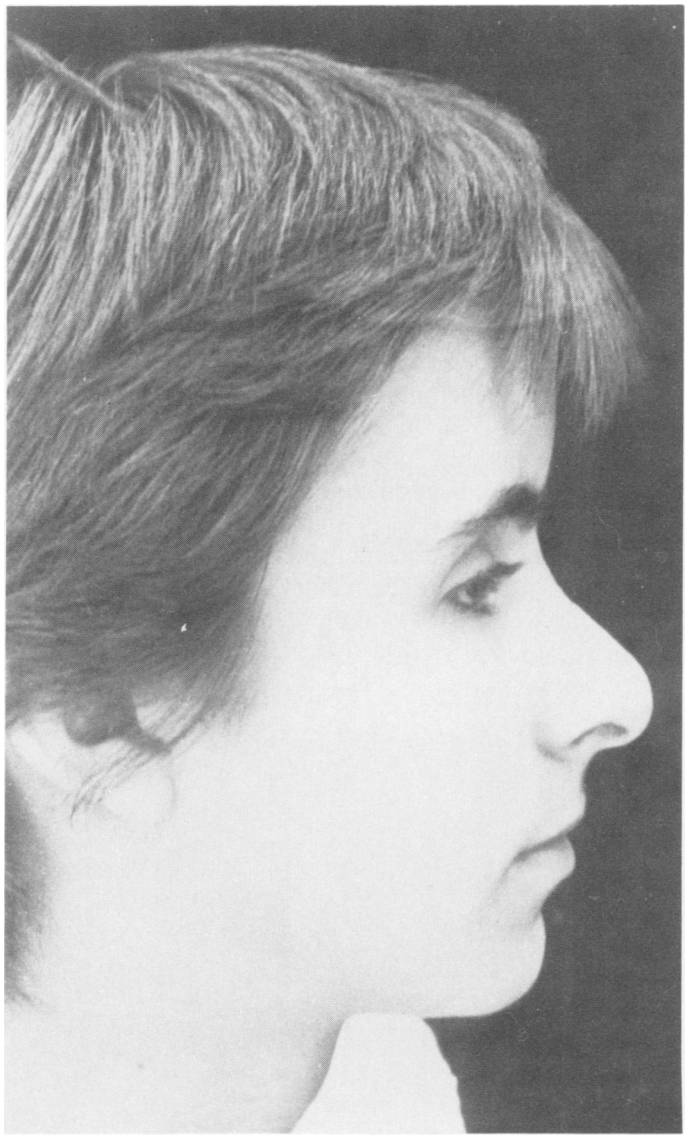

FIG. 1e. J.A., 1 year postoperative.

preoperative, intraoperative and postoperative antibiotics (usually Keflin) as well as profuse irrigation with dilute Betadine solution to try to diminish the incidence of infection.

Future Applications

Cosmetic Surgery

Many subtle variations of the recognizable syndromes are now being noted. The spectrum of manifestation of each syndrome is extremely wide. Especially in instances of hypertelorism and hemifacial microsomia, manifestations can be subtle, and many have undoubtedly gone unrecognized in the past. Many patients in the past have sought cosmetic surgery because of slight facial asymmetry, or slightly increased width between the eyes or breadth of the nasal bridge, and been classified as aesthetic surgery patients. In these individuals psychosocial problems may have been minimal and the same considerations for correction applicable as to any patient seeking cosmetic surgery. Minor variations of the craniofacial procedures have proven applicable, such as extensive freeing of the orbital contents for a more effective medial canthoplasty or medial canthopexy. The radicalness of the procedure must of course correlate with the severity of the defect. There is little doubt however, that such principles have application in cosmetic surgery.

Tumors

Application of the techniques of wide exposure, operating intra and extracranially simultaneously, and the safety with which the cavities about the craniofacial structure may be entered has potentially wide application in the field of tumor surgery. We recently operated upon a patient with a fourth time recurrent meningioma in whom much of the anterior cranial base, including the supraorbital ridges, full thickness of the skull, cribriform plate, and sphenoids back to the clinoids were resected. The structures were immediately reconstructed using a combination of split rib and iliac bone grafts. We have used wide exposure for ressection of hemangiomas, and fibrous dyspoasia in 6 cases. Malignancies in the region of the orbits, extending into the ethmoid or sphenoid sinuses probably are less applicable. They are in general so extensive by the time they are recognized that in the few we have encountered it has been impossible to be certain enough of having achieved cure to go ahead with bony reconstruction at that time. In the presence of earlier malignancies however, it is conceivable that the orbit or anterior cranial base can be resected in the same fashion as previously described, and reconstructed immediately with appropriate bone grafts and soft tissue.

Trauma

Trauma is an area of wide future applicability. We have had experience with reconstruction of major defects about the orbits and upper face secondarily in 14 patients. These were defects that involved displacement of half or more of the orbit originally, and for one reason or another were not corrected acutely. In several instances, brain damage, a cerebrospinal fluid leak, or other major trauma had prevented correction of the facial structural deformity at the time of the original injury, and it remained as the single dominant problem after everything else had been corrected. Using the concepts of wide subperiosteal exposure, large amounts of bone, and in one instance total movement of an orbit from an intra and extracranial approach into a more normal position were used. Also the concepts of doing everything possible at one operative procedure is applicable. Associated soft

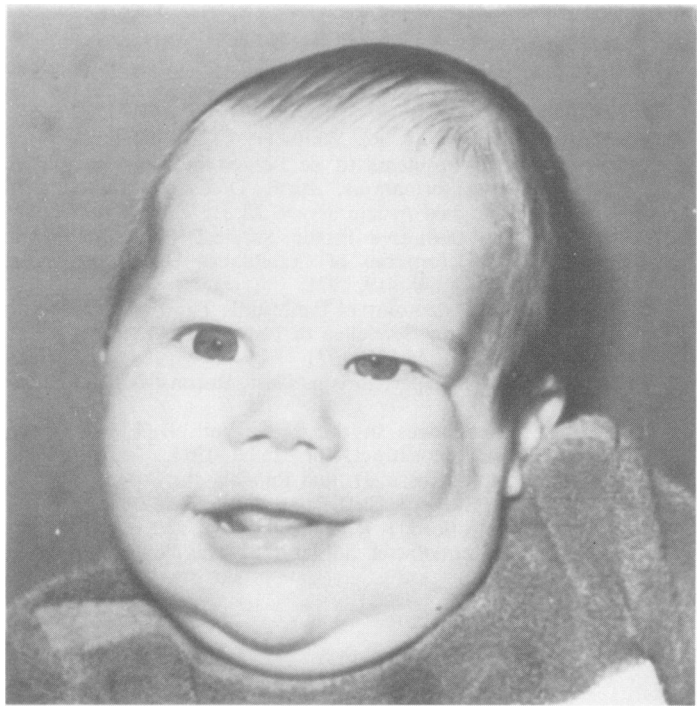

FIG. 2a. Isolated left craniofacial stenosis with deformity of cranium, left temporal and left zygomatic areas.

tissue and bony shifts can be more readily accomplished when the entire dissection is in progress. It is possible to correct bony deficiencies, canthal displacements, orbital and ocular displacements, lacrimal apparatus problems, and soft tissue defects at one operative procedure.

Case Reports

A 16-year-old girl is shown in Figs. 1a to e, two months following an automobile accident with depression of her frontal bone and extensive fractures about both orbits. She had been comatose with a cerebrospinal fluid leak in another hospital and was transferred for handling of those problems plus correction of her fractures. At two months following the accident, bone grafts were used to correct areas of bony deficiency. Because of the difficulties encountered in moving comminuted segments of facial bones, onlay bone grafts have proven preferable. Since facial bones tend to heal with fibrous union only, and come apart into many segments if dissected free, it is a rare situation that en bloc movement can be done as successfully as with onlay bone grafts. The principles of total freeing of the orbital contents sub-periosteally, moving canthi into more normal position, holding the soft tissue contents in position with wires and appropriate bone grafts, and restructuring areas of bony deficiency are important.

In acute upper facial trauma, the same techniques have been used effectively, to give adequate exposure and control of the problems associated with such severe injuries. We have recently operated upon two such patients within 12 hours of injury. A coronal incision, wide exposure, and extensive replacement of the fractures were done successfully in each instance.

Preventive Surgery

The final major area of future application is the field of preventive osteotomies. The concept is that of allowing

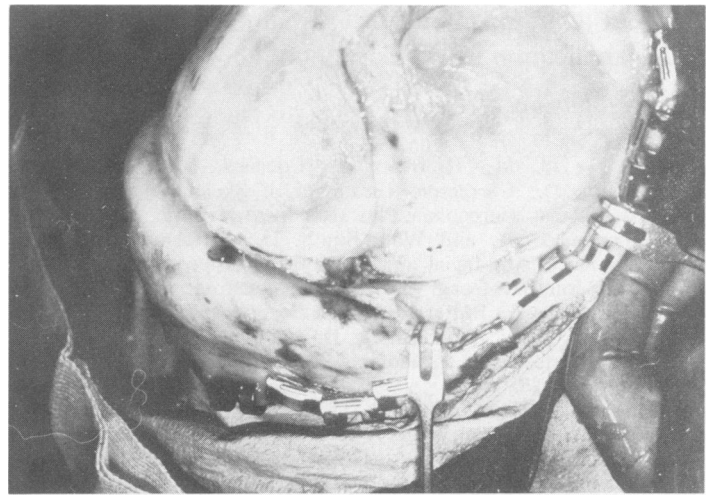

FIG. 2b. Note area of bone removed in central portion to release craniostenosis.

the rapidly expanding brain in the infant to exert its normal influence on upper facial growth. In instances where craniofacial stenosis occurs, the normal influence of the brain on upper facial growth is altered. If such abnormally premature fusion can be released before age five to six months, an extremely beneficial effect may be possible.

We have operated in conjunction with neurosurgeons on six such patients. Four were operated upon six months ago or longer, and the striking changes of the face following such extensive release is seen in the patient in Figs. 2a to c. The procedure is much simpler, shorter, and does not involve bone grafting, but only a resection of bone. In its ideal form, the cranium and upper face will then be molded by the rapidly developing brain.

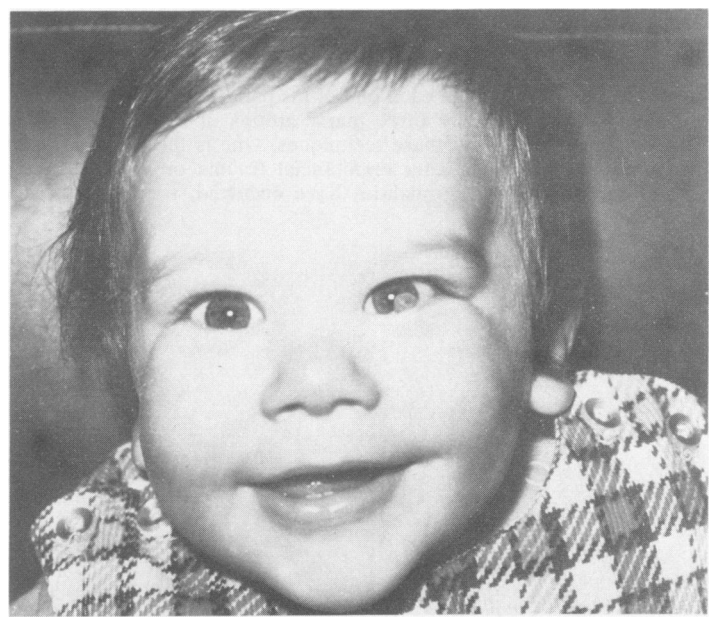

FIG. 2c. 6 months postoperative with improving deformities.

It seems likely that such preventive measures may have wider application as we learn more about facial growth.

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Discussion

Dr. John Burke (Boston, Massachusetts): For many years the isolated techniques and approaches needed in this form of surgery were well known to the neurosurgeon and the plastic surgeon, but the actual treatment of these malformations remained largely confined to limited subcranial procedures. It remained for Dr. Paul Tessier, of Paris, to recognize that these anomalies could be safely and completely corrected by using a combined intracranial-extracranial approach.

It is to Dr. Tessier's great credit that he took it upon himself personally to instruct interested groups, primarily in this country, in these very difficult procedures. Further, the multidisciplinary team needed for the correction of these problems was rapidly formed, and in this Dr. Whitaker's group is one of the leaders.

Today, the craniofacial group is the logical outgrowth of a well-developed cleft palate team, such as has existed at the University of Pennsylvania for years, under the vigorous direction of Dr. Peter Randall.

We believe that the surgical success that Dr. Whitaker has analyzed for us today can only reflect a high degree of leadership and personal involvement on his part in each one of his patients.

It seems that there are three major groups of malformation susceptible to correction by these techniques. One is the stable form of deformity such as seen after craniofacial trauma or ablative tumor surgery. Once these abnormalities have occurred, the malformation does not progress.

The second is the congenital malformation, which has a relatively predictably constant evolutionary pattern.

The third is the group of patients which includes unstable deformities brought about by such conditions as lymphangioma, arteriovenous fistula, fibrous dysplasias, etc. These conditions are produced by capricious and unpredictable changes that are constantly presenting new problems. Dr. Remensnyder wonders if Dr. Whitaker has any thoughts on the management of this third and unstable group.

Dr. Linton A. Whitaker (Closing discussion): I'd like to try to clarify that stable lymphangioma discussion, Dr. Burke. I'm not quite sure I understood the question.

Dr. Burke: I think Dr. Remensnyder's question was, since these deformities created by lymphangioma or arteriovenous fistulas are unpredictably unstable, do you have any thoughts about their management?

Dr. Whitaker: We have used the wide exposure to resect lymphangiomas in several instances. As to stability and progression afterwards, I don't know the answer.

As to whether or not they would ever progress following resection, I think it is the same consideration you would have in doing the resection in any other fashion, with cystic hydromas or lymphangiomas.