Pediatric Facial Plastic and Reconstructive Surgery

James D Smith, Robert M Bumsted

Foreword

This foreword to the new book *Pediatric Facial Plastic and Reconstructive Surgery* gives credit to Drs Robert Bumsted and James Smith for an excellent job with a complicated, extensive, and often underevaluated subject, namely this type of surgery as it applies to children.

They emphasize the importance of teams of different specialists in certain instances and recognize that many of these specialists have become superspecialists within their specialty group. They also recognize that some cases may include specialists from other special facilities in the more complex single and staged procedures. This part of the introduction alerts the medical profession that many of these cases should not be handled by amaterus, as this branch of surgery has grown into a full-fledged specialty of its own. The complex anatomy of this region, the seriousness of the congenital and accidental injuries that can affect this growing anatomy, with all of its organs and vital structures, puts it in a category by itself. This is well illustrated by the drawings of the embryological status relating to the organs and the different regions and systems of the face and neck.

This text has an abundance of photographic documentation of the clinical divisions that are occurring in the epithelial, somatic, and endothelial elements. These illustrations are nicely coordinated with the text and assist in the education and learning process of the great variety of conditions. This is augmented by the identification and use of the modern diagnostic facilities that are necessary to evaluate the extent of certain congenital, neoplastic, and traumatic situations. The text also covers the great variety of treatments, those which have become rather standard, and the newly innovative techniques for this complex group of problems. Many of the chapters have very complete bibliography and reference lists.

The authors make it quite clear that certain abnormalities may require multiple or serial operations, and that they must be carried out by a specialist or certain groups of specialists. Some of the complex clinical conditions cannot be completely or perfectly cured by one or more operations, and the search for second opinions and new ideas is appropriately emphasized.

They very wisely emphasize the necessity of a good rapport with the family group. This essential part of the treatment program is accomplished best by a friendly attitude, adequate communication and explanations, informed consent, and a sense of caring.

The chapter on benign and malignant tumors of the head and neck in children is exceptionally interesting and important. The authors include appropriate classifications, the methodology of diagnosis and treatment, the seriousness of psychological implications, the fact that these are emotionally charged situations for all participants, and that these surgical adventures are associated with life and death, and in some instances, serious deformity in a very young child. The young child who is an innocent victim of trauma in this age of violence and accidents, which may be inadvertent and unexpected, and of minor or horrendous

proportions, is another important chapter. They emphasize general principles and specific surgical techniques in relation to the soft tissues, bones, organs, nervous system about the face and orbit, and, of course, the enveloping skin.

Facial nerve problems have always proven to be especially poignant to the family group. These problems may be congenital or acquired, and may be associated with complete or partial paralysis. These are highly personal situations because of the emotion of human expression. They detail the programs for amelioration and for optimal restitution. It is well recognized, however, that it is not possible to make a paralyzed face normal by any surgical technique. It is also recognized that almost all of these conditions today can be improved considerably.

The chapter on anesthesia identifies this, indeed, as a specialty unto itself. The pediatric anesthesiologist must put the child to sleep, monitor his physiological balance during this interval and awaken him. In doing so, the anesthesiologist must create optimal situations for oxygenation, fluid balance, maintenance of the airway, pulmonary physiology, and an uncomplicated recovery. One of the tricky aspects of anesthesia in children is the miniaturization of the anatomy and the equipment in contrast to the adult. When this is understood, planned for, and carried out on these terms, the patient has the best opportunity for a normal recovery.

One of the most important chapters, on a subject which is often not taught in the classroom, but is self-acquired on the basis of exposure and experience with illness in children, deals with the psychological implications and effects on the child, on the family group, and on the professional attendants. In many complex problems, these effects prove too profound, leaving a permanent mark on the siblings and the family group. Adjustment to these factors and their amelioration requires sensitive and supportive attention on the part of all the professionals involved. A good doctor cannot escape sharing in this stress and this travail.

There is no doubt about the multiple values of this book, both for the specialists who participate in this type of work, and for the children and their families who endure it with hope.

John Conley, MD.

Preface

Pediatrics as a specialty has given way to many subspecialties. General surgeons were the first to specialize in surgical diseases of children and over the past several years there has been increasing recognition of the need for pediatric surgical specialists in the areas of anesthesia, urology, otolaryngology, ophthalmology, neurosurgery, and plastic and thoracic surgery. Facial plastic surgery in children continues to be performed by a wide group of regional specialists. This book brings together the diagnosis and treatment of congenital and acquired surgical conditions of the head and neck as it relates to facial plastic and reconstructive surgery in children.

The majority of facial plastic and reconstructive problems in children arise from congenital disorders. The chapter that begins this book on genetic and craniofacial disorders

will help the reader appreciate some of the new developments in the diagnosis and treatment of genetic syndromes, associations, and sequences. The next several chapters discuss the diagnosis and treatment of congenital abnormalities of the ears, eyes, nose, and neck as well as vascular malformations and hemangiomas of the head and neck. This is followed by a discussion of primary and secondary treatment of children with cleft lip and/or cleft palate. Cleft lip and palate is a major focus of pediatric plastic and reconstructive surgery because of the complex, multiple surgical procedures required to treat these children. Many new treatment developments are emphasized, as well as the importance of the team treatment concept. The next section discusses current treatment of soft tissue and skeletal trauma of the head and neck, as well as wound healing and secondary scar revision. The final chapters cover a variety of related topics including dermatological problems, orthognathic surgery, congenital and acquired facial nerve paralysis, tumors of the head and neck, anesthetic problems with children, and the psychological impact on the child and family coping with a congenital or acquited cosmetic deformity of the head and neck.

The breadth of material in this book will appeal to many physicians and surgeons dealing with pediatric patients who have a congenital or acquired abnormality of the head and neck. This book will be useful to the physician when making a diagnosis and counseling patients and their families, and will be invaluable to the surgical subspecialist treating children with facial plastic and reconstructive problems.

We hope that bring together specialists to discuss the treatment of pediatric facial plastic and reconstructivwe problems will enhance the outcome of those children who have suffered a congenital or acquired deformity.

James D Smith
Robert M Bumsted

Chapter 1: Syndromes and Craniofacial Surgery

Lawrence J Marentette and Robert J Gorlin

The studty of craniofacial anomalies is an ever-expanding field. New syndromes are currently being discovered and new etiologies for established syndromes are being delineated through the field of molecular genetics. The purpose of this chapter is to provide an overview into the different categories of disorders that are responsible for craniofacial anomalies. Additionally, the common intracranial and extracranial approaches in craniofacial surgery will be discussed in order to elucidate the principles of total correction of the cranium and cranial base.

A definition of terms is necessary in order to place in proper perspective the underlying etiology for craniofacial anomalies.

Malformation - an anomaly that is caused by an intrinsic defect in the patient. Examples of this would include the chromosomal abnormalities such as trisomy 21, oculo-auriculo-vertebral spectrum, and cleft lip and palate.

Deformation - an anomaly that is caused by a force extrinsic to the patient. Examples of this would include Pierre Robin sequence and craniosynostosis secondary to fetal head constraint. Congenital deformations of the head and neck are common with most resolving spontaneously in the first few days of postnatal life.

Disruption - can be the most severe form of craniofacial anomaly. Although the fetus has the potential to develop normally, external forces act not only to deform but to destroy the developing tissue. An example of this is amniotic band sequence. In this anomaly, amniotic rupture can lead to constriction of the developing craniofacial skeleton by amniotic bands. The pressure exerted on a developing face is so severe that actual tissue necrosis occurs. This is commonly seen in bizarre, unexplained facial clefting. Other associated anomalies may be present and would include limb reduction defects or amputations, growth restriction, and extrathoracic heart, among others.

Syndrome - characterized by a defined set of physical findings that when found in a prescribed, repeatable association, comprise a very specific clinical entity.

Sequence - a craniofacial anomaly that results from a series of intrauterine events. A classic example of this is the Pierre Robin sequence. In this anomaly, there is extrinsic compression of the mandible resulting in failure of the embryonic tongue to lower. Because the embryonic tongue is remaining between the palatal shelves, they are physically not able to close and therefore, cleft palate results.

Association - not a specific disorder or a syndrome. It is rather a collection of a variety of diagnoses. Its main purpose is to alert clinicians to look for other disorders when a group of these disorders has been identified. The CHARGE association (coloboma, heart defects, atresia, retarded growth, genital hypoplasia, eardrum deformities) is a typical example of this.

Team Evaluation

The craniofacial team evaluation is at the heart of treating patients with these congenital anomalies. Patients are evaluated by all the members of the team in one concentrated area. The team then meets after the examinations have been completed to discuss each member's physical findings and recommendation. Oftentimes two or more specialties are involved in any one particular aspect of the patient's treatment and the timing of their treatment is coordinated through the discussions within the team conference. A typical craniofacial team consists of a craniofacial surgeon, neurosurgeon, otolaryngologist, oral and maxillofacial surgeon, speech pathologist, audiologist, pedodontist, orthodontist, prosthodontist, social worker, geneticist, pediatrician, and ophthalmologist. In some cases, other specialties are needed to participate on an occasional basis in the team evaluation, and their participation is coordinated by the head of the craniofacial team. The patient benefits from having a multidisciplinary evaluation in a single location and the members of the team benefit from the conference discussions by learning from the other members. It is important if at all possible to classify the patient into a category of syndrome, sequence, or other classifications. When this identification is done, the clinicians can then proceed to further evaluate the patient for other defects that are found in association with that diagnosis. It is also important from the standpoint of genetic counseling to let the patient's family know whether this deformity can be manifested in future siblings as well as in the offspring of the patient.

The comprehensive report of the team conference is forwarded to the patient's family as well as to all of the patient's providers. This dissemination of information allows for direct continuity of care between the craniofacial team and their primary providers. Patients are usually recalled on an annual basis to evaluate their progress and results of treatment recommendations and to reassess what other treatments need to be instituted.

Syndromal Craniosynostosis and Craniofacial Synostoses

Crouzon Syndrome

Crouzon syndrome is well known in the literature of craniofacial surgery. It is also referred to as craniofacial dysostosis and craniofacial synostosis. The mode of inheritance has been found to be autosomal dominant. The patients exhibit characteristic facies with maxillary hypoplasia, proptosis, and orbital hypertelorism along with brachycephaly. Craniosynostosis is one of the hallmarks in the syndrome and usually involves the coronal sutures producing brachycephaly. The sagittal suture may be involved instead, resulting inscaphocephaly and the metopic suture may be fused producing trigonocephaly. Occasionally, the cloverleaf skull deformity is seen with pansynostosis.

Examination of the face reveals proptosis of varying degrees secondary to shallow orbits. The maxilla as well as the zygomas are hypoplastic including the root of the nose. The midfacial hypoplasia results in a class III dental and skeletal relationship as well as an anterior dental open and a bilateral maxillary crossbite. Approximately one-half of the patients will have conductive hearing loss. The diagnosis is made on clinical findings and also can be determined from evaluation of the patient's pedigree. It can be distinguished from other common craniosynostosis syndromes by the lack of hand and foot anomalies.

Apert Syndrome

The facial features in Apert syndrome may superficially resemble those of Crouzon, but the finding of syndactyly easily differentiates it from other craniosynostosis syndromes. Apert syndrome occurs mostly sporadically. Proptosis is a hallmark finding in Apert patients as a result of shallow orbits and maxillary hypoplasia. Hypertelorism is a constant finding as well, and the palpebral fissures have a down-slanting appearance. The midface hypoplasia results in a class III dental and skeletal relationship and the maxillary arch findings are more severe than in Crouzon syndrome. The arch is characterized by a very narrow V-shaped appearance with a deep midline furrow and hyperostosis on the alveolus. An anterior dental open bite is seen as well as posterior and anterior maxillary crossbite. The calvarium shows significantly more deformity than that found in Crouzon syndrome. Pansynostosis is much more common and there are marked contour irregularities of the frontal, parietal, and occipital bones. The deformity extends into the base of the skull with marked shortening of the anterior cranial fossa. Asymmetries of the skull base are often seen and associated with corresponding cranial vault asymmetries. The syndactyly involved in Apert syndrome is characterized by fusion of the second, third, and fourth digits of the hands and feet. The diagnosis of Apert syndrome is made on clinical findings and after careful examination can be distinguished from other craniofacial synostosis syndromes.

Nonsyndromal Craniosynostosis

Fusion of one or more calvarial sutures can produce cranial and craniofacial anomalies and yet not be associated with identifiable syndromes. The nonsyndromal group of craniosynostosis involves fusion of the calvarial sutures, and any facial anomaly produced is a result of the cranial and possibly skull base fusion. This is in contrast to syndromal craniosynostosis in which there is primary involvement of the skull base synchondroses as well as the calvarium.

Unilateral Coronal Synostosis

Synostosis of a unilateral coronal suture produces the deformity of plagiocephaly. This is characterized by retrusion of the forehead on the affected side and in more severe cases a teardrop or harlequin orbital deformity of the affected side. Facial asymmetry may occur with the lower facial midlines deviating away from the affected side.

Scaphocephaly

Fusion of the sagittal suture results in the deformity of scaphocephaly. There is an elongated calvarium with a decrease in the bitemporal and biparietal width. This is the most common suture involved in craniosynostosis.

Trigonocephaly

Synostosis of the metopic suture, the midline suture of the frontal bone extending from the anterior fontanelle to the nasofrontal suture, results in the deformity of trigonocephaly, or a triangle-shaped skull. There is a midline forehead vertical ridge at the site of the fused metopic suture. There is also bitemporal narrowing and bilateral recession of the lateral

aspects of the superior orbital rims.

Brachyocephaly

Fusion of the coronal suture bilaterally results in the deformity of brachyocephaly. The forehead is retruded and wide, and there is an increase in the bitemporal and biparietal measurements.

Lambdoid Synostosis

A less common form of craniosynostosis, lambdoid suture involvement may occur unilaterally or bilaterally. If unilateral involvement is found, there is marked flattening of the affected side of the occiput, which may produce ipsilateral prominence of the frontal bone. In the bilateral form, the back of the head is symmetrically flattened and there may be more severe deformity of the frontal and parietal bones. The calvarial deformity is also termed plagiocephaly.

Pansynostosis

Fusion of both coronal and both lambdoidal sutures results in pansynostosis. Oftentimes there is a gapping sagittal suture; however, if this suture also is fused, the anterior fontanelle is exceedingly large and in some cases the brain can protrude through the fontanelle. Because of the marked restriction to brain growth, increased intracranial pressure is an extremely common finding in these patients.

Branchial Arch Anomalies

Oculo-Auriculo-Vertebral Spectrum

This spectrum includes disorders known as hemifacial microsomia, Goldenhar syndrome, Goldenhar-Gorlin syndrome, first and second arch syndrome. The range in severity of the anomalies found clearly demonstrates that this is indeed a spectrum. The hallmark finding is facial asymmetry with varying degrees of facial deficiencies. It is postulated that early vascular disruption of the first branchial arch leads to the unilateral tissue deficiency seen with this spectrum. In the laboratory, Poswillo showed that vascular disruption resulted in the destruction of tissues in the ear and mandibular region of the experimental animal. The degree of tissue destruction seemed to be related to the severity of vascular disruption.

Varying degrees of facial asymmetry are typically seen with this spectrum. There is hypoplasia of the maxilla and zygoma on the affected side as well as of the temporal bone. The mandible is affected as well, ranging in severity from mild hypoplasia to complete aplasia of the ramus and glenoid fossa. The auricle is affected on the involved side with deformities ranging from mild auricular deficiency to auricular dysplasia. The external auditory canal may be stenotic or in more pronounced cases, atretic. Because of the temporal bone and facial deficiencies, the facial nerve may take an aberrant course after it exits the skull base. As a result of the mandibular and maxillary deficiency, the occlusal plane is canted upward on the affected side and cleft lip and palate have been reported in some patients. Skeletal anomalies may also occurr including fusion of cervical vertebrae, platybasia, spina bifida, and scoliosis

among others. Ocular findings include coloboma, epibulbar dermoids, and on rare occasions microphthalmia or anophthalmia.

Chromosomal Syndromes

Trisomy 21 Syndrome (Down Syndrome)

Trisomy 21 is the most well known of all malformation syndromes. Its prevalence is approximately 1:650 live births but may vary from 1:600 to 1:3.000 live births, with nondisjunction the cause of the malformation is roughly 95% of the patients. The risk increases with maternal age. In mothers less than 33 years of age, the prevalence is 0.9 per 1.000, whereas in women greater than 44 years of age, the prevalence is 38 per 1.000. Life expectancy in patients with Down syndrome is approximately 35 years of age. In infancy, causes of death include congenital heart disease, leukemia, and recurrent pneumonias. In the older adult patients, Alzheimer's disease is a common cause and neoplastic diseases are twice as prevalent compared to the general population. They include most commonly leukemia, but also lymphomas and central nervous system tumors are likewise reported.

The craniofacial features of this syndrome make the diagnosis readily apparent. Brachycephaly and flattening of the occiput are common cranial vault findings. Frontal and sphenoid sinuses are absent and maxillary sinuses are hypoplastic in 90% of the patients. Nasomaxillary hypoplasia produces the characteristic midface deficiency, and orbital hypotelorism is a common finding. Because of the nasomaxillary deficiency, relative and/or mandibular excess is an additional finding. Epicanthal folds, up-slanted palpebral fissures, and Brushfield's sports are common ocular findings. The ears tend to be small and the earlobes are small or absent. Craniocervical junction abnormalities are not uncommon and about 10% to 20% of the patients have atlantoaxial instability. The patients exhibit an open-mouth posture with a relative macroglossia and fissured tongue. The lingual papillae are excessively large. Dental eruption is delayed in the deciduous and permanent dentition, and the eruption sequences may be quite bizarre. Malocclusion is extremely common and the findings include a posterior maxillary crossbite, anterior open bite, and anterior crossbite. Associated cardiovascular defects are common and they may include ventricular septal defects, atrial septal defects, and pulmonary stenosis. The diagnosis is readily made by confirming the trisomy chromosome 21 through chromosomal studies.

Fragile X Syndrome

Fragile X syndrome represents 50% of the cases of X-linked mental retardation. It constitutes between 2% and 6% of the population of mentally retarded males. The frequency of the syndrome is 1:1.200 males. Craniofacial features of the syndrome include an increased head circumference with a dolichocephalic-shaped cranium. The forehead is large with prominent supraorbital ridges. There is a broad-based nose and varying degrees of maxillary hypoplasia. The palate is high and arched, and maxillary crossbites and anterior open bites are quite frequently seen. The diagnosis is made by culturing chromosomes in a folic acid-deficient medium for up to 96 hr at elevated pH and supplemented with 5% serum.

Craniotubular Bone Syndromes

Craniometaphyseal Dysplasia

Craniometaphyseal dysplasia is a disorder that involves hyperostosis of the craniofacial skeleton as well as metaphyseal flaring in long bones. Both autosomal dominant and recessive means of transmission have been reported. Hyperostosis develops in the region of the nasal bones early on and progresses to widening of the area of the nasal bones and the nasal process of the maxilla. This leads to one of the diagnostic findings of orbital hyperteloriosm. With time, the increased bone deposition leads to nasal airway occlusion. Hyperostosis is also found in the skull base, which affects the anterior and posterior cranial fossa. In the middle cranial fossa the temporal bone is also affected and patients develop a progressive sensorineural hearing loss. Additionally, 30% of the patients develop facial nerve paralysis. The diagnosis is made through facial examination as well as skull and long bone radiographs. Hyperostosis of the facial bones and skull base is readily apparent radiographically and the long bone survey reveals club-shaped flaring in the metaphyseal area.

Inborn Errors of Metabolism

Hurler Syndrome

Hurler syndrome is one of the more familiar craniofacial syndromes associated with inborn errors of metabolism. The patients classically have growth failure, mental retardation, typical craniofacial features, and skeletal dysplasia. The enzymatic defect in the syndrome is a deficiency of alpha-L-iduronidase activity. The lack of this enzymatic activity prevents intralysosomal breakdown of alpha-L-iduronide containing glucosaminoglycans, dermatan sulfate, and heparan sulfate. The intracellular accumulation of glucosaminoglycans then interferes with the normal function of the affected cells and leads to characteristic clinical signs and symptoms. The frequency of Hurler syndrome is 1:100.000 live births. Craniofacial features include synostosis of the sagittal and metopic sutures leading to scaphocephaly. Overall, there are coarse facial features with hypoplasia of the nasal bones and a broad nasal tip. The cheeks and nasolabial folds are quite prominent and thickened as are the earlobes and the lips. Patients typically manifest an open-mouth posture and nasal airway obstruction is significant. Macroglossia is a constant oral finding and patients exhibit an anterior dental open bite with widely spaced teeth and severe attrition. There is maxillary dental alveolar protrusion and an obtuse gonial angle. Obstructive sleep apnoea is very common due to the macroglossia and narrowing between the face of the sphenoid sinus and the hard palate.

The tongue is quite large and because of the macroglossia the patient assumes a persistent mouth-open posture with protrusion of the tongue. This persistent open posturing leads to development of the anterior dental open bite. The lips are coarse and quite thick and there is increase in the vertical height of the upper lip. The patients very frequently have areas of cystic bone destruction in the mandible usually associated with second primary molars and first and second permanent molars.

The diagnosis can be made early on by evaluating urinary level of glucosaminoglycans. In Hurler syndrome there is more dermatan sulfate found in the urine than heparan sulfate. Cell culture techniques have been useful also in making the diagnosis,

and deficiency of alpha-L-iduronidase can be detected in fibroblast and leukocytes.

Overgrowth Syndromes

Beckwith-Wiedemann Syndrome

Beckwith-Wiedemann syndrome is characterized by macroglossia, omphalocele or umbilical hernia, hypoglycemia, and increased birth weight and length. This syndrome is predominantly sporadic, and chromosomal abnormalities have been discovered in association with Beckwith-Wiedemann syndrome. The growth hormone production is normal in these patients; however, increased levels of somatomedin have been found, which may be the cause of the visceromegaly. Hyperplasia of pancreatic eyelet cells is responsible for the hypoglycemia seen in the syndrome. The most common of the craniofacial features in this syndrome is macroglossia, which may cause obstructive apnea. If surgical tongue reduction is not performed, anterior open bite will develop as a result of the anterior tongue positioning. The open bite may develop, however, in spite of partial glossectomy. Nevus flammeus of the face is also common. Auricular anomalies include linear grooves on the lobule as well as indented areas on the posterior aspect of the auricle. Mild macrocephaly can occur in some cases.

Visceromegaly is very common, with hyperplasia occurring in the pancreas, kidney, and adrenal medulla. There is increased risk of neoplastic development in patients with Beckwith-Wiedemann syndrome. The most common is nephroblastoma, adrenal cortical carcinoma and hepatoblastoma, neuroblastoma, rhabdomyosarcoma, and other visceral tumors have also been reported in association with this syndrome. The diagnosis is made on the clinical findings and evaluation of blood glucose levels to confirm hypoglycemia. Periodic abdominal ultrasounds are likewise indicated for early detection of visceral neoplasms.

Surgical Correction

Surgical correction of patients with craniofacial deformities requires the application of a full range of techniques in facial plastic and reconstructive surgery. Complete coverage of all of these techniques is beyond the scope of this chapter; however, to illustrate correction of upper facial deformities, the frontoorbital advancement and orbital hypertelorism procedures serve to give a broad overview of the type of surgery needed to correct craniofacial anomalies.

Frontoorbital Advancement

This procedure is commonly done in patients with Crouzon or other syndromes in which there is recession of the forehead and the orbits. The ideal age of the patient for this surgery is between 3 and 6 months. When surgery is done at this age, there is no restriction of brain growth and overall results are better than when the surgery is performed at a later time. Also in children under 1 year, the dura has maximal bone regenerative capacities and any gaps left in the cranium are readily filled in with new bone.

The upper facial skeleton is approached through a bicoronal incision that is carried from tragus to tragus. The scalp dissection then proceeds anteriorly in the subgaleal plane

leaving the periosteum attached to the bone. This is helpful in reducing intraoperative blood loss. Dissection is carried down over the supraorbits, preserving the supraorbital nerves, to approximately the level of the lateral canthal tendons. The temporalis muscle is retracted from its fossa to gain access to the greater wing of the sphenoid and the lateral orbital wall. Bifrontal craniotomy is then performed and the bone flap is removed. Next, osteotomies are made in the orbital roof/anterior skull base to allow for remodeling and repositioning of the upper half of the orbits. Beginning medially, osteotomies are made through the nasofrontal suture and then continue lateraly through the floor of the anterior fossa and the roof of the orbits. This extends laterally to connect with the lateral orbital wall. The lateral orbital wall osteotomy is then carried through the midpoint of the lateral orbital rim. Posteriorly the osteotomy is continued back through the sphenoid wing and into the squamous portion of the temporal bone. The osteotomies are performed in a tongue-in-groove fashion so that when this "frontoorbital bar" is advanced, adequate bone contact is maintained for stability in the temporal fossa. Once the osteotomies have been completed, the frontoorbital bar is advanced or may be removed for remodeling if necessary. Fixation is then accomplished in the temporal fossa with either miniplates or wires, depending on the preference of the surgeon. The bifrontal craniotomy bone flap is then replaced and wired or plated to the advanced frontoorbital bar. This results in a gap behind the frontal bone flap that will fill in with new bone if the procedure is performed at a young enough age.

Orbital Hypertelorism Correction

The correction of orbital hypetelorism, like frontal orbital advancement, requires a team approach by craniofacial surgery and neurosurgery. Correction of orbital hypertelorism can usually be done on patients between the ages of 2 and 4 years; however, individual circumstances may dictate that it be done either prior to or after this time frame.

A coronal approach is used to gain access to the upper facial skeleton. This is done using an excision from tragus to tragus followed by a subgaleal dissection to the supraorbital rims. The entirety of the orbits must be exposed; therefore, the dissection also proceeds inferiorly and laterally over the temporalis muscle to expose the zygomas and zygomatic arches. The dissection here, as the arch is approached, is carried through the superficial temporal fat pad and then proceeds on to the superior aspect of the zygomatic arch. The periosteum is incised superiorly and retracted laterally, thus preserving the temporal branch of the facial nerve as it crosses over the zygomatic arch and proceeds up to the frontalis muscle. The lateral dissection is then carried to the infraorbital nerve.

Medially the dissection is carried down over the nose, exposing the medial canthal tendons, which are detached and tagged for reattachment at the end of the procedure. After this, the dissection carries medially along the medial and infraorbital rim to completely connect the lateral dissection with the medial dissection such that the orbits are skeletonized in all four quadrants. It may be necessary to make additional incisions to access the floor of the orbit and these may be made through a subciliary, infraorbital, or transconjunctival-lateral canthotomy approach. Once the orbits are skeletonized, osteotomies are made through the medial, lateral, and inferior orbital rims as well as through orbital walls.

As is typical with hypertelorism seen in, for example, frontal nasal malformation, there is excessive bone in the nasoethmoid region. This bone is excised in paramedian strips,

preserving the midline sutures of the nasal septum and the olfactory apparatus. Bifrontal craniotomy is next performed, and the brain is retraced posteriorly followed by osteotomies through the superior orbital rims and through the roof of the orbits from the intracranial side. Once the osteotomies are completed, the paramedian nasoethmoid ostectomies are performed, excising excessive bone and ethmoid sutures. Following this, the orbits are mobilized and medially translocated.

Bone grafts are used to stabilize the orbits in their medial position and are placed in the gaps left at the lateral orbital rim. Medial canthal ligaments that have been previously identified are then repositioned with a transcanthal/transnasal wiring technique. After securing the ligaments, deficiency in the nasal area is treated with a bone graft to improve prominence of the nasal dorsum. Any other gaps in osteotomy lines may also be bone grafted to further stabilize the medially translocated orbits.

Summary

The field of craniofacial surgery is based on a thorough understanding of various syndromes of the head and neck as well as a knowledge of a normal postnatal facial growth and development. Surgical techniques are constantly improving, resulting in improved outcomes. When the team approach is used, the patient is offered the best possible treatment plan because of the input of the many specialties involved in the evaluation.