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 amateurs, 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 acquired 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 bringing together specialists to discuss the treatment of pediatric facial plastic and reconstructive problems will enhance the outcome of those children who have suffered a congenital or acquired deformity.

James D Smith
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Chapter 1: Syndromes and Craniofacial Surgery

Lawrence J Marentette and Robert J Gorlin

The study 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, ear 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 retraction 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.

**Brachycephaly**

Fusion of the coronal suture bilaterally results in the deformity of brachycephaly. 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 occur 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 spots 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 hypertelorism. 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 laterally 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 hypertelorism, 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 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.
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Chapter 2: Congenital Anomalies of the Nose and Nasopharynx

James D Smith

Congenital anomalies of the nose are related to the embryological development of the face during the first 12 weeks of fetal development. To better understand the clinical findings, pathology, and differential diagnosis, and to make a plan for the evaluation and treatment of these anomalies, we must review the embryological development of the nose and face during those critical first 12 weeks of development.

Embryology

The development of the face begins at about 3 to 4 weeks of age. At this stage the embryo is composed of a brain covered with a membrane; the anterior neuropore is still present and the optic vesicles are placed on the lateral aspect. At this point the neural fold develops as a longitudinal infolding along the dorsal aspect of the embryo. The margins of this infolding form the neural crest, which continues to expand into the neural groove and undergoes closure, creating the neural tube. During the same time the neural-crest cells, which are "pluripotential cells," migrate laterally and are important in the development of the face as they are the tissue of origin of the various germ layers that create the facial structures. As these cells migrate laterally they pass posteriorly to the developing eye and anteriorly over the frontonasal process. At the same time, the fetal head is flexing as the neural-crest cells reach their predetermined location to create the first and second branchial arch mesenchyme. Now the development of the matrix of neural tubes and cellular differentiation occurs, which leads to facial process development by the end of the 4th week.

As this step is completed, the head consists of a large bulging mass at the cephalic end, which contains the forebrain. Two small thickenings just above the stomadeum depression in the epithelium form the nasal placodes. The stomadeum is created by the separation of the first branchial arch into the maxillary and mandibular portions. In a short period of time this depression connects with the primitive foregut, forming the oral plate, which ruptures to form the oral cavity. At the same time the nasal placodes are developing as horseshoe-shaped bulges, forming the medial and lateral nasal processes. As the medial nasal processes approach each other in the midline, the embryo's face begins to have recognizable human features. During this time the changes that occur with amazing rapidity and precision are critical in the formation of many of the congenital anomalies of the head and neck.

The processes that are developing are really ridges of ectoderm overlying proliferating, differentiating, and migratory neural-crest cells. Once the overlying facial features are formed, there is not a dramatic change in appearance, but the smoothing out of the tissues by the tremendous differential growth of the mesenchymal tissue results in enlargement of the fetus, as well as in changes in facial configuration.
During the 6th week there is rapid growth of the medial nasal processes and the maxillary processes toward each other, with fusion to form the upper lips, philtrum, philtral ridges, and columella. The lateral nasal processes become less prominent, but a deep groove is formed between them and the maxillary process that extends from the stomadeum to the medial canthus of the eye. This depression, the nasal maxillary groove, will later be closed by overlying epithelium and will form the nasolacrimal duct.

During the next 2 weeks, the pits of the nasal process burrow deeper and above the stomadeum to form a thin sheet called the nasobuccal membrane, which will rupture to create a nasal cavity with a primitive choana. During this time, the two sides are separated by the forming nasal septum with its anterior septal cartilage and the columella, as a result of the mesodermal flow from neural crest cells into the frontonasal process. Other neural crest cells enter the craniobasal region to form a cartilage model that will ossify and become the posterior septum, ethmoid complex, cribriform plate, and sphenoid bone. The ethmoid complex is derived from lateral and medial mesenchymal masses that surround the olfactory sacs. The olfactory nerves penetrate the area between the lateral and medial masses, and later are surrounded by cartilage that will ossify to become the cribriform plate. The remainder of the posterior septum comes from mesenchymal tissue forming the vomer. The septum is then fused in the midline with the palatal processes starting anteriorly and continuing posteriorly to form the hard palate and separating the oral and nasal cavity. The nasobuccal membrane will have ruptured, allowing communication with the oropharynx via the permanent posterior choana.

If during this 12-week, rapidly moving, complex event there are genetic or external influences that effect the normal movement of cells and cell masses, there will be a permanent effect on the fetus. Later steps in the developmental sequence will be further altered resulting in the congenital anomalies we will be discussing.

**Nasal Dermoid (Nasal Dermoid Sinus Cyst)**

The appearance of a subcutaneous epithelial cyst in the area of the nose may represent a simple epidermal inclusion cyst or a nasal dermoid sinus cyst (NDSC). The former is simply a cyst with a fibrous wall, lined with stratified squamous epithelium, and confined to the subdermal or subcutaneous layers of the skin. An NDSC may have a similar appearance, but it may or may not have an opening to the skin or have an intracranial connection to the dura. On microscopic exam it will have a fibrous cyst wall lined with squamous cell epithelium as well as other skin appendages including sebaceous glands, hair follicles, and sweat glands.

**Embryology**

Nasal dermoid sinus cysts represent an embryological defect similar to the defect that forms a nasal glioma or anterior encephalocele. The external nose forms initially as a cartilaginous foundation that is continuous posteriorly with the ethmoid labyrinth. External to this cartilaginous foundation the nasal and frontal bones form via intramembranous ossification during the 2nd to 3rd month of life. Between the frontal bones and nasal bones there is a space filled with a firm membrane called the fonticulus frontalis. Between the nasal bones and the more posterior cartilage is the prenasal space. At the cranial end of the prenasal space there is a herniation of dura through the foramen caecum, which passes through the
prenasal space and is continuous with the periosteal lining of the nasal bones. As development proceeds, the foramen caecum and fonticulus frontalis are obliterated by bony growth.

Various theories concerning the embryopathogenesis of NDSCs have been proposed. Cysts basically fall into two categories: those that result from a cranial developmental defect and those that result from a cutaneous developmental defect. Pratt feels that there is failure of the skin to separate from the dura mater, which results in ectoderm being carried into the prenasal space for varying distances, including through the foramen caecum and intracranially to form a dermoid cyst or a persistent nasal dermal sinus. A tract may have cystic dilatation anywhere along its course or may undergo fibrous obliteration at any point along the tract. If there is failure of or incomplete retraction and obliteration of the neuroectodermal elements and the foramen caecum does not close, glial tissue may extend along the pathway, producing a glioma or encephalocele.

Diagnosis

Nasal dermoid sinus cysts are usually noted at birth or shortly after. They may range from a small pit in the skin with a few hair extruding somewhere between the glabella and the base of the columella, to midline cysts with or without a skin connection. If there is a skin pit it will intermittently drain small amounts of cheesy debris or cloudy exudate. Occasionally they do not drain and the diagnosis is not made until adulthood. If there is a cyst without a connection to the skin allowing drainage, it will gradually increase in size, remaining unattached to the skin, but thinning the skin and making it difficult to dissect a plane between the two. If there is a sinus tract, it will usually extend cephalic to the skin pit in a subcutaneous plane to the level of the nasal bones. At this point the sinus tract or cyst will create a depression splaying out the upper lateral cartilages and quadrilateral cartilage of the septum, causing the nose to be widened. The tract will usually pass beneath the nasal bones and into the prenasal space, although rarely the tract may stay superficial to the nasal bones and go between the nasal bones and frontal bone to enter the fonticulus nasofrontalis and extend through the foramen caecum. In either path the sinus tract may widen to form another cyst under the nasal bones and then extend superiorly into the anterior cranial fossa via the foramen caecum, the crista galli, or the cribriform plate, and sometimes as far posterior as the basisphenoid.

Intracranial extension is variable and in various reports has ranged from 0% to 85%. Intracranial extension is more frequent if a skin fistula tract is present, but may also be present with cysts and no skin pit.

Untreated NDSCs may become infected and require antibiotic therapy. Sometimes they may be mistaken for an infected epidermal inclusion cyst and be incised and drained. If there is intracranial extension, Paller et al reported a 22% incidence of infection, which included superficial infections, recurrent meningitis, osteomyelitis of the frontal and nasal bones, and frontal lobe abscesses. These complications may press for early surgical therapy.

The majority of NDSCs occur spontaneously, although there have been rare reports of a familial incidence. The reported incidence for midline nasal masses is 1/20,000 to 1/40,000 live births, which include NDSCs, nasal gliomas, and encephaloceles. There is a slight male preponderance in most series.
The differential diagnosis includes epidermal inclusion cysts, which are rare before puberty and are usually attached to the skin. Hemangiomas have a bluish color, are compressible, and are not attached to the skin or underlying tissues. The most critical lesions to differentiate are tumors of neurogenic origin, which will be nasal gliomas or encephaloceles.

Preoperative radiological evaluation is for the purpose of distinguishing between NDSCs, nasal gliomas, and encephaloceles. Today both magnetic resonance imaging (MRI) and computed tomography (CT) are used for this purpose. MRI is the most useful for encephaloceles, identifying brain tissue extending through defects in the floor of the anterior cranial fossa. Using T1- and T-2 weighted images cerebrospinal fluid (CSF) may be distinguished from fat. Barkovich et al found that both the CT and MRI were useful in diagnosing intracranial extension of a nasal dermoid sinus tract, but both also had certain pitfalls. One pitfall was normal widening of the septal cartilage superiorly being interpreted as expansion from a sinus tract in the septum. Fatty change in the frontal and nasal bones may be mistaken for fatty tumors such as the intracranial extension of a nasal dermoid. Finally, the fatty change in the crista galli or perpendicular plate of the ethmoid may be mistaken for intracranial extension. By looking at both sagittal and coronal views one can distinguish marrow from intracranial extension. Both CT and MRI may mislead the surgeon, and this is the reason we prefer to approach an NDSC from the nasal side first to prevent an unnecessary craniotomy.

**Treatment**

The successful treatment of NDSCs is complete surgical excision of all epidermal elements. Since most lesions are diagnosed before 1 year of age, indication for and timing of surgery becomes important. Treatment is indicated for all lesions except for the rare circumstance where the patient has an asymptomatic nasal pit that does not drain, there is no evidence of intraseptal or intracranial cyst, and the patient does not want surgery. Some authors have advocated waiting until the child is 2 to 5 years of age. However, there is the risk of repeated infections of the cyst, which will make dissection more difficult, or life-threatening meningitis if there is an intracranial extension.

My approach has been to evaluate the patient with a CT scan when the diagnosis is first made. If the CT shows a defect in the crista galli or anterior cranial fossa, and MRI is obtained looking for evidence of an intracranial extension. Neurosurgical consultation is also obtained on all patients, in case at the time of surgery there is an unsuspected intracranial extension. If there is no history of infection and the child is less than 1 year of age, I will follow them with plans to excise the lesion at about 1 year of age. If there is a cyst present on the nasal dorsum or in the columella, I watch it for enlargement. If it starts to enlarge, thinning the skin over it, or there is an infection, I will go ahead with surgery at any age.

Our approach is to start by excising the extracranial portion of the NDSC first. If the lesion can be completely excised from this approach, no further surgery is necessary. If the tract extends intracranially, the proximal end is tagged with a hemoclip and the neurosurgeons will remove the intracranial portion via a small bifrontal craniotomy. This may be done at the same sitting, or if the initial procedure has been prolonged it may be delayed for 1 to 4 weeks in very young children.
The key to successful removal is adequate exposure of the nasal dorsum and the ability to continue deep to the nasal bones. Many authors advocate a midline dorsal incision from the glabella to the inferior portion of the cyst or inferior enough to include an elliptical excision of the skin pit, if present. Although this approach gives excellent exposure, it leaves a visible scar directly on the nasal dorsum. I have preferred to use a horizontal incision at the glabella and then to extend lateral limbs inferiorly on each side of the nasal dorsum like the lower limb of an external ethmoidectomy incision. This makes an inverted-U flap based inferiorly, with each limb being in a relaxed skin tension line. The skin pit is excised with a small elliptical excision oriented either vertically or horizontally. This incision gives excellent exposure with an excellent cosmetic scar. If there is a cyst in the tip of the nose or columella, I use a standard external rhinoplasty incision across the midcolumella with lateral incisions along the caudal margin of the lower lateral cartilage. This gives excellent exposure with minimal external scars, but if a sinus tract extends deep and posterior to the nasal bones, dissection may be difficult at the distal extent of the tract. Some authors advocate this approach in all cases because of the excellent cosmesis. Again, a separate incision must be made if there is a skin pit.

The skin is injected with 1% Xylocaine and 1:100,000 epinephrine for hemostasis. First, the elliptical excision is made around the skin pit through the dermis, taking care not to cut into the sinus tract. The skin incision is then made, freeing the nasal skin from the sinus tract and/or cyst to its most caudal extent on the nose. Many times the dermis has been thinned by pressure from the cyst and extreme care must be taken to not "button-hole" the skin. I have found the microscope to be an invaluable aid in this delicate dissection and then following the tract deep into the nose. Once the skin is elevated, the sinus tract may be dissected from its bed on the nasal dorsum. Middle ear elevators like a Rosen or McCabe dissector are especially useful to free the tract. The upper lateral cartilage and septal cartilage will usually be flattened and concave from the pressure of the sinus tract. As one continues cephalically, the tract will usually go between the nasal bones and the cartilage, although occasionally it will extend superiorly over the nasal bones and go between the nasal bones and the frontal bone. With magnification it is easy to follow and distinguish a sinus tract from other tissue. It may be necessary to remove a small amount of the caudal end of the nasal bones or do a medial osteotomy for exposure as the tract extends posteriorly toward the foramen caecum or the crista galli. At this point the tract may end in a blind pouch with just a fibrous tract extending superiorly toward the anterior cranial fossa. With the middle ear elevators, I remove as much of this fibrous tract as possible and then carefully examine the specimen to make sure I can see the blind end of the sinus tract. If there is any question, a frozen section of the fibrous tract is done to make sure there are no epithelial elements left in the most distal portion of the tract. Sometimes this tract may end as a dilated cyst in the septum. Care must be taken to remove all of the sinus tract lining.

If the sinus tract extends superiorly through the floor of the anterior cranial fossa, the dissection is carried to the bony margin. The distal end of the sinus tract is closed with one or two hemoclips, and Gelfoam or Gelfilm is used to fill the defect for an inferior marker when the intracranial portion is resected. Consultation is then made with the neurosurgical team and a decision is made whether to proceed with the intracranial portion at the same setting or delay for 1 to 4 weeks. In our hands, either way has been equally successful and the decision is usually made on the basis of the length of the anesthesia and the patient's age.
The incisions are closed in two layers using either 6-0 nylon or 6-0 fast absorbing catgut in the skin. Normally a drain is not used, but a light tape and pressure dressing is used for 2 to 3 days over the skin, which has been elevated.

If a neurosurgical approach is necessary, a bifrontal craniotomy is used. An extradural dissection is used down to the defect, which is usually in the area of the crista galli. As the dissection is carried out and the bony defect found, the sinus tract may be identified and lifted out, finding the hemoclips that will identify the inferior margin. The sinus tract may end at the dura where it may be peeled off, but more frequently it will have a cyst extending into the falx and a small portion of dura must be excised for complete removal. The dura can usually be closed primarily.

We use perioperative antibiotics unless there is an intracranial extension with infection present. In that case antibiotics would be used for 7 to 10 days.

Postoperative complications are usually limited, but postoperative seromas or infection may occur. With an intracranial resection we have had one temporary CSF leak, which closed spontaneously after 2 weeks. There is concern over the cosmetic appearance of the nose, but so far the postoperative results have been satisfactory.

**Nasal Gliomas and Encephaloceles**

Gliomas and encephaloceles are rare lesions that are made up of glial tissue and arise from similar embryological defects. The incidence is difficult to determine from the literature since they are so uncommon and obviously all cases are not reported. It has been reported that encephaloceles occur in the USA and Europe in 1 in 35,000 live births, but are more common in southern Asia with an incidence of 1 in 6,000 live births. My personal observation is that nasal gliomas and encephaloceles are about equal in occurrence, but both are much less common than NDSCs. Our definition of encephaloceles for the purpose of this chapter includes all types of intracranial herniations that are connected to the CSF system.

**Embryology**

These lesions are thought to arise from an embryological defect: faulty closure of the foramen caecum during the 3rd and 4th week of fetal life as the anterior neuropore is closing. It is thought that there is a failure of the appropriate migration of the neural crest cells, which allows a bony defect to develop along the floor of the anterior cranial fossa.

One theory is similar to the theory for the development of NDSCs. The dura that is in contact with the epithelium fails to retract through the foramen caecum and so instead of having an epithelial tract, one no whas a dural tract with or without actual brain tissue present. In the case of the nasal glioma, there is retardation of the neural crest cell migration, which allows brain tissue to be trapped extracranially, and located intranasally (30%), extranasally (60%), or a combination of the two (10%). About 15% maintain a fibrous connection to the dura, which normally passes through the cribriform plate area.

Encephaloceles are divided into two types, sincipital and basal. In the sincipital encephaloceles there is a failure of the foramen caecum to close just anterior to the crista
galli, allowing the encephalocele to pass between the ethmoid and frontal bone. It will then present externally in one of three locations. The nasofrontal type will present between the frontal and nasal bone in the area of the glabella. As it projects forward, it will cause a downward displacement of the nasal bones and a lateral displacement of the orbital wall. In the nasoethmoidal type the encephalocele will present between the nasal bones and cartilages of the nose. This displaces the nasal bones and frontal process of the maxilla superiorly. In the naso-orbital type, the defect is in the medial orbital wall, between the lacrimal bone and the frontal process of the maxilla. In this type the frontal bone, nasal bone, and nasal cartilages are normal and the mass presents between the lower eyelid and nose.

Basal encephaloceles form a defect in the anterior cranial fossa from the anterior border of the cribriform plate to the supraorbital tissue or posterior clinoid fissure. They will herniate into the nose, nasopharynx, or orbit. They are divided into four types, depending on the location of the defect. Transethmoidal lesions herniate through the cribriform plate and the encephalocele will present intranasally. Sphenoethmoidal lesions have a bony defect in the posterior cribriform plate and will present intranasally or in the posterior ethmoid cells. Transsphenoidal defects are usually large and extend from the cribriform plate to the posterior clinoid with the mass presenting in the nasopharynx. Sphenoorbital encephaloceles extend through the superior orbital fissure and present in the posterior portion of the orbit.

**Diagnosis**

Nasal gliomas are really areas of heterotopic brain tissue without a fluid-filled space that connects with the ventricles or subarachnoid spaces of the brain, but may have a fibrous or glial stalk attachment to the dura. Microscopically, the tissue contains foci of glial cells and fibrous tissue. They are surrounded by a pseudocapsule of fibrous tissue. These lesions are firm, rubbery, and noncompressible, and rarely have mitosis in the cells. As a result they are not neoplastic lesions and expand slowly in proportion with the growth of the child. Most present in or around the glabella of the nose, but only rarely in the midline. The skin overlying the lesion may be erythematous, telangiectatic, or violaceous, but will be mobile over the underlying mass. Those that present intranasally may cause nasal obstruction, septal deviation, or deformity of the surrounding bony structures, or may be large enough to prolapse from the nose. The masses do not enlarge with crying or jugular vein compression (Furstenberg’s sign).

There are rare reports of other head and neck locations for heterotopic brain tissue separate from the subarachnoid space, presenting in a separate compartment of the brain, the nasopharynx, soft palate, pterygomaxillary space, and/or maxillary sinus. These lesions are identical microscopically to nasal gliomas and may even include ependymal cells and immature choroid plexus. Since they lack a connection to the dura, one would assume that they result from a defect during the closure of the anterior neuropore, leaving neural tissue separated from the brain, or possibly from misdirection of the migrating pluripotential neuroectodermal cells. As with nasal gliomas, these lesions grow in proportion to the child.

Encephaloceles do not seem to have a genetic predilection, but are often associated with other congenital abnormalities (30% to 40%). Sincipital encephaloceles most often present in the midline at the glabella or to one side of the nose. The size is variable from small elevations to masses as large as the child's head. The overlying skin may be
hyperpigmented, bluish, thin, smooth, wrinkled, or, in rare cases, with exposed brain and CSF leak. The masses are firm, somewhat compressible, rarely have transmitted pulsations, and may transluminate.

Basal encephaloceles are less common than sincipital encephaloceles and usually present in the nose or nasopharynx. The sphenoorbital type will present with proptosis. The other three types, transethmoidal, sphenoethmoidal, and transphenoidal, will present with unilateral or bilateral nasal obstruction, feeding problems, and airway obstruction, which may be mistaken for a choanal atresia. On examination, there is a polypoid mucosal covered mass pedicled high in the nasal vault or nasopharynx that may pulsate or expand with venous compression (positive Furstenberg's sign). There may be unilateral rhinorrhea. Many smaller encephaloceles may go undetected and the patient will present with recurrent meningitis or spontaneous CSF rhinorrhea.

The diagnosis is best confirmed and the defect outlined with CT and MRI scans. Usually both will be used as the CT scan best identifies the area and extent of bony defects. The MRI best demonstrates the soft tissue components of these lesions. Herniated brain tissue is well identified adn with T1 and T2 weighting, CSF can be identified. With these two modalities a differential diagnosis of NDSCs, nasal gliomas, heterotropic brain tissue, and encephaloceles may usually be made without the use of angiograms.

Treatment

As with NDSCs, the treatment of nasal gliomas and encephaloceles is surgical. The surgical approach for nasal gliomas is similar to that of an NDSC. If after evaluation with MRI and/or CT scan the lesion appears to have no intracranial extension, the lesion is approached by the appropriate external incision for adequate exposure. If the glioma is intranasal, a lateral rhinotomy approach is usually used. The use of the microscope and microscopic instruments is similar to the procedure described for the nasal dermoid sinus cyst. It is important that all of the gliomatous tissue be removed or it will continue to grow with the child. If obvious or occult CSF fluid is encountered, one must assume that the diagnosis was wrong and that one is dealing with an encephalocele. In this case an intracranial approach will be necessary unless a local septal, mucosal, or turbinate flap can be created to seal the leak if it is recognized during the surgical procedure.

Hengerer and Oas report that five nasal gliomas had intracranial procedures as their primary approach, with four being normal, and in one patient a cribriform plate and dural defect was found. They recommended the following indications for intracranial exploration as a primary procedure: (a) a history of meningitis, (b) a compressible intranasal mass, (c) a positive Furstenberg sign, (d) radiographic evidence of an osseous defect or a soft tissue mass in the cribiform plate, and (e) CSF rhinorrhea.

Encephaloceles will tend to enlarge with time, which will increase the cosmetic deformity and potentially the amount of brain tissue and vital structures that may be involved. There is also the risk of meningitis when an encephalocele is untreated. For these reasons, early surgical intervention should be carried out as soon as the patient's condition permits it. If hydrocephalus is present, it should be first corrected with a shunt procedure. Nasofrontal encephaloceles have short, small necks, and may be amenable to an extracranial approach.
The nasoethmoidal and nasoorbital types should be approached intracranially, and the encephalocele separated from the brain with a watertight dural closure. The bony defect may be repaired with a bone graft or alloplastic material like tantalum mesh. The external portion may be removed at a later date or at the same time. If there are skeletal abnormalities in children under 3 years, much of the deformity will correct itself with normal growth. Excessive skin should be resected and the incision closed in a cosmetic fashion. Older children may need either primary or secondary correction of the associated hypertelorism. Sargent et al have described a one-stage reconstruction for these patients.

Intranasal basal encephaloceles should not be approached transnasally as a primary procedure. There will be a known CSF leak and there is the risk of meningitis working through a contaminated field. It is also difficult to obtain a tight dural closure. The primary approach should be intracranial with division of the encephalocele and repair of the dural defect. Osseous repair is similar to the repair for sincipital defects. If there is a residual encephalocele in the nose it can be removed later via a lateral rhinotomy. In transsphenoidal encephaloceles, there may be vital structures such as the pituitary hypothalamus or third ventricle prolapsed into the defect. Exposure is difficult and one must try to reduce the encephalocele rather than transect the stalk. There is a high risk to the operative procedure, and surgical intervention must be carefully weighed in view of the risk-benefit ratio.

Other Anomalies of the Nose

With all of the embryological fusion planes present, it is surprising that there are not more congenital anomalies of the nose. The most common defect associated with defects in the fusion planes of the face are the cleft lip and palate described extensively elsewhere in this book.

Congenital anomalies of the nose may include complete agenesis of the nose and midline clefting of the nose to various degrees. Lateral nasofacial clefts result from inappropriate fusion of the nasal maxillary groove in their severest form. Less severe forms may occur as small defects in the alar margin to actual deficiency or absence of the ala.

Other conditions that have been reported include proboscis lateralis and bilateral proboscis. These are reduplication defects with either an accessory portion of the nose laterally or some degree of reduplication of part or all of the nose. These may be associated with other cranial anomalies and hypertelorism.
The topic of nasal surgery in children has been debated for decades. As in all other aspects of pediatrics, it is imperative that we first do no harm while attempting to correct debilitating conditions. The controversy revolves around whether or not surgical manipulation of the nose will influence it and surrounding structures in a manner that would be aesthetically or functionally beneficial. It must be emphasized that surgery should only be performed after intensive medical therapy has failed to relieve severe obstruction, persistent rhinorrhea, recurrent sinusitis, or upper respiratory illness. Obviously, immediate procedures need to be performed for septal hematoma or abscess or traumatic or congenital deformity causing airway compromise.

Surgeons interested in performing nasal surgery for acquired or congenital disease need to understand the development of the nose and the many surgical options available. Today, innovative instrumentation and techniques allow rapid, accurate diagnosis and give surgeons the opportunity to correct debilitating problems without unreasonable disrupting future aesthetic or function.

Development

Development of the nose begins at approximately 4 weeks of fetal life. At this time the neural tube is closing and neural crest cells begin their migration. In the head and neck neural crest cells form the connective tissues of the face. They are induced by closely associated tissues to differentiate into various types of connective tissue.

Collections of these neural crest cells undergo proliferation, forming the nasal placodes. Other collections proliferate in areas that become the medial and lateral nasal swellings and the maxillary swelling. The medial and lateral swelling soon grow together, forming the two nasal pits, the floor of which is the oronasal membrane. This soon ruptures and then is closed by fusion of the medial portions of the maxillary swellings, thus forming the hard palate. The nasal pits, meanwhile, continue to burrow deep in the mesenchyme until only the nasobuccal membrane remains. This soon ruptures and by week 10, the nose communicates with the choana.

While this is occurring, the cartilaginous framework of the nose is developing. Three paired condensations of mesenchyme in the medial and lateral nasal swellings initially are formed: the trabecular cranii, in the primitive nasalseptum; the tectal condensations, located dorsally around the primitive nares; and the paranasal condensation, located further posteriorly in the lateral nasal swellings. This cartilage model will ossify over the next 4 weeks, becoming the posterior septum, ethmoid complex, and nasal bone.
Postnatal growth and development is predictable and sporadic. The infantile nose is proportionally smaller and more broad with visible nares. Growth is very rapid initially and then slows in early childhood. Another burst occurs during the pubertal growth spurt, after which activity declines. Many factors influence postnatal development of the nose such as occlusion of the teeth, tongue placement, facial musculature, and development of the sinuses. Whether or not the nasal septum is the main determinant of postnatal development is debatable.

Effects of Surgical and Nonsurgical Management on Subsequent Development

The goal of nasal surgery in the pediatric patient is to restore function without deleteriously altering development. It seems logical then to avoid areas of the septum that are "growth centers" and to avoid resection of any tissue if possible. Vetter and his group helped to delineate the metabolically active area of the septum by analyzing small cartilaginous strips obtained at septoplasty from children and adults. They incubated these strips in $^{35}$S-labeled NaSO$_4$, and measured the uptake of this material and showed age-related differences in metabolic activity. In prepuberty the suprapremaxillary area and anterior septum showed the greatest activity, whereas in pubertal children and adults only the anterior end of the septum showed high activity. The conclusion from this study was that these areas appear important metabolically and should be avoided during surgery.

Verwoerd and Verwoerd-Verhoef used a rabbit model to investigate the effects of future development after surgery on various parts of the septum. They found that elevation of the mucoperichondrium unilaterally or bilaterally had no effect on future development and that a small (1 mm) submucous resection resulted in no foreshortening of the nose or maxilla. These rabbits did develop a septal deviation at the point of resection. However, removing a basal strip of the septum resulted in foreshortening of the nose and midface.

Verwoerd and Verwoerd-Verhoef have reviewed several other studies using models that have shown other areas of the septum, along with the premaxilla and the anterior septum, to be crucial in development, including (a) the upper lateral cartilages, whose integrity ensures straight outgrowth of the nose; and (b) the integrity of the T-bar configuration of the septal and upper lateral cartilages. Trauma or disruption of this unit affects the total development of the dorsoseptal cartilage, which in turn causes maldevelopment of the nasal bones and vomer.

The potential effects of large resections as discussed above are discouraging. However, expectant management of a pediatric patient with nasal obstruction may be debilitating also. The dental literature is replete with reports that describe facial maldevelopment attributed to "mouth breathing". Bresolin et al examined 30 allergic children who appeared to breathe through the mouth and compared them to 15 nonallergic children who appeared to breathe predominantly through the nose. All were subject to intraoral and cephalometric analysis. They found that the allergic children had longer faces with narrower maxillae and retruded jaws. However, they readily admit that they had not objectively determined that their patients indeed were breathing through the suspected route. A Canadian group performed a careful study and showed that there was no difference in nasal airflow between "mouth breathers" and normal children. This obscurity then shifts the debate to a question of whether the abnormal anatomy causes the appearance of mouth breathing or is caused by mouth breathing. This
discussion is presented because there are a large number of professionals who believe nasal obstruction and mouth breathing adversely affect development.

A question then arises, do deformities that cause obstruction, specifically, septal deformities, correct themselves over time? Gray examined the septa of 145 babies at birth and at 8 years. He found that all untreated deviations were present or had increased in severity during subsequent development. He correlated this with a high proportion of otitis media. Only surgically corrected septa were straight at follow-up.

This evidence supports surgical manipulation when clinically indicated. The question then becomes, Can surgery be performed with acceptable aesthetic and functional results? Ortiz-Monasterio and Olmedo presented a series of 44 rhinoplasties, which included septoplasties, all done on patients between 8 and 14 years of age. Follow-up consisted of all patients being seen at least 5 years later with results comparable to adult patients. Pirsig presents his more than 15 years of conservative rhinoplasty experience showing poor to excellent results in children with traumatic nasal injuries. Verwoerd et al, despite showing deleterious results of resections of specific portions of the septum, also mention that their findings do not contraindicate a conservative procedure that avoids critical areas of the septum. Healy presents ten children, all with severe obstructing deviations, who had their obstructions relieved via a sublabial route with no disturbance in facial development 10 to 60 months later. Jugo's total septal reconstructive technique also provides good evidence that septal surgery is safe in the younger patient.

**Optimal Timing of Nasal Surgery in Children**

Wide experience, as demonstrated above, has not shown hard evidence to absolutely contraindicate nasal surgery in the younger patient if performed in a conservative manner for appropriate indications. Obviously, in a child with septal hematoma or septal abscess, immediate drainage needs to be performed. A newborn with a dramatically deviated nose may need immediate correction; however, most correct spontaneously. A young child with symptomatic obstruction should have surgery within the guidelines listed below. It may be prudent to perform surgery to relieve obstructing lesions before the pubertal growth spurt, if indeed nasal obstruction causes midface maldevelopment. If a nonobstructing lesion is problematic, surgery should probably be delayed until after the nose is mature. Buck and Brown, in a longitudinal study of nasal growth of children age 6 to 18, showed that 87% of nasal growth is obtained by age 12 in girls and age 15 in boys. Thus, if surgery can be delayed until after the midteens, it should. However, Ortiz-Monasterio and Olmedo reported several children of pubertal age who had rhinoplasty without deleterious results.

**Psychological Aspects**

Surgery in the pediatric population is universally stressful. Parents do not wish to see their children in discomfort and are often distraught themselves when dealing with an operative situation. Children often have great anxiety about parental separation, general anesthesia, postoperative pain, and other aspects of surgery. But at the same time, children, particularly adolescents, may suffer psychological stress over the unacceptable appearance of the nose. The positive benefits and negative factors must be considered on an individual basis. Many reluctant children in whom corrective surgery would be clearly beneficial will often, over a
period of time, accept surgery with proper explanation and guidance on the part of the physician, parents, and others involved. The procedure should be performed for precise clinical indications with special emphasis on restoring function and reasonably relieving aesthetic deformities that can be emotionally devastating. Proper preoperative preparation of the parents as well as the child is crucial. Every effort should be made to allow the child as much control as possible without being detrimental to his/her well-being. A perioperative team trained to work with children in circumstances that all find comfortable is most beneficial. When the above considerations are given proper attention, the surgery will often be a surprisingly smooth experience for the child. If there is any question on the part of the surgeon or any member of the team, including the parents, professional psychological evaluation and treatment, if necessary, should precede surgery.

**Technical Challenges**

The technical challenges are (a) the small operative area, (b) the delicacy of the tissue and the need to always be as conservative as possible, (c) avoiding "growth centers," (d) gaining adequate exposure, (e) aesthetic acceptability, and (f) the underlying pathology causing the deformity. Jugo and Healy use open procedures to perform septoplasty and have obtained excellent results. The new pediatric, rigid nasal endoscopes may dramatically affect surgery by making diagnosis more precise and will allow superior visualization intranasally. More experience with the endoscopic techniques may make open procedures less desirable.

**Congenital Disorders**

The nose is deformed in a multitude of genetic disorders, many of which involve the orbit, midface, palate, or cranial vault. There are also tumors such as dermoids, gliomas, and encephaloceles that present as nasal deformities. These will be covered in other chapters in this book.

**Choanal Atresia**

The newborn with bilateral choanal atresia is usually brought to the otolaryngologist's attention immediately after birth when the patient develops respiratory distress. After obtaining an adequate oral airway the diagnosis can be made by attempting to pass a suction catheter through the nostrils (Table 1A and B). If time and conditions permit, a 2.7 mm rigid endoscope can be used to directly visualize the atretic plate after properly decongesting the nasal mucus membranes. Computed tomography (CT) scanning gives the definitive diagnosis. However, its primary use is to identify the composition of the atretic plate prior to surgery.

**Table 1A. Choanal atresia**

1 to 7,000 live births
2 females to 1 male
2 unilaterals to bilateral
90 percent bony/10 percent membranous
50 percent associated with other congenital anomalies
The clinicians must keep in mind that many patients (20% to 50%) have associated anomalies, ie, the CHARGE association (coloboma, heart defects, atresia, retarded growth and central nervous system (CNS) abnormalities, genital hypoplasia, eardrum deformities). Choanal atresia occurs more often in females (2:1), most often unilaterally, and in 90% the atretic plate is bony.

**Table 1B. Choanal atresia - Diagnosis**

- **Bilateral - Birth - Respiratory distress relieved by oral airway**
- **Unilateral - Later - Unilateral rhinorrhea**
- **CAT Scan Finding:**
  1. Narrow nasal cavities
  2. Lateral bony obstruction
  3. Medial (vomer) bony obstruction
  4. Membranous obstruction
  5. Bony choanal plate.

The definitive cause for choanal atresia is unknown. There are four basic theories: (a) persistence of the buccopharyngeal membrane, (b) persistence of the nasobuccal membrane of Hochstetter, (c) the abnormal persistence or location of mesoderm, or (d) abnormal neural crest migration.

**Table 2A. Choanal atresia - Treatment**

- **Bilateral - Early**
  - Transnasal: 1. Blind puncture
  - 2. Visually assisted - microscope and/or endoscope
  - Transpalatal - More widely used
  - Unilateral - Elective

The treatment is surgical (Table 2A and B). Various methods have been employed: transpalatal, transseptal, and transnasal puncture. Richardson and Osguthorpe found that 83% of transpalatal procedures were successful and Ferguson and Neel also reported excellent results in a small number of patients. We prefer to use the brilliant illumination of the rigid fiberoptic endoscope to perform this procedure intranasally in those cases in which the stenosis is membranous or when the bony plate is not thick and when the nasopharynx does not have a lateral bony constriction. In the older child, the endoscopic approach is also preferable as a larger size of structures will allow this surgery to be more easily performed. However, in the infant with bony atresia who does not respond to medical management, the transpalatal approach is quite appropriate. In using the transpalatal approach, we prefer the inverted horseshoe approach rather than midline splitting as we feel this is functionally superior and has fewer complications.

**Table 2B. Choanal atresia - Key Surgical Aspects**

1. Conserve periosteum and mucosa
2. In addition to atresial plate, remove posterior septum and lateral bony buttresses.
Nasal Duplication

Fortunately, this entity is exceedingly rare (only three reported cases) as the obvious deformity; if uncorrected, it would be psychologically devastating. The patients present with two nearly complete external noses and hypertelorism with a nonfunctional nasal airway.

The origin of this deformity is unknown. The abnormality obviously arises before the nasal placodes develop. Interestingly, there is a report of "tripe nostrils" in which the author also postulates that an extra nasal placode is theoretically possible. However, a recognizable nose or pseudonose structure does not develop.

Treatment is initiated early for functional and cosmetic reasons. The first goal is to provide an adequate nasal airway using procedures to perforate the atretic plates. The second portion of the procedure is to remove the medial portion of each "nose" and one septum is removed. The remaining lateral portions are approximated to give a more normal appearance.

Proboscis Lateralis

This is another rare deformity that presents as hypertelorism with a nose-like structure located at the inner canthus and oriented in a plane perpendicular to the normal nose. Midfacial and nasal development may be affected. The embryology is also unknown but probably involves a similar mechanism of abnormal nasal placode development. Whether Hengerer and Oas' theory of abnormal neural crest migration, as they mentioned in their theory of choanal atresia, is involved in some or all of these nasal deformities, has not been proven.

Treatment is surgical, first to ensure an adequate nasal airway, and second to establish a more acceptable countenance.

"Cyrano de Bergerac" Hemangiomas

This deformity is caused by a capillary or capillary-cavernous hemangioma primarily involving the nasal tip, giving rise to the "Cyrano" appearance. Multiple modalities have been used to treat hemangiomas of the head and neck, such as excision, steroids, and expectant treatment. Spectacular results due to spontaneous involution and without active medical management frequently occur.

This conservative mode of thinking may be the best choice for the Cyrano lesions also. Thomson and Lanigan retrospectively reviewed their experience and found that although at time difficult, the conservative treatment, at least initially, is probably the best. In selected patients, surgical treatment may be the more conservative approach. For example, with persistence and growth, there are selected cases in which the appearance is psychologically disturbing or in which bleeding is a significant problem. Although carbon dioxide freezing and laser surgical excision have been found to be appealing, in our experience the straightforward surgical excision utilizing a modified external rhinoplasty approach is quite successful. Bleeding, surprisingly, is easily controlled by the usual methods.
Birth Trauma

Deformities of the nose, independent of the genetic or embryologic effects, are commonly caused by trauma in utero or during delivery. It is common to see nasal flattening at birth; however, this normally corrects itself. Closed digital manipulation or selected surgical instrumentation of severely displaced structures is appropriate. However, open approaches are seldom, if ever, indicated. It is surprising that, even in those infants in whom the external nose appears markedly displaced, self-correction is usually the rule. Jazbi studied 100 consecutive cases from 7,129 live births and found a frequency of occurrence of less than 1%. The deformities were generally caused by septal dislocation. In older children whose deviations were not corrected at birth, septoplasty may need to be performed by any of the various acceptable approaches, considering each patient and the deformity on an individual basis. Healy has obtained excellent exposure with minimal morbidity using a sublabial approach.

Septoplasty

If the projection of the nose and midfacial development in general is dependent upon the nasal septum, then it is logical to strive to attain an acceptable nasal airway prior to the "point of no return". As previously mentioned, Gray showed that watchful waiting did not allow septal deviations to correct themselves. If a patient has an obstruction, then a minimal, closed procedure is our usual approach. The sublabial approach of Healy certainly seems reasonable. We have not yet used the total septal reconstruction approach of Jugo, though his experience would indicate that this is a reasonable approach.

Septorhinoplasty in Older Children and Adolescents

In children whose deformity will not be sufficiently corrected by septoplasty alone, consideration of reconstructive septorhinoplasty is appropriate. These are primarily children in whom the nasal pyramid as well as the septum are deviated and there is significant nasal obstruction. There has been no firm evidence to show that simple osteotomies to reposition the bony pyramid has resulted in growth deformities. On the other hand, the temptation to perform complete reconstructive septorhinoplasty, including resection of the dorsal tissue, should usually be resisted. However, dorsal augmentation should not be unduly delayed. It has been our philosophy to correct severe deviations by repositioning and augmentation at an age when these problems become physiologically or psychologically important, with the full understanding that further correction may be desired after full growth has been obtained.

Cosmetic septorhinoplasty in older adolescents is quite rewarding; however, the psychological aspects demand careful consideration. In our experience involving both adults and adolescents, there are a greater proportion of psychological pitfalls in the adolescents; however, these are usually more easily managed than when present in the adult. To manage them, they must first be identified and if the surgeon is not fully capable in dealing with these problems, psychological consultation should be obtained. However, this has been surprisingly infrequent. The surgeon who is comfortable in dealing with adolescents will find the adolescent among his most satisfied and satisfying patients.
Proper, informed consent is especially important in these patients, with specific attention to the possible effects on future growth. The younger the adolescent, the more likely there will be further growth changes, changing the appearance over time. In addition to the age guidelines given by Buck and Brown concerning growth, each patient must be considered on an individual basis with regard to size and with reference to parents and siblings, with regard to whether the patient has completed pubertal growth, and particularly, with regard to whether there had been growth in the last 6 months.

Even when all these factors are considered, there may be further growth. A slight alteration in structure will not significantly affect the appearance; however, if there is significant change, it will usually be as a result of increased dorsal projection, which can later be managed with minor revisional surgery. There is increasing evidence that careful conservative septorhinoplasty in younger patients can be safely performed. Fedor has recently reported satisfactory results in 22 patients where conservative surgical guidelines were carefully followed.

The guiding surgical principle of septorhinoplasty in children is to never overoperate - less is better. Attempts are made to achieve the desired result by repositioning tissues whenever possible. Incising, reshaping, and resecting of tissue is only done as a last resort with resection of tissue kept to an absolute minimum. It should be remembered that it is much easier to later correct errors of omission than those of commission.

Table of Guidelines

1. Do as little and as few surgical maneuvers as possible to achieve satisfactory results. Repositioning is preferable to reshaping and incising. Reshaping is preferable to resecting.

2. Use only lateral osteotomies and avoid medial osteotomies unless absolutely necessary. Unless absolutely necessary, do not separate the upper lateral cartilage from the septum.

3. Use sharp dissection to modify the cartilaginous profile. The bony profile in the adolescent can be generally remodeled with a rasp, as the tissue is immature and responds readily to rasping. This aids in avoiding overresection.

4. Do not do extensive tip surgery in the younger patient, and in the adolescent use the most conservative tip modification approach to produce the desired result. Particularly avoid overresection of the lateral crus.

5. Augmentation measures are generally satisfactory in the adolescent.

Nasal Trauma

All children sustaining significant trauma to the nose should receive a thorough, detailed examination, whether treatment is apparently indicated or not. (For a discussion of soft tissue injuries, see the chapter by Farrior and Clark.) Of particular importance, it should be noted that soft tissue defects should have early repair, and that scars of the nose in children
and adolescents are erythematous for an extended period of time before "fading". Nasal fractures are a common event throughout childhood. Trauma in this area may influence further development of the premaxillary and maxillary septal and nasal bony elements. Unfortunately, on occasion, even after accurate diagnosis and adequate treatment, further growth and development may be affected. An important anatomical feature to remember in assessing and treating nasal injuries is that the smaller the child, the larger the cartilaginous proportion of the external pyramid. The nasal bones are formed on the surface of the cartilaginous nasal capsule and are initially separated in the midline by an open suture. There is a large overlap of the nasal bones on the upper lateral cartilages. If these are detached, upper lateral cartilage can be prolapsed into the nasal cavity, creating an obstruction. Fractures of the septal cartilage may occur with or without nasal, bony fractures. An untreated greenstick fracture of the septum may result in progressive deformity.

Septal hematoma occurs more frequently in children due to the loose attachment and mobility of various anatomical components. If untreated, this frequently leads to septal abscess with cartilage resorption and subsequent "saddle nose" deformity. The hematoma should be evacuated as early as possible and the septal components sutured together with a mattress type suture, following which the nose is packed. If an abscess has occurred, immediate evacuation with thorough irrigation should be undertaken. Close inspection of the cartilage for evidence of absorption should be done at the time of surgery. A drain is left in place, the nose is packed, and appropriate antibiotics are administered for at least 2 weeks. If the cartilage has absorbed, the patient must then be observed closely after healing and should dorsal depression become evident, early grafting (usually with conchal cartilage) will prevent progressive "saddling".

Nasal Obstruction Due to Other Causes

Nasal obstruction is not always secondary to septal deformities; consequently, an accurate etiologic diagnosis is required. Allergy and other metabolic conditions such as cystic fibrosis require proper management. Other causes that may require surgical management include nasal polyposis, chronic hypertrophic sinusitis, turbinate hypertrophy (especially pneumatization of the middle turbinate, ie, concha bullosa). It is our practice to include a coronal computed tomography (CT) scan as part of the diagnostic evaluation in all children with airway obstruction not obviously due to septal nasal deformity or obvious medical causes. Many of these patients will have an excellent response to functional sinus surgery and/or turbinoplasty. (The details of evaluation and surgery are beyond the scope of this chapter.)
This chapter deals with congenital neck masses. By definition, *congenital* means present at birth, and a mass lesion is one that represents an abnormal regional increase in volume. By definition, therefore, congenital neck masses would be exceedingly rare. If, however, we consider congenital to encompass what is present at birth becoming obvious later, then the incidence of congenital neck masses increases dramatically. A number of congenital cervical anomalies will not present as masses, eg, anomalies of the skin.

Congenital masses will be described by tissue types, and anomalies of the branchial arch apparatus will be described collectively. Vascular and lymphatic masses will be discussed in another section of the book and will not be described here; neither will the more frequent benign and malignant true neoplasms.

**Embryology of the Neck**

Only the briefest outline of the embryology of the neck is possible in this format. The neck develops from the derivatives of the branchial arches. In the human only five arches are represented, I, II, III, IV, and VI - arch V is not detectable in the human. The branchial arch apparatus is first identifiable in the 3.0-mm embryo as tiny, smooth, rounded projections immediately anterior to the notochord in the area between the cephalon and the chest. At this point the embryo consists of two masses, a head and a chest/abdomen. There is no neck.

Each branchial arch consists of a central core of mesoderm that differentiates into a skeletal bar, an artery, nerve, and muscles. The external surfaces are covered by ectoderm and internally they are lined by entoderm. The arches expand ventrally so that by the 5-mm stage they are recognizable as raised cylindrical structures. Between adjacent arches externally are grooves or clefts, and internally similar depressions are called pouches.

Each arch is the precursor of definitive structures as is each cleft and each pouch. The more important derivatives of the arches in the human are listed in Table 1.

Continued growth in a ventral direction brings right and left branchial arches together in the midline where they fuse to form the neck. The lateral surface of the neck is smoothed by a projection from the ectodermal surface of the second arch called the operculum that grows caudally to fuse with a smaller projection from the third arch. There is, therefore, a potential space between the operculum and the lateral surface of the third arch, the cervical sinus of His. Theoretically, a second branchial cleft cyst develops if remnants of the cervical sinus persists. The potential anomalous development of the branchial arch apparatus are listed in Table 2. Only the more common and a few rare anomalies are noted.

The final position of the third pouch has not been definitely decided. Internal sinuses
developing from the lateral pharyngeal wall including the pyriform sinus might be from third or fourth pouches.

**Table 1. Derivatives of the branchial arch apparatus**

<table>
<thead>
<tr>
<th>Cleft (ectoderm)</th>
<th>Derivative</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st</td>
<td>Ext aud canal</td>
</tr>
<tr>
<td>2nd</td>
<td>Cervical sinus</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Pouch (endoderm)</th>
<th>Derivative</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st</td>
<td>Eustachian tube/middle ear/mastoid cells</td>
</tr>
<tr>
<td>2nd</td>
<td>Tonsil</td>
</tr>
<tr>
<td>3rd</td>
<td>Inf parathyroid, thymus</td>
</tr>
<tr>
<td>4th</td>
<td>Sup parathyroid, ultimobranchial body</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Arch (skeleton)</th>
<th>Derivative</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st</td>
<td>Malleus, incus, mandible</td>
</tr>
<tr>
<td>2nd</td>
<td>Stapes, styloid process, stylohyoid lig; sup half body of hyoid bone</td>
</tr>
<tr>
<td>3rd</td>
<td>Inf half body of hyoid bone; gt cornua</td>
</tr>
<tr>
<td>4th and 6th</td>
<td>Laryngeal cartilages</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Nerve</th>
<th>Ligament</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st arch</td>
<td>Temporalis, masseter, pterygoideus, mylohyoid, t tympani, t palatini</td>
<td>VIth Vth</td>
</tr>
<tr>
<td>2nd arch</td>
<td>Muscles of face, stapedius, stylohyoid, postdigastric</td>
<td>VIIth</td>
</tr>
<tr>
<td>3rd arch</td>
<td>Stylopharyngeus, constrictors</td>
<td>XIt IXth</td>
</tr>
<tr>
<td>4th/6th arch</td>
<td>Laryngeal constrictors</td>
<td>Sup and rec laryngeal</td>
</tr>
</tbody>
</table>

**Table 2. Developmental anomalies of the branchial arch apparatus**

<table>
<thead>
<tr>
<th>Arch</th>
<th>Anomaly</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st</td>
<td>Aplasia/dysplasia malleus, incus, mandible</td>
</tr>
<tr>
<td>Groove</td>
<td>Anomalies of external auditory canal</td>
</tr>
<tr>
<td>Pouch</td>
<td>Rare - diverticulum</td>
</tr>
<tr>
<td>2nd</td>
<td>Anomalies stapes and styloid process</td>
</tr>
<tr>
<td>Groove</td>
<td>Branchial cleft cyst</td>
</tr>
<tr>
<td>Pouch</td>
<td>Sinus in tonsil, fistula pharynx to neck</td>
</tr>
<tr>
<td>3rd</td>
<td>Cervical thymus, thymic cyst, aplasia parathyroid</td>
</tr>
<tr>
<td>4th</td>
<td>Congenital laryngeal stenosis or cleft</td>
</tr>
<tr>
<td>Pouch</td>
<td>Sinus from pyriform sinus, aplasia parathyroid gland.</td>
</tr>
</tbody>
</table>

**Anomalies of the Skin**
Congenital masses that arise from the ectoderm are exceedingly rare. There are, however, a number of skin-related problems that are of interest to the surgeon.

**Pterygium Colli (Winged Neck)**

A pterygium is a fold of skin with a core of connective tissue extending from the mastoid process to the point of the shoulder. Pterygia are found in Turner's syndrome, Noonan's syndrome, the multiple pterygium syndrome, and the leopard syndrome.

**Turner's Syndrome**

Turner's syndrome is caused by the absence of an X chromosome in a female. It is characterized by a short stature, webbed neck, widely spaced nipples, sexual infantilism, short metacarpals, cubitus valgus, coarctation of the aorta, and midfrequencies sensorineural hearing loss.

**Noonan's Syndrome**

Noonan's syndrome is characterized by short stature, webbed neck, hypertelorism, and cardiac anomalies. With development the face becomes almost triangular. Noonan's is probably a dominant syndrome. Females are usually fertile, whereas male fertility is variable.

**Multiple Pterygium Syndrome**

This syndrome consists of pterygia of the neck, antecubital and popliteal areas, fingers, and axillae, as well as a short stature and cleft palate. The multiple pterygium syndrome is probably recessive inherited with a background of consanguinity.

**Leopard Syndrome**

This is a mnemonic (1) for a syndrome that consists of multiple lentigines, electrocardiographic abnormalities, ocular hypertelorism, pulmonic stenosis, abnormal genitalia, retardation of growth, and sensorineural deafness. Mild pterygium colli are frequently present.

If cosmetically desirable, pterygia are treated by excision and Z-plasties.

**Midline Cervical Clefts**

These are rare anomalies in which there is an absence of normal tissue in the midline of the neck. Instead, there is a band of fibrous tissue extending from the mandibular symphysis to the sternum. Clefts present in the neonate as a linear raised area covered by thin, pink skin. The cleft tissue grows more slowly than the surrounding normal tissue, and untreated, it tethers the mandible inferiorly. A computed tomography (CT) scan or magnetic resonance imaging (MRI) will identify the depth of the lesion and also will ensure the presence of normal structures such as the thyroid gland. Midline cervical clefts are treated by excision and repair with either Z-plasties or a broken-line closure.
Anomalies of the Musculoskeletal System

Anomalies of the skeleton of the neck occur not infrequently but usually do not present as a mass.

The vertebral column or the clavicle might be affected. Only two skeletal anomalies will be discussed here.

Anomalies of the Vertebral Column

Klippel-Feil Syndrome

This is a condition in which the neck is abnormally shortened because of developmental anomalies of the vertebrae. There might be hemivertebrae or fused or absent vertebral bodies. The cervical vertebral column is frequently represented by a shortened almost solid block of bone with narrow or absent intravertebral discs or fused articular facets. The result is a short or almost absent neck. Rotation of the neck and head is grossly limited or absent, but flexion and extension at the atlanto-occipital joint is normal. The posterior hairline might be at the interscapular level and the chin might almost be resting on the sternum. One-third of these cases have associated hearing problems. The shortness of the vertebral column causes prominence of the trapezius muscles and the clinical impression of a winged neck that should not be confused with a pterygium.

As the vertebral anomalies in this syndrome are variable, difficulty of intubation for general anesthesia varies accordingly. In mild cases intubation might not present a problem in childhood, but in more severe cases, blind nasal intubation or intubation over a fiberoptic endoscope might be necessary.

Anomalies of the Clavicle

The clavicle develops from at least two segments - medial and lateral - that fuse. If fusion fails, a pseudojoint in midclavicle might present as a mass-like lesion. A simple radiologic study identifies the problem. No specific treatment is indicated, but if any type of surgical intervention is planned, then the underlying vascular structures, such as the subclavian artery and vein, must be protected.

Cleidocranial Dysostosis

In this syndrome there is aplasia or dysplasia of the clavicle occasionally with absence of a part or whole of one clavicle. There are also anomalies of the skull and frequently mental retardation and sensorineural hearing loss. The characteristic clinical finding is the ability to bring the shoulders into close apposition in the midline.

Torticollis (Wryneck)

Torticollis is defined as a tethering of the neck to one side with concomitant rotation of the chin to the contralateral side. The usual cause is contraction of the sternocleidomastoid muscle. Other causes might be maldevelopment or inflammatory disease of the cervical spine.
In the so-called congenital torticollis, the sternocleidomastoid muscle frequently develops a firm tumor-like swelling in its lower third. This represents a mass of fibrous tissue and is lily white and gritty in consistency - the white tumor of the newborn. The cause of these tumors is speculative ranging from in vivo venous occlusion to a hamartoma to even the ever-present probability of birth trauma. This condition can be treated by vigorous stretching exercises four or more times daily. If, however, the muscle continues to contract, then surgical section is advisable through a small horizontal incision in the lower neck. This should preferably be done before the first birthday. Muscle lengthening techniques give no better results. Persistence of torticollis results in asymmetry of the face with hypoplasia of the "down side."

The Branchial Arch Apparatus

Anomalies of the First Branchial Cleft

Anomalies of the first branchial cleft are those of the external auditory canal and are classified as aplasia, atresia, stenosis, and duplication anomalies. Here, we are concerned only with duplication anomalies. Work subdivided duplication anomalies into two types:

Type 1 - A skin-lined tract parallels the existing external auditory canal from a blind pouch laterally to the region of the middle ear medially.

Type 2 - Anomalies also consist of a squamous epithelial-lined tract ending in a blind pouch laterally or inferiorly that opens directly into the external auditory canal. Work considered these to be a composite containing ectodermal and mesodermal derivatives. In both cases, the tract becomes filled with keratin and might present because of infection, or as a mass in the upper neck or anywhere in the periauricular area. Clinically, they are diagnosed as recurrent abscesses usually treated by incision and drainage. Alternatively, because of the keratin material, they are frequently called extratemporal bone cholesteatoma.

Duplication anomalies usually require surgical excision. Because both types of anomalies have varying relationships to the facial nerve, the first step in surgery should be identification of the facial nerve in the usual manner and then careful excision of the tract. We recommend a standard parotidectomy incision that includes an ellipse of skin around any external fistula. After the facial nerve is identified, removal of the tract begins from lateral to medial. The tract is dissected through the branches of the facial nerve as necessary, and followed to its deepest point.

Anomalies of the Second Branchial Arch Apparatus

Anomalies of the Second Branchial Groove/Cleft

Persistence of the second branchial groove might present as a sinus, cyst, or fistula.
Second Branchial Sinuses

A sinus is a blind tract open on an epithelial surface. Second branchial cleft sinuses open on the skin, are usually sited along the anterior border of the sternocleidomastoid muscle at the level of the hyoid bone or thyrohyoid membrane and are frequently bilateral. They may drain small quantities of clear fluid probably produced by heterotopic salivary glands, which are sometimes seen in the wall of the sinuses. Tracts at this level are usually about 2 cm long, penetrate the platysma, and end blindly. They are easily excised.

Second cleft sinuses should not be confused with the rare first arch fistula that might open in the same position and track upward and posteriorly toward the external auditory canal.

Second Branchial Cleft Cysts

Cysts of the second branchial cleft might be present at birth, in early childhood, in late teenage or young adulthood, or in the fourth to fifth decade of life. The trigger is usually inflammation or frank infection. The typical position of a second branchial cleft cyst is deep to the sternocleidomastoid muscle at about the level of the hyoid bone, frequently protruding anteriorly beyond the anterior border of the muscle. Their sizes vary considerably, and in the adult might expand rapidly to be many centimeters in diameter. The popular embryological explanation of a branchial cleft cyst is a persistence of the epithelial remnants of the cervical sinus of His. Although this is a simple concept that provides a reasonable explanation, it does not quite fit the embryological pattern as Fraser, in a careful study, showed that the sinus is obliterated from its depths. Although second cleft cysts can be easily diagnosed clinically, it is wiser to confirm the diagnosis by ultrasonography and/or CT scans. As these cysts might be associated with sinus or fistulous tracts that might ascent to open into the region of the tonsil fossa or might descend to the lower neck, these patients should have their mouths and pharynges thoroughly examined.

Branchial cleft cysts are treated by surgical excision via a horizontal incision. Uninfected cysts are easily separated from the surrounding tissues. As the cysts lie lateral to the carotid sheath, they should be carefully dissected, preferably by blunt dissection.

Occasionally there are tracts leading from the deep surface. One tract might run superiorly between the internal and external carotid arteries and might even penetrate the middle constrictor muscle to open internally into the oropharynx close to or through the tonsil fossa. The lower tract might extend inferiorly toward the lower anterior border of the sternocleidomastoid muscle. The surgeon, therefore, must be prepared to follow and excise these tracts. Incomplete removal will result in the potential for recurrent infection or draining sinuses.

Acutely infected cysts are much more difficult to excise because their walls are thicker, surgical landmarks are obscured, and the characteristic lymphoid nodules in the cyst walls might be substantially enlarged. It is better, therefore, if possible, to cool an infected cyst with vigorous antibiotic treatment before attempting excision.

Second Branchial Cleft Fistulae
A fistula of the second branchial groove and cleft consists of an epithelial-lined tract with an internal opening in or adjacent to the pharyngeal tonsil and an external opening along the lower third of the anterior border of the sternocleidomastoid muscle. The tract runs between the external and internal carotid arteries penetrating the middle constrictor muscle superiorly and the platysma inferiorly, and can be outlined by contrast radiography. Branchial fistulae are excised electively if they tend to have recurrent infection or for cosmesis. Usually two ladder-pattern horizontal incisions are necessary, one at the level of the external opening and the second at the level of the hyoid bone. The tract must be traced to the constrictor muscle, and ligated and severed at this level.

**Persistent Branchial Fistulae**

Incomplete excision of branchial pouch or cleft anomalies usually results in recurrent infections in the neck. Investigation of these cases is not easy. A barium swallow, computed tomography, ultrasonography, and, if there is an external opening, contrast radiography, might all be of value. Frequently, however, the neck needs to be explored surgically using the details of previous surgery as guidelines.

**Derivatives of the Third Branchial Pouch**

The corresponding final place of the third branchial pouch has not been precisely determined. A possibility might be at the level of the vallecula or thyrohyoid membrane. Similarly, there is some controversy about the position of the fourth pouch, which traditionally has been supposed to be the pyriform sinus. It would seem wise, therefore, to consider third and fourth pouch derivatives together, particularly as the parathyroid glands are derived from both pouches, and the thymus gland from the third pouch. The thymus gland and parathyroid glands are frequently closely related in the developed organism.

Persistence of the third/fourth endodermal epithelial pouches occurs rarely. They occur as a blind sinus tract off the pyriform sinus. The usual method of presentation is a spontaneous infection in the neck in the vicinity of the thyroid gland frequently diagnosed as thyroiditis. If an abscess forms, it is usually incised and drained, only to recur sometime in the future. We have seen cases with seven drainage procedures over a 10-year period. There might also be a persistent fistulous drainage through one of the neck incisions and very rarely a chronic phlegmon in the neck at the level of the thyroid gland. The correct diagnosis can be made by barium study that frequently, but not always, identifies a tract leading from the pyriform sinus. Occasionally, when the barium study is negative, the internal opening can be seen by direct laryngoscopy. If a third/fourth branchial pouch sinus is diagnosed, treatment is by excision via a horizontal collar-type incision, which gives access to the region of the thyroid gland. If possible, the tract should first be injected with methylene blue. The old scar and surrounding tissue is excised, with care being taken not to injure the recurrent laryngeal nerve. The tract should be followed to the pyriform sinus where it is ligated and amputated.

**Thymus Gland**

Arrest of the thymus glands in the neck is rare. They may present as cystic masses at birth or in adulthood. Less than 80 cases have been reported in the world literature. Graeber et al studied 46 patients with thymic cysts; of these, 7 were in the neck and 36 in the
mediastinum. The neck masses were in the midline, paramedian, or lateral planes. There are, therefore, no distinguishing features for cervical thymic cysts, but there is frequently an associated hypertrophy of the ipsilateral parathyroid gland and consequently a few of the reported cysts have concomitant disorders of calcium metabolism. It is imperative, therefore, that cyst neck masses should be investigated by ultrasonography, CT scans or MRI, and screening of calcium metabolism. The superior mediastinum must also be assessed.

**Thyroglossal Duct Cysts**

The thyroid gland begins as a diverticulum from the floor of the embryonic pharynx just caudal to the tuberculum impar at a point that will become the foramen cecum. The diverticulum descends to the front of the neck in close relationship to the developing hyoid bone. The thyroid gland is in position pretracheally by the 8th week. The connection between the foramen cecum of the tongue and the thyroid gland forms the thyroglossal duct. The duct usually disappears but might persist anywhere along its length. These persistent areas expand to become thyroglossal duct cysts.

Thyroglossal duct cysts are more frequent in children but might present in adults. The oldest that I have seen is in a 75-year-old man. The trigger for presentation is usually inflammation or frank infection. They are characteristically in the midline of the neck or just lateral to, or just inferior to, or at the level of, the hyoid bone. There is, however, considerable variation in the positions of thyroglossal duct cysts.

Thyroglossal duct cysts are treated by excision. There must be preliminary investigation by radioisotope study, CT scan, or ultrasound to ensure that the mass is not an ectopic thyroid gland. Excision is accomplished by the Sistrunk procedure through a horizontal incision at the level of the hyoid bone. The cyst usually separates easily from the surrounding tissues, and the central 2-cm block of the hyoid bone is cleared of muscle attachments and removed. A block of tissue about 0.5 cm in diameter is then cored from the central hyoid to the foramen cecum and excised at that level. Failure to excise a thyroglossal duct completely will result in a persistent sinus or recurrent infection.

**Ectopic Thyroid Gland**

During its descent from the foramen cecum, the thyroid gland might come to rest in any point between the foramen cecum and the usual pretracheal position. Ectopic thyroid tissue, therefore, might be present in the substance of the tongue, around the hyoid bone, almost at the level of the cricoid cartilage or in front of the third to fourth tracheal ring, as is usual in the adult. Ectopic thyroid tissue is solid in contrast to a thyroglossal duct cyst. It is usually attached to the tongue and moves upward when the tongue is protruded. Any solid mass in the midline or just off the midline of the anterior neck, where there is a suspicion of ectopic thyroid tissue, should have the following investigations performed. Ultrasonography determines its consistency, and might also show the presence of absence of a normal thyroid gland. In addition, a technetium scan will identify functioning thyroid tissue. It is important
to ensure that ectopic thyroid tissue is not the patient's only functioning thyroid. Ectopic thyroid tissue should be excised only for cosmetic reasons, although there is controversy about whether or not there is an increased incidence of malignant degeneration in these masses.

**Teratomas**

Teratomas are compound tumors composed of tissue from more than one germ layer. They arise from cells that maintain their embryonic totipotentiality to differentiate into any of the three primary germ layers. Therefore, teratomas might contain skin, muscle, bone, etc. Understandably, teratomas are more frequent in association with the gonads but can occur anywhere on the body. Touran et al reported a teratoma in the neck of a newborn with metastases. Through the time of their report (1989) less than 150 cases of cervical teratomas had been described. Generally, however, cervical teratomas have less tendency to malignancy than those arising elsewhere.

Teratomas, therefore, might present as a mass in the neck or pharynx or any combination thereof, in a neonate, but also might be diagnosed in adulthood. The diagnosis is sometimes aided by the radiologic finding of calcification in the mass. As there is a definite tendency to malignant degeneration, teratomas should be excised as soon as possible. Preliminary investigation with CT scans, MRIs, and even ultrasound might be of value to determine the extent of the lesion and the surgical approaches that might be indicated. After excision, these patients should be carefully followed.
Vascular lesions are the most common congenital abnormality seen in humans. One in three infants will be born with a vascular mark of some type. Lesions range from small, barely noticeable vascular birthmarks to large, deforming lesions that may be quite grotesque and even life-threatening. Many fade rapidly, but others may increase in size rapidly.

Birthmarks have played a part in folklore for thousands of years. The question is whether maternal thoughts, desires, or trauma can produce a mark in the unborn child. In antiquity it was felt that ungratified desires, such as craving a certain food, like a strawberry, could produce a similarly shaped birthmark in the unborn child, leading to the term strawberry hemangioma. The notion that mothers could imprint their babies carried into the medial field through the Renaissance and affected medical teaching into the late 19th century. There were a few brave souls who argued against the theory of maternal impression and concluded that birthmarks were the result of faulty embryological development and not maternal impressions. Unfortunately, there is still the lingering suspicion by some people that maternal impressions may occur, which is perhaps reinforced by parental convictions. With this lingering doubt, the moth of a newborn child presenting with a vascular malformation may have feelings of guilt that she has done something wrong during the pregnancy and thus marked her child for life.

Birthmarks have had an interesting effect on art and literature. Mulliken and Young found a paucity of portrait paintings showing vascular birthmarks. The only one found was a portrait by Picasso in 1906 that showed a woman with a lymphatic anomaly of the lower cervical area. In literature the vascular birthmark has been associated with tragedy in stories by Shakespeare, Hawthorne, and V S Pritchett. For some reason people seem to shrink away from someone so marked by nature. Even today Mikhail Gorbachev, who has a visible vascular birthmark on his forehead and scalp, had it airbrushed out by newspaper photographers in Russia. In the USA it is the political cartoonists' favorite trademark when depicting Mr Gorbachev. Other important people have learned to turn their head in photographs so their birthmark is not visible. Birthmarks today continue to be a source of embarrassment for those individuals affected by these vascular lesions.

**Emotional Aspects**

It is natural for all parents to expect a perfect, normal baby at the end of a 9-month pregnancy. When this expectation is not realized, there is an emotional impact on the parents. If the lesion is in the highly visible area of the head and neck, or is large, there will be a major challenge to the parents' self-esteem and ability to parent. Their response will have a critical effect on the child's emotional adjustment to the birthmark. Many times the parents' reaction may be overprotective and not allow the child to develop his/her own potentials. As in many other cases of serious illness or congenital deformities, the stress of a large vascular
birthmark on the family unit may be so great that the family will break up. One of the parents will become so obsessed with the affected child that the needs of the spouse and other children are neglected. This will increase the stress and guilt feelings on the affected child, who may feel it is his/her fault the family has split up.

Harrison has divided personality development in an affected child into six stages. In infancy, the reaction is mainly on the part of the family. There is guilt on the mother's part, who blames herself for what she has done or eaten, feeling that maternal impressions do occur despite assurances to the contrary. With hemangiomas, only 30% are visible at birth, but within a few months all hemangiomas rapidly expand and grow, creating a great deal of anxiety in the mind of the parents. They want something done "now" to restore their "normal" child. During this time, when a new baby is normally shown off, the parents are reluctant to dress up and show off their new child for fear of negative reactions from friends and strangers. This sense of shame will affect the parenting process.

The toddler stage is from 12 to 24 months of age. At this stage the child will first become aware of the vascular lesion as the child begins to develop his/her own self-awareness. At this age all children begin to fear separation from parents, clinging at bedtime and being shy with strangers. This is a normal developmental stage, but if parents do not understand this they may interpret this reactions as frustration about the birthmark and press for early removal. Helping parents understand that these are normal developmental conflicts rather than concerns about the birthmark will help them deal with this stage in a more relaxed manner.

In the preschool period the child starts to identify with the parent of the same gender, but there are also feelings of inferiority and inadequacy in relationship to the adult competition. Bodily defects, such as a vascular birthmark, may become a focus for feelings of inferiority and badness, and as a result the child will act sad or regress to babyish behavior. The child may vacillate between positive attachment to the birthmark and feelings of inferiority or badness because of the birthmark. As a consequence the child may vacillate between wishing for surgery to get rid of the birthmark and positive feelings about his/her body, which the birthmark is part of, and not wanting to have it off. At this point the surgeon may want to counsel the parents to delay surgery and seek counseling for the child.

In the school-age child there is the beginning of separation from home. In the school situation, the child must develop competence in the academic arena, in sports, and in making friends. The birthmark may get mixed up in these developmental conflicts. The child may believe that the birthmark is the reason he cannot make friends or is not doing well in school. For the parents it is also a difficult time to sort out how much is related to the birthmark and how much is a lack of competence in peer relationships.

In the first 1 to 2 years of school the child seems to be accepted, but by age 7 to 8 years teasing and isolation become more acute with significant stress on the child's psychological development.

The next stage is adolescence, when there is the normal struggle with sexuality and a need to become more independent. It is a normal time for high anxiety in relationships, a need for peer acceptance, and concern for personal appearance. The presence of a vascular
birthmark compounds all of these problems and feelings. Rejection by the opposite sex is felt to be secondary to the birthmark. The normal child-parent conflict is intensified with the child possibly feeling rejected by his/her parents or blaming them. The normal embarrassing incidents of adolescence are compounded and made worse by the physical mark.

As one approaches and enters adulthood some acceptance will take place, but still there is the acute feeling that people are staring or rejecting one because of one's appearance. It may result in withdrawal or isolation to protect oneself. With help, the person can overcome these feelings, but there is still the desire to be rid of this abnormality. This leads to the hope by all of these patients, no matter what their age, that there will be a new miracle treatment to rid them of this unwanted birthmark. They are often willing to try any new treatment and will be happy with minimal improvement that the physician feels is a poor result at best.

Classification

The classification of vascular lesions has been an area of confusion for centuries with descriptive, anatomico-pathological, and embryological classifications. It has led to a confused body of literature and nomenclature using terms such as hemangioma, cavernous and capillary hemangioma, port-wine stains, nevus flammeus, lymphangioma, cystic hygroma, and hamartoma, just to state a few. In 1982, Mulliken and Glowacki proposed a histological classification based on the correlation of cellular features of the vascular lesions of infancy and childhood with the physical examination and natural history. On the basis of cell kinetics they identified two major types of birthmarks: hemangiomas, those demonstrating endothelial hyperplasia; and malformations, those with normal endothelial turnover. Table 1 compares the older terminology for vascular birthmarks to that proposed by Mulliken and Young.

Table 1. Translation from old terminology for vascular birthmarks into hemangioma or malformation

<table>
<thead>
<tr>
<th>Hemangioma</th>
<th>Old terminology</th>
<th>Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Capillary</td>
<td>Capillary</td>
<td>Malformation</td>
</tr>
<tr>
<td>Strawberry</td>
<td>Strawberry</td>
<td>Capillary</td>
</tr>
<tr>
<td>Capillary-cavernous</td>
<td>Capillary-cavernous</td>
<td>Capillary</td>
</tr>
<tr>
<td>Cavernous</td>
<td>Venous</td>
<td>Venous</td>
</tr>
<tr>
<td>Venous</td>
<td>Venous</td>
<td>Venous</td>
</tr>
<tr>
<td>Hemangio-lymphangioma</td>
<td>Venous</td>
<td>Lymphatic</td>
</tr>
<tr>
<td>Lymphangioma</td>
<td>Lymphatic</td>
<td>Lymphatic</td>
</tr>
<tr>
<td>Arteriovenous</td>
<td>Arteriovenous</td>
<td>Arteriovenous</td>
</tr>
</tbody>
</table>

With this classification the term hemangioma would be reserved for those vascular lesions that are common in infancy, which have a rapid growth phase characterized by hypercellularity and endothelial multiplication. This proliferation may result in a large cell mass that will necessitate the dilatation and formation of new feeding and draining vascular channels. These channels on angiography will give the appearance of a high-flow vascular lesion. There are also a few hypercellular tumors of vascular origin in adults that meet the
definition of hemangioma.

The second category, vascular malformations, exhibit a normal rate of endothelial cell turnover throughout their natural history. They are by definition congenital lesions, present at birth, although they may not all be obvious at that time. Some may be a vascular malformation or abnormality of the vessel wall that presents in adolescence or adulthood as the result of progressive ectasia. These vascular malformations grow with the child but may have gradual or sudden expansion secondary to flow-pressure changes, collateral formation, or hormonal modulation. These vascular malformations may be subdivided into capillary, venous, arterial, arterial-venous fistulae, and lymphatic anomalies. The capillary, venous, and lymphatic, or a combination of these, are "low-flow" lesions and the arteriovenous lesions are "high-flow".

**Table 2. Characteristics of vascular birthmarks**

<table>
<thead>
<tr>
<th></th>
<th>Hemangioma</th>
<th>Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical</strong></td>
<td>Usually nothing seen at birth, 30% as red macule</td>
<td>All present at birth; may not be evident</td>
</tr>
<tr>
<td></td>
<td>Rapid postnatal proliferation and slow involution</td>
<td>Commensurate growth; may expand as a result of trauma, sepsis, hormonal modulation</td>
</tr>
<tr>
<td></td>
<td>Female:male 3:1</td>
<td>Female:male 1:1</td>
</tr>
<tr>
<td><strong>Cellular</strong></td>
<td>Plump endothelium, increased turnover</td>
<td>Flat endothelium, slow turnover</td>
</tr>
<tr>
<td></td>
<td>Increased mast cells</td>
<td>Normal mast cell count</td>
</tr>
<tr>
<td></td>
<td>Multilaminated basement membrane</td>
<td>Normal thin basement membrane</td>
</tr>
<tr>
<td></td>
<td>Capillary tubule formation in vitro</td>
<td>Poor endothelial growth in vitro</td>
</tr>
<tr>
<td><strong>Hematological</strong></td>
<td>Primary platelet trapping: thrombocytopenia (Kassabach-Merritt syndrome)</td>
<td>Primary stasis (venous); localized consumptive coagulopathy</td>
</tr>
<tr>
<td><strong>Radiological</strong></td>
<td>Angiographic findings: well-circumscribed, intense lobular-parenchymal staining with equatorial vessels</td>
<td>Angiographic findings: diffuse, no parenchyma Low-flow: phleboliths, ectatic channels High-flow: enlarged, tortuos arteries with arteriovenous shunting</td>
</tr>
<tr>
<td><strong>Skeletal</strong></td>
<td>Infrequent &quot;mass effect&quot; on adjacent bone; hypertrophy rare</td>
<td>Low-flow: distortion, hypertrophy, or hypoplasia High-flow: destruction, distortion, or hypertrophy</td>
</tr>
</tbody>
</table>
The appeal of this simplified classification is that it is based on an accurate history, physical examination, and follow-up, but does not require complicated diagnostic techniques or biopsies. Table 2 presents the characteristics distinguishing hemangiomas from vascular malformations during infancy and childhood.

**Hemangioma**

**Natural History**

Hemangiomas are the most common tumor of infancy. About one-third will be visible at birth as a small macular spot or rarely as a full-size hemangioma. They are more common in fair-skinned families and occur in a 3:1 ratio female to male. There does not appear to be a familial incidence. About 60% occur in the head and neck region with 25% in the trunk area and 15% on the extremities.

Hemangiomas have two distinctive phases. During the first 1 to 6 months of life there is a rapid proliferation and expansion of the hemangioma. At this time there is a proliferation of endothelial cells that form syncytial masses with and without lumina. In the early stages this lesion will consist of solid masses of proliferating cells where lumen formation is difficult to appreciate, but later vascular spaces are not so compressed and capillary-sized lumina may be seen lined with plump endothelial cells. The proliferative stage is the hallmark of hemangiomas and the most distinctive feature in separating them from vascular malformations. During this phase if the lesion is in the superficial dermis, the skin will become raised with varying shades of crimson color and should be referred to as a superficial hemangioma. If the lesion involves the deep dermis and subcutaneous tissue, the skin over it will be smooth with normal color, but with a bluish hue from the underlying hemangioma and will frequently have dilated veins or telangiectatic vessels on the surface. This should be referred to as a deep hemangioma. As might be suspected many hemangiomas will present with involvement of both areas and as such will have clinical characteristics of both, but the histological pattern of proliferative endothelial cells is consistent throughout the tumor.

The involution phase begins at about 6 to 10 months of age and may be divided into an early and late stage. After the initial phase of rapid growth, which may last a few weeks to a few months, the hemangioma stabilizes and for a short period of time seems to grow at the same rate as the child. As involution begins to occur the color changes from a bright crimson to a more purple color. During this time there may still be some remaining areas of proliferation while involution is beginning in the center of the lesion. The surface begins to take on a grayish mottled appearance, especially near the center. The lesion begins to soften, the skin becomes wrinkled, bleeding and ulceration cease to be a problem, and the lesion no longer swells when the child cries. As involution proceeds it seems to spread in a centrifugal pattern, starting in the center and proceeding to the edges. As involution is completed the long-term result is variable. In some the skin will be near normal with mild atrophy and a few telangiectatic vessels; in others there may be significant atrophy with the appearance of scarfring. In some there is almost an empty sac of atrophied wrinkled skin, whereas the remaining may have fibrofatty infiltration leaving a mass with varying degrees of abnormal skin covering the surface. Histologically, there is decreased endothelial cellularity and progressive deposition of perivascular fibrofatty tissue. The vascular channels decrease as fibrosis increases and the remaining vessels dilate, with flattened, more normal-appearing
endothelial cells. Most clinical studies would indicate that complete resolution occurs in over 50% of children by age 5, in over 70% by age 7, and continued improvement until puberty. Usually the earlier involution begins, the better the final appearance will be.

**Differential Diagnosis**

The most important differential diagnosis is between hemangioma and vascular malformation. The clinical history is the most singly helpful distinguishing feature. Hemangiomas are not normally seen at birth, but within the first 1 to 2 months appear and rapidly grow at a rate beyond the child's growth. A vascular malformation will be present at birth and grow commensurately with the child. The early color of the hemangioma is usually bright crimson, and as involution proceeds it changes to dark purple, to a lilac, and ends as a yellowish to whitish color. Vascular malformation have a consistent color that is dependent on the depth of location and whether it is arterial, venous, capillary, or lymphatic. On palpation the hemangioma will have a firm or rubbery feel, whereas the venous malformations will be soft and compressible. In some circumstances the differential diagnosis will be more clear with time and the patient may need to be followed. In deep lesions, especially of the cheek or parotid area, a hemangioma is more likely to be confused with a lymphatic malformation, as both are rubbery to cystic on palpation and noncompressible. Hemorrhage into a lymphatic malformation may further confuse the picture. Both may rapidly expand under certain circumstances. In these circumstances I have found simple aspiration with a 20- or 22-gauge needle to be helpful in the differential diagnosis. If one gets clear, straw-colored fluid, the diagnosis of a lymphatic malformation is confirmed. Obviously pure blood would indicate a hemangioma or vascular malformation. Aspiration is relatively safe in this situation since pressure is easily applied to control bleeding. This is in contrast to potential hemangiomas of the viscera, especially the liver, where aspiration should never take place.

Another lesion that may occasionally be mistaken for a hemangioma is a pyogenic granuloma. It is a reactive proliferative lesion that histologically resembles a hemangioma and may be called a "capillary hemangioma" by the pathologist. These lesions appear suddenly, usually in children and occasionally in adults. They may occur on the cheek, eyelids, extremities, or on the mucosal surface of the oral mucosa or nose. They are usually granulomatous-type lesions that bleed easily and are pedunculated. They will occasionally occur in a capillary venous malformation (port-wine stain). These may be treated by removal and cauterization of the base with silver nitrate. If they recur, laser therapy or excision and closure may be necessary.

Radiographs are not usually needed for the differential diagnosis, but in deep lesions may be helpful. Computed tomography (CT) with contrast will help differentiate hemangiomas from vascular malformations. During the proliferative phase of hemangiomas the lesion will have a well-circumscribed homogeneous pattern, whereas during involution it will become heterogeneous with a lobular architecture. Venous malformations demonstrate a heterogeneous pattern with occasional calcifications. Lymphatic malformations show cystic elements with enhancement of the septa. The CT will also show the extent of the tumor, especially if resection is being contemplated.

Magnetic resonance imaging (MRI) scans are also helpful in distinguishing these lesions. Hemangiomas have high-flow characteristics with solid tissue of intermediate intensity.
on T1-weighted images and appear bright on T2-weighted appearances of high flow with flow voids and lack of solid tissue, which helps differentiate them from a hemangioma. Venous malformations are characterized by a low-flow pattern that has an intermediate intensity on a T1 image slightly greater than muscle and are very bright on T2 images. With gadolinium the venous channels enhance. The MRI is especially helpful in identifying multiple venous malformations in muscle. Lymphatic malformations have low intensity compared to muscle on T1 images, and on T2 images are bright. They may exhibit either cystic spaces with varying signal intensities or multiseptated spaces. Combined lesions may share characteristics of both types of malformations. The MRI may obviate the necessity for arteriograms in most low-flow lesions.

Angiography is not usually indicated in head and neck hemangiomas unless for some reason surgery is being contemplated or there is some unusual circumstance. The characteristics of a hemangioma are a well-circumscribed mass and intense, persistent tissue staining in a lobular pattern. Some hemangiomas have high blood flow, which suggests arteriovenous shunting and may be confused with tissue shunting seen in artiovenous malformations.

Complications

Several complications may occur in hemangiomas, usually during the proliferative phase. The physician must be alert to these as some may produce long-term or permanent disabilities.

In rapidly growing superficial hemangiomas the epidermis may be lost and cause ulcerations and/or bleeding. The bleeding is usually venous oozing or more rapid bleeding from a small punctate area. Both of these are easily controlled with pressure, but may be frightening to the parents, especially when there is an abrasion and bleeding while the child is asleep, leaving blood over the bedclothes. In rare circumstances these ulcerated areas may become infected, producing a septicemia or extensive necrosis and slough of soft tissue.

Rapidly enlarging hemangiomas surrounding or in the orbit may permanently affect vision. A lesion, such as in the upper eyelid, which obstructs vision for even short periods of time, may result in permanent effects on vision, such as amblyopia or failure to develop binocular vision. Periorbital hemangiomas may place pressure on the growing cornea causing distortion, which may produce astigmatism and/or myopia. Strabismus may result from paralysis secondary to infiltration of the muscles or secondary to amblyopia. All infants with hemangiomas in the orbital or periorbital area should have ophthalmological consultation and close follow-up. In lesions that are affecting vision, a more aggressive treatment plan may be necessary.

Another life-threatening area of obstruction is the subglottic area. These infants will develop biphasic stridor in the first 1 to 2 months of life, in contrast to a congenital subglottic stenosis in which there will be stridor from birth. The symptoms rapidly become worse and are worse with crying. They will respond temporarily to racemic epinephrine or subcutaneous epinephrine. About 50% will exhibit a cutaneous hemangioma elsewhere on the body. The diagnosis may be suspected by the appearance of a subglottic swelling on x-ray. It is confirmed by direct laryngoscopy and bronchoscopy with the typical findings of a soft
compressible subglottic mass that may or may not have telangiectatic vessels or a bright red appearance. The mass is not usually circumferential, but comes from the posterior and/or lateral walls. In contrast the congenital subglottic stenosis is firm, not dilatable and circumferential.

Other potential areas of obstruction in the head and neck are the nose and external ear canal. Theoretically a hemangioma that obstructs both sides of the nose could produce airway problems in the infant, who is an obligate nasal breather. In practice, total obstruction is rare and when it does occur it is slow enough for the infant to adapt to mouth breathing. However, one must be aware that these infants have the potential for significant sleep apnea secondary to this obstruction, which in severe cases may interfere with feeding and result in failure to thrive or right heart failure. If a hemangioma obstructs the external auditory canal the child will have a conductive hearing loss. This is not a problem if the hemangioma involutes and the canal opens up, unless it is bilateral. If it is bilateral then some type of amplification should be used by 6 to 8 months or there will be a delay in the child's speech development.

Skeletal distortion is a concern, but is relatively rare in hemangiomas. It is much more likely to occur in vascular or lymphatic malformations.

Bleeding from a superficial ulcerated lesion is unusual and is easily controlled. Of much more concern is the rare condition of Kasabach-Merritt syndrome. This is a generalized clotting disorder that manifests itself with petechiae, ecchymosis, or internal bleeding in a child with a large hemangioma. There appears to be platelet trapping during the proliferative phase, which results in a profound thrombocytopenia. This is a potentially fatal condition from bleeding into the hemangioma with pressure on vital structures or internal bleeding into the viscera or central venous system. Infants with a large hemangioma (greater than 5 cm in diameter) are at a greater risk and should be evaluated with a complete blood count including a platelet count. If thrombocytopenia is present, coagulation studies including prothrombin time (PT), activated partial thromboplastin time (PTT), fibrinogen, fibrin degradation products, and fibrinopeptide A should be obtained to look for evidence of a consumptive coagulopathy. The other serious complication to be watchful for in the Kasabach-Merritt syndrome is infection and septicemia. If this is suspected, wound and blood cultures should be obtained.

A final possible complication is congestive heart failure. This is rare in hemangiomas isolated to the head and neck, but may occur if they are large or multiple or there are associated visceral hemangiomas. It is felt this is the result of high output failure secondary to multiple hemangiomas requiring large amounts of blood. The infant will usually present at 2 to 8 weeks of age with congestive heart failure, hepatomegaly, and anemia. There is a high mortality rate associated with this condition. It is treated medically and if this is unsuccessful, consideration must be given to embolization and/or surgical intervention.

Treatment

One can imagine the fright and concern that new parents experience as suddenly from nowhere or from a small innocuous red birthmark there is the appearance of a rapidly expanding crimson tumor that is disfiguring their newborn child. In rare cases a large portion of the head and neck may be involved. The frightened parents seek medical help to arrest the growth of the tumor and restore their child to normal. The concerned physician may make
recommendations that are detrimental to the child if he is unaware of the natural history of hemangiomas versus vascular malformations. Concern that this represents a highly malignant tumor may lead to injudicious or unnecessary surgery. Parents are concerned that everything be done and may pressure the physician into doing something. It makes a sympathetic and concerned physician who is willing to listen to the parents' concerns and reassure them that watchful waiting is usually the best treatment. Care must be taken to watch for potential complications such as eye involvement, congestive heart failure, or coagulopathies and treat these entities expeditiously.

**Historical**

For centuries multiple types of therapy have been attempted to rid the newborn of this bright red stain. In antiquity various methods of folklore were used, such as rubbing the spot with the afterbirth, having the mother lick it, etc. Physicians for centuries have attempted ligation and/or excision and continue to be the source of reports for treating large "hemangiomas". The observation that hemangiomas that ulcerate would go on to heal and involute led to the widespread use of artificial ulceration, using agents such as astringents, caustics, refrigerants, and various bacterial and viral inoculums to produce an infection and scarring. As galvanic electricity became available in the late 19th century, electrolysis and thermocautery were widely used and were the forerunners of today's more sophisticated laser technology. Sclerosing agents were used widely in the 19th and early 20th centuries, and continue to be used in various forms today. Radiation therapy was widely used in the 1930s to 1950s. The rapidly proliferating hemangioma is very sensitive even to small dosages of 300 to 600 R, but there are well-documented risks of associated malignancies years later. As a result, the consensus is that radiation therapy does not have a place in the treatment of hemangiomas today.

Compression therapy was first advanced in the early 1800s and has been more recently suggested for the treatment of hemangiomas involving the extremities.

Spontaneous involution received some notice, but it was Lister's report of a prospective study in 1938 that documented the natural history of 93 hemangiomas that grew rapidly in the first few months of life, then went on to regress and disappear by about 5 years of age. Since this significant observation of spontaneous involution, the literature is replete with numerous articles debating which lesions will and will not involute. Unfortunately, the confusion is enhanced by the inconsistent terminology combining clinical findings with pathological descriptions of capillary, cavernous, and capillary-cavernous hemangiomas. The classification described by Mulliken and Glowacki limiting the term hemangioma to those rapidly proliferating lesions in the infant, which are the ones that will most likely undergo spontaneous involution no matter how fast they grow or how large they become, leads to a rational treatment plan for these unfortunate children.

**Contemporary Management**

**Observation**

Watchful waiting as a tumor is enlarging on a daily basis is most difficult and alarming for both parents and physicians. The former want something done and the latter wish
to do something. Inactivity in the face of a rapidly progressive tumor is galling for a surgeon whose life is spent doing something to improve patients' well-being. For those surgeons attuned to the importance of appearance it is particularly difficult to stand by and watch a beautiful infant rapidly become distorted and disfigured by this devastating lesion while the anguished parents are pleading for something to be done. One must temper this natural inclination to do something with the knowledge that hemangiomas will involute and any treatment we recommend must guarantee a better or more rapid outcome than spontaneous involution. Our overriding motto should be "do no harm".

At the initial consultation the physician must be willing to take the time to listen to the parents and explain the pathogenesis and natural history of hemangiomas in layman's terminology. It is particularly helpful if he has personal or textbook photographs demonstrating the stages of spontaneous involution. Special care must be taken to reassure the mother that it is not her fault or some activity or eating pattern that produced this growth. It is necessary to assure the parents that this is a benign tumor and that the need for active intervention is unlikely. During this initial visit one must be sensitive to the desperate feelings of the parents and if one senses doubts, to offer a second opinion from a reliable and knowledgeable physician. Many parents will go from physician to physician until they find someone who is willing to actively treat their child's growing tumor. During this initial visit photographs and measurements should be taken to document changes in the future.

Finally, close follow-up to reassure the parents and watch for possible complications should be arranged. The parents should be encouraged to call if they have concerns. If there is concern for vision or airway problems, the appropriate consultations should be arranged. Depending on the size and location, return visit intervals may be every 1 to 3 months during the proliferative phase and as long as 6 to 12 months once the lesion has stabilized and involution is starting.

Local Complications

If complications occur, the patient will need to be seen on more frequent intervals. The most common local complication is during the proliferative phase when ulceration with or without bleeding may occur. The ulceration from epithelial breakdown in a superficial hemangioma is treated with local care including gentle cleaning, topical antibiotic ointment, and a nonadherent dressing changed at frequent intervals. This may be an avenue for cellulitis and septicemia, which should be treated with the appropriate systemic antibiotics. It may take several weeks for the area to reepithelialize.

Bleeding from an ulceration or punctate area is another troublesome complication. Parents are particularly concerned, since they feel bleeding may become serious and uncontrollable. The bleeding can be controlled by gentle pressure with a nonadherent pad for at least 10 min. If bleeding continues, the child should be brought to the office or emergency room where cautery with silver nitrate or electrocautery will solve the problem. I have found it useful to teach the parents how to control the bleeding with pressure and then use a silver nitrate stick to cauterize the bleeding point. In a short period of time this ceases to be a problem. Neither ulceration or bleeding are indications for surgical removal, but persistent local bleeding may be an indication of a systemic coagulopathy and should be evaluated.
Steroid Therapy

Steroids were first used to treat thrombocytopenia associated with hemangiomas, but it wasn't until 1963 that Zarem and Edgerton noted a coincidental shrinkage of the hemangioma as well. Reported response rates have ranged from 30% to 90%. The exact mechanism of action by steroids is not clear. It is clear that the rapidly proliferating hemangioma is much more responsive than the stable or involuting lesion. It may be that the steroids have a direct effect by causing vasoconstriction of the channels and sinusoids of the immature hemangioma or by somehow modulating the control of endothelial proliferation. At the present time there is a great deal of research going on regarding angiogenetic factors that may shed light on the mechanism of steroid action, as well as provide new medical agents that may be used to treat hemangiomas. Crum et al have noted that there may be more active inhibition of angiogenesis by steroids in the presence of heparin or heparin fragments.

Mulliken gives the following guidelines for the use of systemic steroids:

1. A rapidly growing lesion that seriously distorts facial features.
2. A lesion where there is recurrent bleeding, ulceration, or infection.
3. A lesion that interferes with essential, normal, physiological functions (breathing, vision, eating, or hearing).
4. Large or multicentric hemangiomas causing bleeding (secondary to thrombocytopenia) and/or high cardiac output failure.

He recommends oral prednisone at 2 to 3 mg/kg/day for 2 to 3 weeks. In 7 to 10 days some response should be noted with decrease rate of growth, color change, and/or softening of the lesion. If there is no effect, steroid therapy should be discontinued. If there is a response the dosage should be tapered and if possible switched to an every-other-day dosage. Pulse therapy of 4 to 6 weeks is used and then a rest period, depending on the response of the lesion. In some cases there may be a rebound phenomenon that will require further courses of prednisone. Direct intralesional injection of long-acting steroids has also been advocated by some authors to minimize the long-term effect seen with systemic steroids. The efficacy of both methods seems to be comparable depending on the lesion, location, and report. Intramuscular or intravenous Decadron has a rapid effect on some subglottic hemangiomas.

Side effects of short-term high-dose steroids are minimal, but there are temporary effects such as slowing of growth, decreased appetite, hypertension, facial edema, and increased risk of infection, especially for otitis media and pneumonia. Because of these potential complications, it is advisable to use steroid therapy in conjunction with a pediatrician or pediatric endocrinologist.

Chemotherapy

There are sporadic reports of the use of chemotherapy in the treatment of hemangiomas. Hurvitz et al reported success with using cyclophosphamide in hemangiomas...
not responsive to other treatments.

**Orbital and Periorbital**

Hemangiomas of the upper eyelid may affect vision by producing astigmatismmmmm from pressure on the cornea itself or amblyopia. Large hemangiomas that occlude vision, even for short periods of time, may result in permanent visual loss. It is imperative that these infants be examined and followed by an ophthalmologist.

Rarely a small hemangioma may be surgically resected, but most of the time these lesions are full thickness in the lid and steroid therapy is the treatment of choice in the proliferative phase. Direct intralesional injection of a long-acting steroid, such as triamcinolone acetate or Depomedrol, has been used extensively. The technique is to use a 27-gauge needle with multiple punctures to spread the material throughout the lesion. A maximum of 40 mg of triamcinolone should be used at any one time. If the lesion extends into periorbital tissue there is a risk for hemorrhage or hematoma and risk to vision. In these situations systemic steroids are preferred. During treatment patching of the noninvolved eye may be necessary to minimize the development of amblyopia and strabismus.

**Subglottic Hemangioma**

Subglottic hemangiomas are a relatively rare cause of upper airway obstruction in infants. A child with cutaneous hemangiomas who presents with stridor and upper airway obstruction should be suspect for the presence of a subglottic hemangioma. The diagnosis is confirmed by direct laryngoscopy and bronchoscopy with the typical appearance of a bluish-red compressible mass or biopsy of the lesion. In an excellent review article, Shikhani et al reviewed all of the reported cases from 1913 to 1985 summarizing the myriad of treatments used in the past. Because of the acute airway obstruction, most patients have ended up with a tracheotomy. Treatments essentially mirror those used for the cutaneous lesions including sclerosing agents, cautery, open surgery, and observation. In the past, tracheotomy and observation were the recommended therapy. With involution by 2 to 3 years of age the child could be decannulated. Unfortunately, as pointed out by Shikhani et al, some series report over a 50% mortality rate secondary to complications of the tracheotomy. With better education of caregivers, better tracheotomy tubes, and in-home monitors, this figure has been significantly reduced, but there is still a risk plus the problems with wearing a tracheotomy tube and delayed speech development. Radiation therapy has the same risks as for cutaneous lesions and is not recommended. Open surgery in the infant larynx for subglottic hemangioma should also be condemned. Intralesions steroid injections have been used with and without tracheotomy. If they are used without tracheotomy the child must remain intubated for 5 to 10 days.

In the early 1970s the CO\textsubscript{2} laser began to be used for laryngeal lesions and the logical extension was to use it for subglottic lesions. The CO\textsubscript{2} laser works well, but there is the risk of cicatrical stenosis if the lesion is large or circumferential. Fortunately most subglottic hemangiomas involve posterior and one or both lateral walls. They should be treated segmentally, allowing for reepithelialization. No more than one-third of the circumference should be treated at one time. With recent experience using the neodymium:yttrium-garnet (Nd:YAG) laser for oral cavity and supraglottic submucosal hemangiomas, we have been
encouraged to try it on a subglottic hemangioma. A noncontact probe was used at 40 W and 0.3 sec. The patient was left intubated for 3 days to allow for swelling to decrease. In one case the results have been excellent, but obviously more cases are necessary to make a strong recommendation for the Nd:YAG laser over the CO₂ laser.

**Congestive Heart Failure**

Congestive heart failure is rare in children with hemangiomas limited to the head and neck. However, if a child presents with congestive heart failure and a head and neck hemangioma, care must be taken to look for other systemic hemangiomas, especially of the liver. If this is the case, steroid therapy is the first choice of therapy followed by angiography and embolization of the lesion if possible. If this is not successful or not indicated, surgical intervention may be the only alternative.

**Radiotherapy**

Although radiotherapy is not recommended, it may be necessary in some desperate situations such as for hepatic hemangiomas.

**Coagulopathy**

The Kasabach-Merritt syndrome, which is characterized by thrombocytopenia, is usually a self-limiting condition, but still may be fatal. Many children may tolerate a low platelet count, but if they become symptomatic with a bleeding problem, treatment must be instituted. Steroids are the first line of therapy and some would recommend using it with heparin. Platelet infusions are only a temporary measure since the problem is platelet entrapment. In those who fail steroid therapy, Hurvitz et al have recommended cyclophosphamide treatment.

**Surgical Therapy**

An attractive idea is that early surgical removal of the hemangioma may prevent its rapid growth. The fallacy of this approach is that it may always remain small and the resultant scar may look worse than allowing the natural course of events to occur, or the transformation may be a field event beyond the bounds of simple excision from the very onset. As stated earlier surgical intervention is seldom necessary, and then one must weigh the potential surgical outcome against the natural history of the lesion.

Rarely is surgery indicated for head and neck hemangiomas during the proliferative phase, although reports continue to appear in the literature where this is being recommended. Significant complications, including skin slough and facial nerve paralysis, are reported. Deep hemangiomas of the parotid, which expand rapidly, may create the concern that this represents a malignancy. Batsakis has pointed out that malignant tumors of the parotid gland are medical curiosities, but that hemangiomas that undergo involution make up the majority of tumors of the parotid gland in infants. Rapid growth is not an indication for surgery.

Early childhood, ie, before the child enters school, may be an opportune time to do resections for redundant skin or fibrofatty tissue residuals after involution has taken place.
Care must be taken to make these resections in relaxed skin tension lines to minimize scarring. Hemangiomas of the tip of the nose, sometimes called the "Pinocchio nose", are particularly obvious and will benefit from surgical intervention. These may be approached from an external rhinoplasty incision or a direct excisional approach.

Final scar revision, removal of excess fibrofatty tissue, and cosmetic touch-ups should be left until complete involution takes place. Ideally one would prefer to do this before puberty when hypertrophic scarring is more of a problem. General plastic and reconstructive principles and techniques must be used to obtain the best functional and cosmetic results. Abnormally pigmented or textured skin should be removed in serial excisions or in some areas by tissue expansion techniques for skin coverage of excised areas.

**Laser Therapy**

Various lasers have been used in the treatment of vascular lesions for nearly 20 years. The argon laser with a wavelength of 480 nm is preferentially absorbed by red cells. It has been useful in treating port-wine stains, but does not penetrate deep enough to affect a proliferating hemangioma. Other potential lasers are the copper vapor laser and the flashlamp pumped dye laser. Each has their own specific characteristics and place of use. Waner and Suen have advocated the use of the copper vapor or flashlamp pumped dye laser in treating the early proliferative phase of superficial hemangiomas. They point out the low scar rate, less than 1%, and the effectiveness of controlling lesions in the small numbers they have treated so far. Obviously more cases and longer follow-up are necessary to see if this treatment should be used in all cases.

The Nd:YAG laser has been successful in treating a variety of vascular malformations. It has a wavelength of 1.050 nm with the ability to penetrate up to 1 cm, but does produce skin changes and scarring. It may have a place in the treatment of bleeding or ulcerated lesions where skin changes are inevitable. However, where the epithelium is intact, the scarring left by the laser may be worse than the skin left by involution. I have used it in a few situations where it was obvious there was redundant skin that would later have to be excised in the hopes of hastening involution and decrease bleeding.

**Vascular Malformations**

**Pathogenesis**

The peripheral vascular system arises in situ from the primitive mesenchyme cells, but they are surrounded by pericytes and smooth muscle cells that arise from neuroectoderm. There is a complex development that takes place in three stages.

First, there is a period of an undifferential capillary network composed of interconnected blood lakes, but with no identifiable arterial or venous channels. At about 48 days in the human embryo, separate venous and arterial stems appear on either side of the capillary system in what is called the retiform stage. The final stage occurs by 2 months of life and involves the gradual replacement of these immature networks by mature vascular channels.
It is less clear how the lymphatic system develops. The most widely accepted theory is the "centrifugal theory", which postulates that the lymphatic system arises from a process of budding from the embryonic venous system to form five lymphatic sacs: two jugular, two posterior, and one retroperitoneal. Endothelial sprouting from these sacs form the mature peripheral lymphatic system at about the 9th week of development.

Many theories have been advocated as a cause of vascular malformations. There is some evidence based on the patterns of port-wine stains, hydrosis over malformations, etc, that there is a defect in the autonomic nervous system through the perivascular cells, which have contributed to vascular malformations. Multifactorial aberrations of normal development may also cause vascular malformations. Sequestered or maldeveloped areas of primitive capillary sets could result in venous or combined lymphatic and venous malformations. Failure of regression of arteriovenous communicators in the primitive rete could produce arteriovenous malformations. Abnormal dynamics in the developing system can also explain the particular morphology of some malformations. In lymphatic malformations the deep lymphatics are normal, but there is a failure of the cutaneous lymphatics to communicate with them, so it would appear there is a maldevelopment or sequestration of parts of the developing lymphatic system that form the lymphatic malformations.

In evaluating vascular birthmarks, the first step is to separate true hemangiomas from vascular malformations. At this point vascular malformations may be divided into "low-flow" and "high-flow" malformations. "Low-flow" anomalies may be further divided into capillary, lymphatic, and venous lesions; "high-low" anomalies are arteriovenous malformations that may have microscopic or macroscopic shunts. Each of these categories will be discussed separately.

Clinical Evaluation

Vascular malformations are much less common than other vascular birthmarks such as hemangiomas and vascular stains. Pratt examined 1,096 neonates and found a 1% incidence of hemangiomas, 10% incidence of capillary marks on the eyelids, forehead and nose, and a 42% incidence of macular birthmarks (nevus flammeus nuchae) in Caucasian infants. There were only 5 out of the 1,096 who had a true vascular malformation. Although vascular malformations are relatively rare, they persist throughout life, whereas the nuchae stains and capillary marks fade and hemangiomas involute.

At the initial evaluation it is sometimes difficult to distinguish hemangiomas from vascular malformations. Malformations will grow with the patient and are histologically stable. They do not regress, but under certain circumstances will enlarge, extend, or cause future problems, such as arteriovenous fistulas, and a few are progressive malformations. Some will enlarge secondary to hormonal stimulation such as pregnancy or taking birth control pills. Arteriovenous fistulas may not manifest themselves until adulthood. With this variability in mind, the patient must be followed to evaluate the clinical course of the lesion to make a better prognosis. In most circumstances if the lesions are stable, not affecting function or a significant cosmetic problem, a wait-and-see attitude is indicated. If this approach is to be taken, it is important that the patient be evaluated for other potential involvement such as the viscera, extremities, central nervous system, cardiovascular system, hemostatic abnormalities, and, in older patients, the possibility of a malignant lesion. It is not
within the scope of this chapter on vascular lesions of the head and neck to cover all the
described syndromes and lesions of vascular etiology in detail as this information is available
in textbooks devoted to the subject, such as the excellent work by Mulliken and Young.

Investigative studies are not normally necessary for vascular lesions of the head and
neck. Clinical examination and longitudinal follow-up are adequate for the majority of these
lesions unless there is a clear indication that management of the condition will be influenced
by the studies. Our desire to make an academic diagnosis should not be an indication for
expensive or unnecessary investigations. On the other hand, if active treatment is necessary,
thorough investigation is mandatory before any intervention is carried out.

First of all, one must determine if there are any hematological abnormalities. A history
of bleeding, recurrent thrombosis, or persistent anemia should raise suspicion that there is an
abnormality associated with the lesion. As previously discussed, with the profound
thrombocytopenia seen with some hemangiomas, a less severe localized consumptive
coagulopathy with normal or near-normal platelet count may occur in some vascular
malformations. These patients may be stable, but under the stress of surgery the coagulopathy
may become clinically significant with the possibility of localized and systemic bleeding.
Personally, I have seen this occur in two patients with lymphatic malformations. These
patients should be evaluated with a complete blood count, including a peripheral blood smear
and clotting studies including bleeding, prothrombin and partial thromboplastin time,
fibrinogen levels, fibrin degradation products, and fibrinopeptide levels. These studies should
be done preoperatively as well as postoperatively.

The mainstay of radiological evaluation for vascular lesions has been the angiogram,
but with newer modalities such as digital subtraction angiography (DSA), computed
tomography (CT), and magnetic resonance imaging (MRI), more useful information is
obtained. Although plain x-rays may be useful in evaluating skeletal structures of the
extremities involved with vascular lesions, they are seldom indicated in the head and neck.
CT with and without contrast is much more useful in evaluating the skeletal effects of a
vascular malformation of the head and neck. Not only is good skeletal detail seen, but spatial
relationships with location and size of the lesion may be visualized and some idea of its
vascularity may be determined if contrast is used.

Although MRI does not give good bony detail, soft tissue detail seen in a three-
dimensional plane is excellent. With its highly sensitive densitometry characteristics, it is
useful in distinguishing tissue planes, fat, mucus, fluid-filled cysts, lymphatic and venous
malformations, as well as giving some indication of whether a lesion is high flow or low flow.
Combined, CT and MRI give very useful information if surgery is contemplated or as a
noninvasive technique to follow the growth characteristics of deep lesions. They may replace
the necessity of an angiogram in some, but not all, situations, especially high-flow lesions.
They may be useful in directing the angiographer.

Angiography is a more invasive technique with higher risk, but with newer contrast
agents and improved catheters and techniques it has a very acceptable morbidity rate. The
angiogram will give the physician vital information as to the extent of the lesion, the feeding
and draining vessels, and whether embolization, thrombosis, or occlusion are viable treatment
options. It will help distinguish if a lesion is high flow or low flow and will differentiate an
arteriovenous (AV) malformation from a venous malformation.

**Capillary Malformations (Port-Wine Stains)**

Vascular birthmarks with a typical vivid reddish stain of the skin have been referred to as port-wine stains for centuries. In antiquity it appeared as if a deep red wine had been spilled on the patient's skin; hence the name. The Latin term for these vascular lesions is *nevus flammeus*. In the terminology used for this chapter taken from Mulliken and Young, these birthmarks would be classified as capillary malformations. However, the term *port-wine stain* is so ingrained in medical literature that we will bow to this usage. The term *capillary hemangioma* is confusing and should be abandoned as it is frequently used to describe hemangiomas discussed earlier. Capillary malformations or port-wine stains are not proliferative lesions and thus should not be called hemangiomas.

Port-wine stains are present at birth and remain throughout life without involution. The skin discoloration is usually evident at birth or shortly thereafter. There is equal sex distribution and the incidence is approximately 0.3% of all births.

Histologically these lesions are characterized by ectatic capillary to venular-sized channels located in the papillary and upper reticular dermis. The walls are thin and lined by flat, normal-appearing endothelium with normal cell turnover.

The port-wine stain is macular and sharply demarcated. It grows in proportion to the growth of the child. If the lesions are near mucous membranes there may be involvement of the underlying mucosa as well. Most patients will describe color changes with emotion, internal and external temperature changes, and exercise. The color ranges from a pale pink to a deep red. As the child matures, the pale pink color will deepen to a darker red in early adulthood, to a deeper purple in middle age. As the patient ages, the surface becomes more irregular and corrugated with multiple nodular lesions of ectatic vessels or venous lakes.

Port-wine stains are associated with many other malformation syndromes, especially on the trunk or extremities. In the head and neck area one may see other underlying developmental defects such as a meningoencephale or arteriovenous fistula.

The Sturge-Weber syndrome is one of the more common and feared vascular malformation complexes associated with port-wine staining. It is characterized by vascular anomalies of the upper face dermis (port-wine stain), and choroid and ipsilateral leptomeninges. The dermal lesion will always involve the V1 dermatome, but may extend to V2 and V3 as well. In many cases there will be hypertrophy of the underlying connective tissue and skeleton, giving the patient an asymmetrical, sometimes grotesque appearance that is progressive through puberty. Focal or generalized seizures are common presenting symptoms of neurological involvement. Some of these patients will have varying degrees of mental retardation. In rare circumstances there may be other neurological findings of hemiparesis, hemisensory disturbances, and growth disturbances of the contralateral extremities.

CT examinations will show intracranial calcifications, sometimes even in infancy. Arteriograms will show capillary and venous anomalies of the leptomeninges. Other
anomalous abnormalities may be seen and may be responsible for the progressive
degeneration and atrophy of the involved cerebral hemispheres seen in some patients.

Patients with port-wine stains of both V1 and V2 are at risk for the development of
glaucoma. Those infants suspected of having Sturge-Weber syndrome or port-wine stains
surrounding the orbit should have ophthalmological evaluations for glaucoma early, and then
on a regular basis at 6- to 12-month intervals. The glaucoma may be refractory to medical
therapy and require surgical treatment.

The history of treatments for port-wine stains is similar to the range of treatments for
hemangiomas, both being equally ineffective with potentially serious side effects. Scarification
by various methods has been one of the more frequently described treatments. Various
methods of cutting, puncturing with needles, electrocautery, ultraviolet light, various forms
of radioactive materials and radiation therapy, cryotherapy and even sandpaper, have been
described. If effective at all, these treatments traded scarring for the stain. Tattooing has been
used for many years, but the results are inconsistent, leaving a blotchy papular surface with
an unnatural, fixed appearance that does not blend well at the edges, and in children may
result in hypertrophic scarring.

One of the most acceptable forms of therapy without side effects is cosmetic
camouflage. (The two most popular products are Covermark and Dermablend, which may be
purchased at many large department store cosmetic counters.) It is a sophisticated blend of
opaque, waterproof cream that is covered by an appropriate shading cream. The biggest
disadvantage is the time it takes to apply (20 to 30 min daily), and the cost. Teenage girls and
adult women are more likely to make the commitment of time necessary to use this method.
I have had personal experience with one 10-year-old boy who seemed to be well adjusted to
his port-wine stain, but after his mother used some makeup to cover this he became much
more outgoing and self-assured.

Excision is another alternative for small areas or in facial aesthetic units. If primary
closure is possible, a linear scar in relaxed skin tension lines may be an excellent alternative.
If larger areas are involved, serial excision, skin expansion techniques, or full- or split-
thickness skin grafts may be used to resurface the area. The disadvantage of skin grafts are
their unpredictable pigmentation, scar hypertrophy at the edges, recurrence in the graft or at
its edges, and abnormal skin texture. For these reasons skin grafts are usually discouraged.
However, some patients are willing to trade these deficiencies for the bright red or purple
port-wine stain they have lived with all their lives.

Laser treatment for port-wine stains was first reported by Goldman and Rockwell in
1973. In the 1970s the argon laser became the laser of choice for treating port-wine stains.
It has the advantage of emitting light in the blue-green spectrum that is preferentially
absorbed by red hemoglobin and melanin and penetrates only 1.0 to 1.5 mm beneath the
surface of the skin. As the light energy penetrates the skin, it is changed to heat-producing
thrombosis with collapse of the vessels and damage to the dermal collagen. The adnexal
elements of the skin, sweat glands, and hair follicles survive, whereas there is superficial
necrosis of the epidermis and dermis. The end result is a controlled dermal scar with
epidermal regeneration from the adnexal elements and a decrease in the size and number of
blood vessels, which leaves the skin smoother and lighter in color.
Side effects of the argon laser treatment are atrophic and hypertrophic scarring. The former is an acceptable complication and to a certain degree is the expected result from the dermal scar produced. Hypertrophic scarring is much less acceptable as it creates a raised, irregular surface that is cosmetically unacceptable and may actually be physically uncomfortable from itching and irritation. It is more common in children under 12 years, and in all ages in the nasolabial fold and upper lip. Unfortunately, in children in whom physicians would like to use the argon laser, the results are poorest, with either little or no response or hypertrophic scarring.

The technique for the argon laser is to do a test patch 1.0 to 1.5 cm square at the edge of the lesion in the most inconspicuous spot or in an area that may be excised with a good cosmetic result if there is hypertrophic scarring. If there is a good result the lesion may be treated in stages. The area to be treated is anesthetized with 1% Xylocaine without epinephrine. One characteristic is that the anesthetic very rapidly absorbs, so that only an area that can be treated in 5 to 10 min is anesthetized at one time. Each session will last 1 to 1.5 hr, depending on the area to be treated. We have tried narrow stripes, wide stripes, and checkerboard patterns, but have ended up choosing to treat anatomic units, which seems to give the best cosmetic result. Reepithelialization comes from the remaining adnexal units, not from the edges of the treated areas. The problem with the geographic patterns was the difficulty of blending the egdes between the treatment areas. They tended to have hyperpigmentation at the edges that left a visible pattern result. Following treatment, the skin is covered with an antibiotic ointment daily until the surface reepithelializes, which usually takes 7 to 14 days. The patient should avoid direct sunlight as much as possible, and should use a good sunscreen for 6 to 8 months. The final result will be visible in 3 to 4 months, although further lightening may take place over the next 12 to 18 months. The expected result is a lightening of the color, and in the those with thickened or an irregular skin surface, a significant smoothing. Although many times the results are mediocre in the eyes of the surgeon, most patients are happy with any improvement, especially women who use cosmetics for camouflage. If there is significant lightening of the lesion they can use less makeup with a better result.

Other lasers have been used to treat port-wine stains. The CO₂ laser vaporizes the lesion including the epidermis, dermis, and ectatic vessels. This is a nonspecific burn and is essentially the same as using dermabrasion. Scarring is significant. In general, it is not a recommended treatment.

The most recent laser to be used is the tunable dye flashlamp laser. It is tuned at 576 nm, which should be the ideal wavelength for maximum absorption by the ectatic vessels. In clinical experience it seems to work better on children, patients with light port-wine stains, patients who might scar easily, and patients who have not responded well to the argon laser. It does seem to require more treatments than the argon laser, but if the overall results are better with less scarring and a better textured skin, it will be worth it.

Laser therapy seems to be the best treatment available for port-wine stains today, but newer lasers, especially the copper vapor laser, and refined techniques hold promise that our results will continue to improve.
Telangiectatic Stains

*Nevus flammeus neonatorum* is a very common entity that may be confused with the uncommon port-wine stain. It has several synonyms including stork bite, salmon patch, and angel's kiss. These lesions occur most commonly on the forehead or nape of the neck and are pink, irregular, macular spots that blanch with pressure. The majority fade during the first year of life, but some may persist into adolescence and even into adulthood. Rarely there may be a familial incidence.

There are several other syndromes that include capillary-lymphatic malformations, hyperkeratotic vascular stains, angiokeratomas, and congenital telangiectasis, each with their own characteristics. Again, for a more complete description of these entities one is referred to Mulliken and Young's text on vascular birthmarks.

A final telangiectatic condition is the unusual familial syndrome of Rendu-Osler-Weber or hereditary hemorrhagic telangiectasia. It is inherited in an autosomal dominant pattern with an incidence of 1 to 2 per 100,000. These lesions are characterized by discrete, bright red maculopapules, 1 to 4 mm in diameter, that appear on the mucosal surfaces of the nose, lips, and oral cavity, as well as on the face, palmar surfaces of the hands, nailbeds, and conjunctiva. They are also found on the mucosal surfaces of the gastrointestinal tract, urinary tract, tracheobronchial tree, vagina, and the parenchyma of the liver and central nervous system. The lesions may occur in childhood, but more commonly appear after puberty in the third and fourth decade. The condition is usually progressive with age and the patient's symptoms become more difficult to control. The vascular papules tend to ulcerate and bleed, leading to recurrent and difficult to control epistaxis, hemoptysis, hematuria, and melena. There is a family history in 50% to 70% of patients. However, many patients are unaware of the inherited syndrome, but just assume it was a family characteristic to have frequent nosebleeds or other bleeding problems. Severe complications may occur if the patient develops pulmonary or hepatic arteriovenous fistulae with high-output cardiac failure. Bleeding into the brain or spinal cord will produce neurological symptoms.

The most troublesome problem for most patients is epistaxis. It may occur spontaneously at socially awkward times such as during meals, with exercise, during sexual intercourse, and with emotional stress. As the patient becomes older and the bleeding becomes more severe, the patient may become incapacitated from anemia, cardiac failure, and the necessity for frequent hospitalizations for treatment and blood transfusions. Frequent blood transfusions, if necessary, increase the patient's risk to hepatitis and AIDS.

Treatment of the epistaxis starts with simple measures of pressure, superficial cauterization with silver nitrate or electrocautery, packing with Gelfoam and thrombin, or various hemostatic agents such as Avitene or Instat. One should avoid gauze packing as it produces more bleeding when it is removed. Care must be taken with overzealous cauterization as it may produce septal perforations. As these methods fail, one can progress to the use of the Nd:YAG laser, which is quite effective on the visible lesions, but as new ones appear it will need to be repeated. Systemic estrogen or estrogen creams have been recommended, but there is conflicting information as to their efficacy. If the lesions are surgically accessible through an alaotomy, one may remove the mucosa of the septum and floor of the nose, leaving perichondrium, and cover it with a split-thickness skin graft. The problem is that the lesions
may recur through the graft or at its margins, and there will be crusting and sometimes and odor associated with the skin graft.

Vascular spiders are small lesions with a central arteriole that has superficial vessels radiating from it. With pressure on the lesions using a glass slide, one may see central pulsations, and, as it is released, a centrifugal rush of blood to the edges. These lesions may appear on the face, arms, hands, and fingers. They occur in children and/or adults as well as temporarily during pregnancy. Because they appear during pregnancy and in patients with liver failure, there is evidence that these are hormonally modulated by estrogen. These lesions may be treated with the argon laser or a needlepoint cautery to destroy the central arteriole, although recurrences are not unusual.

Venous Malformations

Venous malformations are developmental abnormalities of veins that are dysmorphic in configuration and structure. At times they may be combined with capillary or lymphatic malformations. They may be further classified as localized, diffuse, deep, or superficial.

Venous malformations may present as skin or mucosal varicosities, spongy masses, or as diffuse lesions spreading through multiple tissue planes. If the lesion is deep, the overlying skin may be normal or have a bluish hue. More superficial lesions will have a deep purple color.

On a physical examination the lesions are soft and compressible, but nonpulsatible. After compression they refill slowly. Patients note that with straining or in a dependent position they will fill and this may be associated with a vague feeling of pressure or discomfort. They will grow in proportion with the child, but may enlarge rapidly following trauma or coincident with puberty or pregnancy. Occasionally, after direct trauma, they may develop arteriovenous shunting and develop into an arteriovenous fistula. In older lesions phleboliths from thrombosis may be palpable or visible on x-rays.

Histologically these lesions will have large venous spaces (formally referred to as cavernous hemangiomas) lined with flat, normal, epithelial cells and sometimes evidence of recent and old thrombi.

Venous malformations may involve muscles, especially the masseter muscle. If they are deep they may be mistaken for unilateral masseter muscle hypertrophy, even on a CT scan. MRI is particularly useful in separating out differences in these soft tissues and will help make the diagnosis. There is also a rare infiltrating intramuscular vascular lesion that has been called an intramuscular hemangioma or angiolipoma. It should not be confused with venous malformations.

Venous malformations of the facial bones are rare, but they may cause skeletal hypertrophy and bleeding around teeth. X-rays show a radiolucent defect with a honeycomb appearance.

Therapy for venous malformations has suffered from the same plethora of modalities as other vascular lesions. These again include irradiation, electrocoagulation, cryotherapy,
intravascular magnesium, and various sclerosants.

Before embarking on therapy, it is important to determine if the lesion is in the low-flow category or the high-flow. If there is any question, angiography should be carried out. The anatomical boundaries must be determined, either by MRI or angiography. One must determine if there is any localized intravascular coagulopathy that could lead to devastating bleeding at the time of resection. This must be evaluated as suggested previously with coagulation studies looking not only at the PT and PTT functions, but also studies for a consumptive coagulopathy.

If possible, total excision is the treatment of choice, but this is possible only in small to medium-sized lesions that are well demarcated. Unfortunately, surgery may be limited to partial resection or contour resection only. One of the frustrating points in surgical excision is that as the lesion is dissected and the blood supply is interrupted, the whole area collapses to a small amount of tissue and the edges are so ill-defined that it is impossible to tell if total excision has taken place.

Subtotal resection may lead to postoperative expansion of the remaining lesions, but it is worth the risk if there is significant improvement in contour, cosmetic abnormalities, or feelings of pressure and discomfort. In some cases removal of skeletal lesions may be indicated.

Selective angiography and embolization has been used, but runs the risk of necrosis of the adjacent soft tissue and overlying skin as reported by Demuth et al. Persky has reported a sclerosing technique for direct injection of contrast media and 95% ethyl alcohol into low-flow venous malformations with good results. Multiple treatments could be performed, and surgical resection of residuals was carried out in some cases.

The Nd:YAG laser has been used successfully for the treatment of mucosal malformations. In our own experience this method has been quite successful for pure venous malformations, especially if they are ectatic. The CO₂ laser has been more successful in treating the variegated combined venous-lymphatic malformations seen on the dorsum of the tongue.

Arteriovenous Malformations

Of all the vascular anomalies, arteriovenous malformations are the most feared and difficult to treat. Arteriovenous malformations are high-flow lesions that enlarge not by cellular hyperplasia but by hemodynamic mechanisms. Fortunately they are rare lesions in contrast to low-flow anomalies.

Cervicofacial arteriovenous anomalies may be noted soon after birth and grow proportionally with the child, but many will not be visible until years later when they become symptomatic. Rapid expansion may occur after local trauma, attempted excision or ligation, or with hormonal changes at puberty and during pregnancy.

The presenting complaint will frequently be a pulsatile tinnitus or buzzing heard best at night when it is quiet. There may be intermittent episodes of stabbing pain. In lesions
closer to the surface, sudden bleeding from a tooth socket or epistaxis may unmask an unsuspected lesion. On examination, the overlying skin may have a mild bluish-red hue with elevated temperature. There will be a palpable thrill with a bruit on auscultation. Congestive heart failure may be present if the arteriovenous fistula is large or of long standing. Arteriovenous shunting may diminish blood flow to adjacent areas and cause ischemic pain and/or necrosis of the skin. Recurrent severe episodes of bleeding may be life threatening. Some lesions may cause local bony destruction or involve the bone itself, producing radiolucent or multiloculated areas similar to low-flow anomalies. If these involve the mandible or maxilla, they may be manifested by a loose tooth with massive bleeding when it is extracted or bleeding around the tooth. As in other vascular anomalies, there is always the danger of disseminated intravascular clotting secondary to thrombotic consumption or local destruction of clotting factors.

The pathophysiology of arteriovenous fistula starts with a direct connection between the arterial and venous circulation. With direct flow from a high-pressure to a low-pressure system, two circuits develop, one with a high resistance through the capillary bed and one with low resistance directly from the artery to the vein. As more and more blood passes through the low resistance circuit, the arteries enlarge and become tortuous, whereas the veins undergo commensurate hypertrophy to accommodate the flow. As the fistula enlarges, the veins become pulsatile as there is direct blood flow from the artery unimpeded by the capillary bed. As this flow increases, blood pressure in the artery beyond the fistula decreases and eventually there is a reversal of flow as the collateral circulation develops, and this supplies the fistula as well. As this happens, blood may be "stolen" from the area supplied by the distal artery, producing ischemia of the area and possibly leading to necrosis. As the shunting increases in volume, the systemic blood pressure would drop, but this is compensated for by increasing blood volume and cardiac output. If shunting reaches a critical point, the patient may suffer from high-output cardiac failure.

The treatment of arteriovenous malformations is hazardous and the results are frequently disappointing. If the patient is asymptomatic, it is best to follow the patient closely. Intervention is indicated for life-threatening problems such as hemorrhage, pain, pressure ischemic ulcerations, or heart failure. Treatment consists of embolization, surgical excision, or a combination of both.

In these high-flow lesions angiography is imperative. Angiograms are difficult to interpret because of the rapid blood flow. Contrast media may be preferentially swept through proximal arteries and veins, failing to demonstrate more distal arteriovenous fistula and collateral shunting. The dye may be so diluted in large venous lakes at the center of the lesion that minor arterial feeders are hidden. It is important that the intracerebral circulation also be evaluated to assess shunting between the internal and external carotid circulation.

In the operative therapy for these lesions, it has been well shown by multiple authors that proximal ligation of the feeding vessels is not only useless, but will result in disaster, especially in the cervicofacial area where there is such a rich collateral circulation. The lesion may undergo a rapid increase in size as collaterals are picked up and develop from the contralateral as well as the ipsilateral circulation and the internal carotid system. Ligating proximal vessels precludes the use of embolization in the future.
Embolization would appear to have the same risks as proximal ligation, but the goal is to embolize the center of the lesion first and work outward. It may be a curative, palliative, or preoperative effort. Ideally, embolization should be a preoperative effort to decrease blood flow for the surgical resection, which should be planned for 48 to 72 hr later. If surgical resection is impossible because of the location and/or structures involved, embolization may be attempted as a curative procedure. The goal is to block as many shunts as possible, decreasing the size of the lesion and hoping that collateral circulation will be slow to develop. It is imperative that the venous lakes at the center of the lesion, especially if they are in bone, be obliterated as well, or they will act as sumps attracting new blood supply. If the embolization attempt is meant to be curative, we prefer to use vascular springs that are permanent and will not pass through into the distal circulation. In bony lesions, a direct stick into the venous lakes is possible, packing them with multiple springs. For preoperative efforts, absorbable materials such as Gelfoam are used. Palliative embolization may be used for relief of symptoms such as pain and bleeding, and when the lesion cannot be removed surgically.

As we have condemned proximal ligation of the arterial supply, the goal, if surgery is contemplated, should be en bloc resection as if for a tumor. Embolization will not reduce the amount of resection needed, but the goal is to make the blood loss manageable. Several other techniques may be used such as temporary mattress sutures around the periphery, temporary occlusion of feeding vessels with percutaneous transcatheter balloons or temporary clamps, or profound hypotensive anesthesia. Large lesions of the scalp, ear, and midface that have failed multiple therapies may require large resections of tissue with resurfacing by regional myocutaneous flaps or distant free flaps. Even with the best of intentions, these frightening lesions frequently recur. However, these patients may be helped for a significant length of time, and some will be cured.

**Lymphatic Malformations**

The lymphatic anomalies suffer from the same semantic problems as the other vascular anomalies that we have discussed. To be consistent with the terminology suggested by Mulliken and Young, as used throughout this chapter, all lymphatic anomalies would come under lymphatic malformations. This change in terminology, although technically correct, is difficult to use, since common usage of terms, such as lymphangioma and cystic hygroma are so entrenched in our medical literature and everyday language. Even recent articles that espouse the classification suggested by Mulliken and Young will revert to common usage terms, creating confusion as to the conditions they are describing and treatment they are suggesting. In the past, the term lymphangioma and cystic hygroma have been confusing and have been suggested by some to represent distinctly different entities. Today it is generally accepted that they are the same entity. For our purpose we will consider lymphatic malformations to be lymphatic anomalies consisting of spaces that may be unilocular, multilocular, or diffuse, lined with flattened epithelium, and containing a pale yellow fluid. There may be combined lesions with venous and lymphatic components, which are referred to as lymphatic-venous or venous-lymphatic malformations. As in venous malformations, these lesions do not undergo cellular proliferation as in a neoplasm. In general, they grow commensurate with the growth of the child, but may undergo rapid expansion during upper respiratory infections or with hemorrhage into the lesion.
It is generally accepted that lymphatic anomalies arise as a result of maldevelopment of the primitive anterior, posterior, or retroperitoneal lymph sacs. This could be a failure of these sacs to form a venous connection or some interruption of the drainage path of the lymphatics. The lack of egress of the lymphatic fluid produces large cysts or more compact anomalies if present in a restrictive tissue environment.

Lymphatic anomalies have an equal sex distribution. They are most common in the cervicofacial region, but also occur in the axilla, extremities, and trunk. These lesions occur at about the 6th to 9th week of gestation and may be detected as early as the 12th week by ultrasound. In a review article by Cohen et al, it was found that if a lymphatic malformation was detected in the antenatal period, 73% had elective termination of the pregnancy and 22% resulted in fetal death in utero. The most common location was in the posterior nuchal region. Some of those detected had spontaneously resolved by birth. There is a well-documented correlation between these cystic lymphatic lesions in the fetus and various chromosome abnormalities, especially Turner's syndrome. If this condition is found on ultrasound, amniocentesis for karyotyping is indicated.

The majority of lymphatic malformations are detected in the newborn nursery or during the first year of life, but Kennedy reported 45% of his cases were over age 20. These lesions may present as large multiloculated cystic structures to small cutaneous or mucosal blebs. The large cystic lesions may transluminate, and do not collapse with pressure as does a venous malformation. Large lesions may compress or distort the parapharyngeal airway and trachea, and extend into the mediastinum. Lesions may involve the orbit or periorbital tissue causing exophthalmus and muscular problems or visual disturbances similar to those seen in the hemangiomas of the orbital area.

Cellulitis of lymphatic malformations is a relatively common occurrence. The lesion will become tense, warm, and erythematous with rapid enlargement. This rapid enlargement may be life threatening if it intrudes on the airway by direct extension or extrinsic pressure, especially if the tongue or laryngeal structures are involved.

It is possible for the lymphatic abnormality to diffusely involve the tongue musculature, resulting in various degrees of macroglossia. Sometimes the mucosal surface of the tongue and/or cheek will be covered with small cystic or hemorrhagic vesicles that will intermittently bleed. Skeletal hypertrophy occurs with lymphatic malformations adjacent to the skeletal structures, especially of the mandible and maxilla.

Although growth in proportion to the child is characteristic, the question of spontaneous regression or improvement has not been answered. There are sporadic reports of spontaneous involution that is slowly progressive over a period of 2 to 6 years.

The treatment of lymphatic malformations is even more controversial than the other vascular anomalies. History is filled with the usual plethora of attempted modalities that were tried in other vascular lesions. These include a multitude of sclerosing agents, cautery, and irradiation. Because of the wide variety of presentations, to do nothing may be the least harmful. In general, the main therapeutic thrust is a well-planned surgical excision.
Two conditions that require emergent intervention are airway obstruction and infection. Airway obstruction is usually secondary to tongue involvement with the tongue being either infiltrated or pushed up and posterior by cystic lesions in the floor of the mouth. Because of the tongue position, these infants may be very difficult to anesthetize and intubate without losing control of the airway. Aspiration or incision of the cysts in the floor of the mouth may give temporary relief, but usually they are so multiloculated that significant decrease in size is not obtained. If there is significant airway obstruction present, a tracheotomy must be performed, which has its own unique risks for mortality. Some lesions may become so invasive that the naso-oro-hypopharyngeal areas will become partially obstructed. If oral alimentation is compromised, a gastrostomy may be necessary to prevent a failure-to-thrive condition.

Infection is the second complication requiring emergent treatment. In some cases a viral upper respiratory infection may cause rapid enlargement, but without cellulitis. If cellulitis occurs, it is usually from oropharyngeal organisms and should be treated as such. Penicillin should cover most oral organisms, but in children there may be a significant number of infections with staphylococci and hemophilus. In the acute stages, the child may be septicemic with a high fever, as well as with the localized cellulitis. In this situation the child is usually quite sick and should be hospitalized for blood cultures, aspiration and culture of the cyst fluid, and intravenous antibiotics. With the rapid changes in antibiotics, a specific recommendation may soon be out of date, and it would be best to consult with a pediatric infectious disease specialist for the best antibiotic to cover the most common organisms. At present we tend to use one of the cephalosporins or clindamycin. Once infected, these patients seem to be more susceptible to repeated infections. In this situation if there is just cellulitis without systemic signs, a prescription for oral antibiotics may be sufficient, to be used when early symptoms appear. If infections become frequent, one may be able to prevent recurrences with the use of prophylactic antibiotics such as those used for recurrent otitis media and sinusitis.

Surgical excision is the treatment of choice in symptomatic lymphatic malformations. Surgical excision should be preceded by good planning. In large lesions, radiological evaluation with either CT or MRI should be used to determine the extent of the lesion. They can be evaluated quite well with either or both modalities, giving the surgeon the perspective of what vital structures are at risk and if there is extension into the mediastinum. If lesions are relatively small, circumscribed, and easily outlined on palpation, x-rays are not needed.

Timing is controversial. If lesions are small and not causing complications, a wait-and-see approach is indicated. This is particularly true for parotid lesions, where a differential diagnosis between a hemangioma that will involute and a lymphatic malformation may be difficult without invasive studies. Since there are sporadic reports of regression, this hope may be held out to the parents, but with the warning that this outcome is unusual. Allowing time to pass and the child to grow may make surgical excision safer for the child in terms of fluid and blood loss, as well as easier in that vital structures such as nerves and blood vessels will be larger.

In large cystic lesions that are rapidly enlarging with the potential for causing complications, especially airway compromise, the surgeon's hand may be forced even in the neonate or first few months of life. These are usually cystic lesions involving the anterior
cervical and submaxillary triangles, but may also extend into the posterior cervical triangle. The rationale for approaching these lesions early is to prevent the necessity for a tracheotomy and/or gastrostomy. If the lesion is bilateral, staged excision may be best. The surgeon must be prepared for a prolonged, tedious dissection. It should not be added to a long surgical list at the end of the day, or one will be tempted to compromise the dissection. These lesions may be approached from a variety of incisions, depending on the extent and location of the lesion. Many times a horizontal midneck incision will suffice to remove lesions from the mandible to the clavicle, especially if extended across the midline or more posterior. If better exposure is needed, a superior and/or inferior limb of a half H may be used. The superior limb of the half H may be extended upward in the preauricular area to complete a parotid incision, if a parotidectomy becomes necessary. A classical McFee or inferior and superior horizontal incisions are also very cosmetic. Care must be taken to find and preserve vessels and nerves. Basically, the technique is similar to a conservation lymphadenectomy (Bocca technique) where all anatomic structures are preserved. One only needs to review the literature to see the high rate of complications such as facial nerve paralysis, Horner's syndrome, and paralysis of cranial nerves X, XI, or XII to see why people would recommend no surgery. The ramus mandibularis must be found and preserved, as well as the greater auricular nerve, sympathetic trunk, VII, X, XI, and XII cranial nerves, phrenic nerve, brachial plexus, and cervical nerves. The sternocleidomastoid muscle and internal jugular vein may usually be preserved and obviously the carotid artery must be preserved. As the dissection is carried out, the cysts may be accidentally ruptured or may have to be transected to preserve vital nerve or vascular structures. Although the goal is complete resection, this is frequently impossible. An attempt is made to dissect as much of the shiny cyst wall as possible, even after rupture has taken place. If the lesion extends into the floor of the mouth or substance of the tongue, an attempt is made to dissect these cysts as completely as possible, and where not possible, to at least open them. I have found it helpful in some of these deep cysts to lightly fulgurate the lining with the cautery.

Parotid involvement combined with a cervical lesion or as an isolated lesion needs special attention. Usually the entire parotid is involved, although the majority of the gland (80%) is lateral and inferior to the facial nerve. If one is forced to treat an infant or very young child, the facial nerve may be very small. Microscopic dissection or loupe magnification should be used to reduce risk to this vitally important nerve. Tedious, meticulous dissection is the watchword. If the physician does not have the experience or patience to do this, the patient should be referred. The physician must do no harm! Cysts in the deep lobe are dissected as best as possible without injuring the nerve, or are opened with a hemostat.

When the resection is completed, there is the question of how to drain the surgical site. In general, I prefer to use suction drains. One must be careful to have controlled any open vascular channels associated with the lesion. Upon occasion it seems that there is a localized coagulopathy condition leading to either significant bloody drainage and/or a hematoma. In those situations, reinspecting the wound and penrose drains may be necessary. One must account for and replace fluid loss, especially blood and protein, if there are large amounts of drainage in a small infant. Drainage may be necessary for several days to prevent a postoperative seroma or reaccumulation of lymphatic fluid. If there is a relatively rapid recurrence of cystic structures at the edge of a resection, reoperation is indicated. If the child has had cellulitis prior to surgery, prophylactic antibiotics should be used.
In those patients who have large lesions that have been observed without noticeable regression, surgical excision is indicated at age 1 to 5 years, depending upon its size and location. The same surgical principles as previously described should be used.

In patients with significant macroglossia demonstrated by extension outside the oral cavity with mandibular and/or maxillary distortion, a reduction glossectomy is necessary. This may be done with vertical and/or transverse wedge resections. One must be careful to preserve nerve function and enough muscular bulk to avoid interference with deglutition and speech. Mandibular or maxillary distortion may require orthognathic and/or orthodontic procedures.

Other treatment modalities may be used as adjunctive procedures. The CO₂ laser is useful in vaporizing lymphatic involvement of the larynx and hypopharyngeal structures. The Nd:YAG laser may be used on combined venous-lymphatic malformations, but does not work as well in pure lymphatic lesions where there is no pigment to absorb the laser rays unless high power-density ratios are used. Vaporization of the lymphatic lesions of the tongue and oral mucosa is better accomplished with the CO₂ laser.

There are sporadic reports using chemotherapeutic agents in the treatment of lymphatic malformations. Ogita et al have used a sclerosing agent OK-432 for direct injection into the lesion. In nine patients treated, eight had complete resolution of the lesion. Cyclophosphamide has also been reported as having a good response in two cases.

**Summary**

In summary, vascular anomalies are difficult lesions to treat. However, if careful evaluation is carried out and an appropriate diagnosis is made, the appropriate therapy may be chosen.
Pediatric Facial Plastic and Reconstructive Surgery
James D Smith, Robert M Bumsted

Chapter 6: Otoplasty
Wayne F Larrabee, Douglas Kibblewhite, and Jeanne B Adams

History

Otoplasty is surgery to correct protruding ears. Ely first wrote about otoplasty in 1881. In the *Archives of Otology* Ely described a 12-year-old boy who came to the Manhattan Eye and Ear Hospital with a complaint that his companions ridiculed him because of the prominence of his ears. Ely, through a postauricular incision, excised skin and cartilage to correct the protruding ear. In 1889, Keen of the Jefferson Medical College noted Ely's contribution but described a better approach to the deformity. Ely had advocated excising skin on the anterior as well as the posterior surface, which left an anterior scar. Keen described an operation solely removing posterior skin and cartilage and noted that cartilage as well as skin must be removed to obtain an adequate result. Luckett in 1910 noted the importance of the antihelical fold. Writing in *Surgery of Gynecology and Obstetrics*, this New York surgeon described again the common problem of a child being teased for his "donkey ears" and analyzed the anatomy of this deformity. He noted lack of an antihelix in these children and described a procedure to create such an antihelix by excising a crescent-shaped piece of cartilage and closing the defect with buried mattress sutures. In addition to delineating the anatomical deformity, he very perceptively noted:

> In an ear with a very thin flexible cartilage, I think it would be possible to reconstruct the antihelix and set the helix close to the head without excising a segment or even incising the cartilage, simply by fluting or folding the cartilage at the proper site, and passing the suture in such a manner as to maintain the fold.

Morestin in 1903 and Owen in 1965 described a technique of placing buried sutures to produce a controlled change in auricular shape. Mustarde, however, writing in the *British Journal of Plastic Surgery* in 1963 refined and popularized this method, which is now commonly attributed to him. Gibson and Davis' studies of the influence of incisions on the deformation of rib cartilage, and, more recently, Murakami's analysis of pig ear cartilage bending created by surgical incisions, have influenced the application of cartilage scoring techniques to otoplasty by Stenstrom and others. These authors have demonstrated that incisions made through perichondrium and into cartilage cause the area incised to become more convex.

Anatomy

Ears may be described in terms of their form, their proportions, and their position on the skull. Small discrepancies in these variables are immediately obvious to the observer, but ears do vary widely between individuals. The framework of the ear is a single, complex piece of fibrocartilage to which thin, translucent, well-vascularized skin is tightly adherent,
more so anteriorly than on the posterior surface. With increasing age, the cartilage becomes stiffer and less pliable, and the overlying skin becomes less tightly attached.

The helix is a gently curved incomplete oval, broader at its root or spine at the external auditory canal and thinning as it curls down toward the lobule. It is partially concave at its spine, becomes almost tubular in its mid-portion, and flattens out again at the lobule. The antihelix originates at the antitragus and follows the curve of the helix superiorly. Approximately two-thirds of the vertical distance toward the top of the ear, it divides into a sharp fold, the inferior crus of the antihelix, which proceeds anteriorly to dive under the overhang of the spine of the helix, and a less prominent broader fold, the superior crus of the antihelix, which fades out into the area under the superiormost part of the helix. The area between these two crura is the fossa triangularis. Posterior to the superior crus is the scapha, a narrow shelf between helix and antihelix. Medial to the antihelix is the kidney bean-shaped conchal bowkl, divided by the spine of the helix into a superior portion, the cymba conchae, and a somewhat larger inferior portion, the cavum conchae. The anterior portion of the cavum conchae is the external auditory meatus. The tragus is a firm, thick plate of cartilage partially covering the opening to the ear canal, and is continuous posteriorly with the variably developed intertragal notch and antitragus, which itself blends into the antihelix. Between the tragus and the spine of the helix the ear cartilage is discontinuous. This incisura terminalis is useful for an endaural approach to the ear canal, which avoids cutting cartilage. The final part of the auricle to consider is the lobule. This dependent fibrofatty tissue lacks cartilaginous support and hangs from the framework of the ear. It is variable in size and shape and in its attachment to the skin of the cheek.

Several points can be defined on the ear as a whole to allow quantification of the internal proportions of the ear and its position on the skull. The most superior portion of the ear at the top of the curve of the helix is the supraaurale, and the inferiormost part at the tip of the lobule is the subaurale. Similarly, the most anterior and posterior points on the helix are termed the preaurale and postaurale. The attachment of the ear to the skull is called the otobasion, which is a line lying anterior to the tragus. Its superior extent where the helix blends into the skin superior to the zygomatic arch is the otobasion superiorus, and the inferior attachment of the base of the tragus to the cheek skin is the otobasion inferiorus.

The aesthetic proportions of the ear are difficult to describe, although disproportion is immediately evident to the casual observer. Generally, the width of the ear, defined as its widest part, should be approximately 55% of its length. The vertical midpoint of the ear is located at the root of the spine of the helix. One-third of the ear lies above the inferior crus of the antihelix. A line drawn through supraaurale to subaurale will make an angle of 15° to 30° to the vertical, and about 22° is ideal. This posterior angulation of the ear has been said to parallel the nasal dorsum; however, the nasal dorsum demonstrates a great deal of individual diversity itself. The otobasion lies at an angle of 8° to the vertical.

The ear's location on the skull is important. In the vertical plane, the top of the ear lies at brow level, whereas the subaurale lies at the level of the columella. In the anterior-posterior direction, the ear should lie one ear-length from the lateral orbital rim (or, about 6.5-7.5 cm). Finally, one must consider the projection of the ear from the mastoid plane. Although this has been described in terms of the angle the ear makes with this plane, it is generally easier to
measure the distance from the anterolateral portion of the helix. This distance should be between 15 and 20 mm.

Thus the ear as a whole should be positioned correctly on the skull in the superior-inferior and in the anterior-posterior places, and its angle of inclination should be correct. Finally, its protrusion from the skull should be neither too much (bat ears) or too little (pinned-back appearance). It is difficult to relate ear position to bony landmarks, but with these specifications as a rough guide, the eye of the surgeon as observer should be able to make the fine adjustments to produce an aesthetic placement.

**Embryology**

The precursors to the human ear appear in the 6th week of fetal life in the shape of six hillocks of condensed ectoderm and mesoderm surrounding the first branchial groove. Mesenchyma of the first and second branchial arches thus creates the external ear. About 85% of the external ear develops in the second branchial arch and its derivatives. The first arch contributes only to the anterior portion of the ear including the tragus, the helical tragal sulcues, and the crus of the helix. The conchal cartilage and intertragal notch develop from the mesoderm along the first branchial groove and do not derive directly from the auricular hillocks. The auricle develops in the region of the neck and later migrates to the side of the head. This accounts for some of the congenital anomalies where the auricle is significantly displaced inferiorly. In the 8th to 12th weeks of gestation the helix develops rapidly and therefore, projects forward and overlies the still underdeveloped antihelix. At this stage, it is normal for the ear to protrude; however, if in the succeeding 12th to 16th weeks the antihelix fails to unfold, the helix continues to overhang and the protrusion will persist. At birth, the normal ear is usually completely formed and rests at an angle of 30° from the head. The ear continues to grow following birth and attains approximately 85% of its adult size by 3 years of age. Slow growth may continue until approximately 6 years of age, at which time the ear has reached the average adult size of approximately 6x3 cm. The distance from the mastoid peristeum to the helical rim changes little after 10 years of age, although the ear may continue to elongate more than 1.5 cm along its vertical axis during the lifetime of the individual.

**Surgical Concepts and Approaches**

There is a plethora of deformities associated with the auricle, and any surgical approach must be quite individualized. There are, however, some common factors that need to be addressed in many otoplasties. The three most common problem areas in auricular deformities are (a) the lack of an antihelical fold, or the “unrolled” ear; (b) a deep conchal bowl or, perhaps more correctly, a high conchal wall; and (c) deformities, usually protrusion, of the lobule. There three problem areas are relatively distinct and can be addressed individually with a variety of surgical techniques. In this chapter we will describe some of these basic approaches and then present the approach of two surgeons to otoplasty to give an example of how these techniques can be combined.

There are three basic approaches to remodeling the auricular cartilage into a more desirable shape. The first of these is a suture technique that uses a permanent suture to remold and hold the pliable auricular cartilage in its desired shape. The various suture techniques are
most useful in children and young adults whose cartilage is quite pliable. They can give quite natural results, but are not always effective in individuals with more rigid cartilage. The best example of this technique is Mustarde's technique of otoplasty, which depends on the placement of permanent buried sutures both to create a new antihelical fold and to maintain the new shape of the ear. An antihelical fold is created by the surgeon and maintained with a bayonet forceps. Three to four points corresponding to the proposed position of mattress sutures are then marked with methylene blue. These marks are then tattooed into the ear cartilage with a needle and methylene blue. The actual surgical approach is posterior. An ellipse of skin is excised and the posterior auricular cartilage meticulously cleaned of all fibrous tissue, retaining the perichondrium. The mattress sutures are inserted through the full thickness of cartilage and perichondrium using the dye marks for guidance and then tightened to create a fold.

The problem of a prominent conchal bowl can also be addressed with a suture technique. This approach has been popularized by Furnas with his conchal mastoid sutures, which he uses to approximate the conchal cartilage to the mastoid periosteum and thus flatten the excessively deep conchal bowl against the mastoid cortex. Objections to the use of permanent, buried sutures include infection due to the presence of a foreign body, palpable or visible sutures, and ridging of the skin where the sutures cross the posterior sulcus.

The second basic approach to remolding cartilage consists of full-thickness incisions or excisions of cartilage to correct the prominent ear. This is currently used most commonly in the concha through either an anterior or posterior approach. One can remove some aspect of the high conchal wall to allow the auricle to settle back to a more normal position. These cartilaginous defects can then be sutured to obtain the desired result. This type of excision allows one to reduce the size of the prominent concha to change the position of the auricle. In general, when full-thickness incisions or excisions are made to create an antihelical fold, one runs the risk of creating sharp edges and ridges. Most surgeons prefer to use the full-thickness cartilage incision or excision only in the concha.

The third basic approach to cartilage remodeling in otoplasty depends on the tendency of cartilage to bend toward intact perichondrium, and to bend away from a surface where the cartilage has been scored. Through a variety of approaches, both anterior and posterior, the cartilage is either abraded with light sandpaper, forceps teeth, dermabrader burrs, or special fine-toothed instruments designed specifically for this purpose in the area where the contour effect is difficult, leading to the occasional full-thickness incision with palpable sharp edges; thus, fine abrasion is preferred. To create a new antihelical fold, the surgeon will gain access to the anterolateral cartilage surface either through an anterior or posterior skin incision and lightly abrade the perichondrium and cartilage at the point where the new fold is desired. A major proponent of this approach has been Stenstrom, who has extensive clinical experience to show that abrasion with a rasp of the anterior antihelical area will cause furrowing and convexity to develop in this area and, thus, create an antihelix.

There can be multiple deformities of the lobule. The most common is a protuberance of the lobule, which can be addressed by either excising a small wedge of postauricular skin or doing some type of procedure on the cauda helicis. Some advocate excising the cauda helicis to produce a flat lobule. Excision can, however, result in protuberant skin with no structure to pull it medially. A cutting and repositioning of the cauda helicis or a simple
suture placement to move it medially works better in most cases. A lobule that is too large and dependent can be treated by excising a full-thickness wedge in the preauricular crease. Similarly, a very large lobule can be reduced by simply excising a crescent-shaped piece of skin and subcutaneous adipose tissue just medial to the rim of the lobule.

**Newborn Ear Taping**

A variety of auricular deformities can occur prenatally either from abnormal development or malpositioning of the fetus in the utero. A large prospective study in Japan has shown that approximately 50% of these will spontaneously correct themselves in the first month of life. The process is actually more complex with about 45% of ears normal at birth and 84% normal at 1 year. Lop ears, for example, are present in 38% of babies at birth and only 6% at 1 year, whereas the incidence of protruding ears actually increases from 0.4% at birth to 5.5% at 1 year. The protruding ear may be caused by mechanical factors in the first year of life. Although many ears that are abnormal at birth will return to normal by 1 year, there is no way to predict which ones will. Thus, the simple nonsurgical correction described below is applicable in every case.

Recent reports have demonstrated that deformities involving malposition of a formed auricle can be corrected nonsurgically by splinting the ear in the correct position within a few days of birth. The new position is maintained for the first 2 weeks. On removal of the splint, the ear remains in its new position. The physiologic explanation for the ability of newborn auricular cartilage to be permanently remodeled in the immediate newborn period is unclear. Cartilage is an avascular network of chondrocytes embedded in a type II collagen-proteoglycan matrix. The proteoglycan matrix is stabilized by hyaluronic acid, a single polymer of which may be linked with as many as 100 proteoglycan monomers. The very high estrogen levels that exist in the neonate at birth and for the first few days of life directly increase hyaluronidic acid synthetase levels. The enzyme affects the production of hyaluronidic acid, possibly allowing remodeling of the cartilage in the new position. Relaxin, a polypeptide hormone involved in parturition, also demonstrates very high serum levels in the immediate pre- and postpartum period. Relaxin has a profound effect on the fibroblasts and collagen of the pubic ligament, and has been recently shown to have a demonstrable effect on skin during tissue expansion. This effect is thought to occur through a disruption of the cross-linking of collagen in the extracellular matrix. High relaxin levels might also allow the auricle to remodel in the immediate postpartum period. Clinical experience shows that, whatever the underlying cause, the splint must be applied in the first few days after birth to be effective.

**Two Individualized Approaches to Otoplasty**

Multiple techniques are available to address the anatomical problems seen in the deformed ear. Surgeons tend to select those techniques that work best in their hands. To obtain a practical approach to otoplasty, two individual surgeons' approaches will be discussed. Obviously, each surgeon's technique is an amalgamation of the individual creative contributions of the pioneers in this field.

In our practice (WFL) a careful analysis of the ear is first performed to describe the specific anatomical deformity that needs correction. In general, the ear or ears involved have
some combination of a prominent concha, a lack of antihelix, and a malposition of the lobule. The ear with a prominent concha only can be treated quite easily with the conchal setback or conchal reduction procedure, which is described below. The ear with solely a lack of an antihelix is treated with a Mustarde suture technique only. For the majority of ears there are a combination of these two problems and both techniques are utilized. Problems with the lobule are addressed independently. In the physical examination cartilage pliability as well as symmetry is noted. Any specific deformity in the area of the lobule is described. If there is a tendency toward a "telephone ear" where the midportion of the ear is closer to the head than the superior and inferior portions that is noted. Standard photographs are taken. These generally involve frontal, posterior, and lateral views. Measurements of the distance from the mastoid skin to the helix are made on both sides.

The procedure can be performed under either general or local anesthesia. Children are best done under general anesthesia and adults can usually be managed quite nicely with local. Children are not operated on until age 5 to 6. A subcutaneous field block is all that is required for the anesthesia. Careful aseptic technique is used throughout the procedure. The patient is usually given prophylactic antibiotics due to the incision of cartilage and the permanent sutures placed. A stockinette dressing is placed over the entire head with openings created for the ears and face. A plastic eye sheet drape is used to maintain sterility of the operative field. If a conchal prominence is present, one first manually places the ear in an appropriate position and places a few dots in the postauricular sulcus to define where the new sulcus should be located. These dots generate a small wedge of tissue for excision. It is important not to overexcise skin in this area and not to create too shallow a postauricular sulcus. Incisions are then made perpendicular to the perichondrium of the posterior aspect of the auricular concha, and all intervening soft tissue is removed. In children it is important to use care because of the more superficial location of the facial nerve. Simply removing the excess soft tissue allows the ear to lie in a more natural position. The depth of the soft tissue is frequently underestimated and as can be seen, it is considerable. Simply removing this soft tissue in many cases will provide adequate setback. Once the auricle is in position against the mastoid peristeme, the surgeon can then remove cartilage as needed from the concha to create the desired ear position. By using a #11 blade, one can carefully shave small segments of the apex of the concha (actually the eminentia) until the ear is appropriately placed. By shaving parallel to the ear with the finger on the anterior surface, an automatic contouring is performed so that no sharp edges are visible. This part of the operation is quite straightforward but must be individualized with variable amounts excised from the different prominences, depending on the patient. The eminentia cymba concha, eminentia cavum concha, and eminentia fossa triangularis are individually treated to achieve symmetry and appropriate position of the ears. Once the appropriate amount of setback has been achieved, attention can be turned to the antihelix. With the Mustarde suture technique, usually three horizontal mattress sutures are used to define the antihelix. Mustarde originally described the technique using white silk sutures. We prefer 4-0 Mersilene, as it is less reactive. It also has good holding characteristics when the knots are snugged individually. Three sutures are usually placed to create the antihelix; more or fewer may be required in a given case. To facilitate suture placement, it is sometimes helpful to fold the antihelix with the forceps into the desired position and then to mark the proposed suture sites externally on the skin. Using a 20-gauge needle with methylene blue dye, one can perforate through these holes and mark the underlying cartilage for the suture site. From the postauricular incision one can then carefully dissect the skin flap up to the helix and visually see the dye marks where the suture.
should be placed. It is crucial to remove all soft tissue attachments from the perichondrium to allow remodeling of the fold. Sutures are best placed from the postauricular approach with one finger on the anterior aspect of the ear to ensure proper level of the needle. The needle should pass through the posterior perichondrium, cartilage, and the anterior perichondrium (but obviously not through anterior skin), and then back in the same fashion. It is quite important to pass through the anterior perichondrium or the suture may tear through and not maintain its position. After a single horizontal mattress suture is placed and tied with a surgeon's knot, it is snugged and visually examined to see if it is creating the fold of the desired position and contour. This individual suture is then loosened and the other sutures placed. If the sutures are tied after each individual placement, it becomes quite difficult to place the sequential sutures. Once all of the sutures have been placed, they are sequentially tied from superior to inferior while the ear is held in its new position externally. Care is taken not to include any unneeded soft tissue in the sutures themselves. The contour of the antihelix is then carefully inspected and if there are any asymmetries or problems the sutures are replaced at this time.

The majority of the problems with the antihelix can be quite nicely handled by the simple Mustarde technique. There are, however, times when the cartilage is thickened and will not easily conform with a suture technique. We are prepared at the time of surgery in these cases to proceed with an abrasion of the anterior surface of the antihelix similar to that described by Stenstrom. Stenstrom described the mechanical tendency of the ear to curl away from the area where perichondrium is excised. Thus, by using a small rasp to abrade the perichondrium and thin the cartilage slightly on the anterior part of the antihelix, one can facilitate bending with the Mustarde sutures if the cartilage appears somewhat intransient.

The ear is then set back into its new position against the mastoid cortex and three conchal mastoid sutures passed from the mastoid periosteum to the perichondrium of the auricle. It is a technical error to excise too much of the conchal bowl when doing a combined conchal and antihelical procedure because it may leave to little conchal bowl to obtain adequate purchase for the Mustarde suture.

After the ear has been placed into its new position the lobule is evaluated. If the lobule is slightly prominent, it can usually be corrected by excising a wedge of skin posteriorly. If this is not adequate or if the prominence is quite significant, a suture is placed from the cauda helicis to the more medial soft tissue to pull the cauda helicis and thus the lobule medially. Although the cauda helicis can be excised in these cases, it sometimes leaves the lobule still protuberant with no firm structure with which to mobilize it. Other problems of the lobule such as excessive lobule size are then addressed at this point in the operation.

After obtaining hemostasis with the bipolar cautery, a rubber-band drain is placed in the postauricular area and the incision meticulously closed using a subcuticular 5-0 nylon or Prolene. The subcuticular suture is left long, so that it can be pulled out at a later date without pulling the ear forward. After the second ear is recontoured, careful examination is performed to ensure symmetry between the two ears and any corrections are made as needed. Although it is not necessary to significantly overcorrect the ear, there is usually some minor overcorrection initially as the ear does tend to uncurl slightly from the scalp over the first few weeks after surgery. A dressing is then placed over the ear. Mustarde initially used wet lamb's wool, which when dry would form a stiff cast for the ear. We usually pack the ear with
bacitracin-impregnated cotton. This is packed anatomically with a deeper piece of cotton in the conchal bowl and a more superficial roll lying between the antihelix and the helix. A final, single piece of cotton is placed over this. A mastoid dressing is then placed over both ears. Though many surgeons leave these dressings in place for 4 or 5 days or more, we routinely change them on the first day to inspect the ears for hematomas or other problems and then replace the dressing. The second mastoid dressing is left in place for about 5 days. The suture is removed at 7 to 10 days by simply pulling out the subcuticular nylon. We ask the patient to wear a simple headband at night for another 4 to 6 weeks postoperatively to ensure the area is not accidentally damaged during sleep.

This combination of techniques - the conchal setback and/or reduction, Mustarde suture technique, and occasional rasping of the anterior antihelix - is adequate for the vast majority of cosmetic auricular procedures. The Mustarde technique is especially good in younger children where the ear cartilage is pliable and easily molded. It is more difficult in older patients. We have had very occasional episodes where the ear has partially unfurled at some stage after the operation, usually months or years later. It is relatively straightforward to replace the suture in those cases. Conservatism is essential with the conchal setback technique to avoid the all too common "telephone ear" appearance with the midportion of the ear being further back against the head than the superior and inferior poles. A major advantage of the technique described is that individual minor variations in the antihelix, concha, or lobule can be addressed without difficulty at the time of surgery.

The approach of Feuerstein and Adams is somewhat different and demonstrates that various approaches can be used to achieve similar results in otoplasty surgery.

The anesthesia employed depends on the age of the patient. Most parents or guardians of children under 10 years of age request general anesthesia, whereas older children and adults may comfortably be operated on under local anesthesia. The periauricular hair is not cut. Following surgical preparation of the skin, towels are sutured around the ear. An auricular-mastoid pedicle flap is then created, but no skin is removed initially. Skin excision should be reserved for the conclusion of the procedure so that careful tailoring of the postauricular skin can help prevent formation of the "telephone ear". That is, one may wish to leave a wider strip of postauricular skin in the middle third, and this need may not be appreciated until the very end of the procedure.

Next, one attempts to create two auricular "compartments". That is, an incision should be made along the margin between the concha and the antihelix, starting at the mastoid-conchal area inferiorly and continuing upward, carefully following the conchal curvature just medial to the antihelix into the inferior crus and then along the lower margin of the inferior crus to the level of the crus helicis. At this point the helical rim should be separated from the inferior crus by excision of a segment of cartilage. One effects complete severance of the concha from the antihelix so that the antihelix with its scaphoid portion and superior crus will easily fold and decrease the conchal-scaphal angle to a normal 90°. Because an adequate fold of the superior crus may still not be attained, further manipulation may be necessary in the superior crus and body of the antihelix. The use of a rotating wire brush to thin the cartilage may be useful.
Normally this crus has a gentle curve, which blends with the inferior crus as it reaches the level of the concha. One should not allow the cartilage incisions in this region to extend through to the perichondrium on the lateral surface. Such care will prevent formation of "sharp" lines, telltale evidence of otoplastic surgery.

Following complete mobilization of the conchal and antihelical auricular components, one must decide whether buried sutures of the Mustarde type are required for further correction. It is recommended that such sutures be used only to hold the repositioned cartilage in place until sufficient scar tissue has formed. Figures demonstrate the insertion of two types of sutures. The first demonstrates the conchal mastoid suture, which is indicated when a conchal setback is required; this suture passes from the conchal cartilage into the mastoid periosteum. The second suture demonstrates the vertical mattress Mustarde suture placed along the antihelix and its superior crus. As mentioned previously, it may be preferable to utilize a strong absorbable suture to obviate future "spitting" of sutures, granuloma formation, and secondary infection.

On completion of the otoplasty procedure, one should carefully evaluate the frontal appearance of the auricle to determine whether there is excessive residual protrusion of the antitragus or lobule. Antitragal prominence can readily be reduced by excision of a wedge of cartilage from the apex through the postauricular wound. The lobule may be accurately tailored to balance with the remainder of the ear by appropriate resection of skin wedges on the posterior aspect of the lobule. Actually the initial postauricular incision may be extended into the lobule. During surgery all bleeding vessels should be carefully cauterized. Use of fine bipolar cautery prevents excessive damage to the cartilage. In addition, a small Penrose drain inserted through the inferior aspect of the postauricular incision should be used routinely. The drain may readily be removed in 24 hr.

Interrupted silk sutures are recommended. The use of absorbable catgut for skin closure, although it obviates the necessity for removing sutures, is not suggested because of the increased incidence of keloid formation. Nonabsorbable sutures, such as silk or nylon, should be used. They may be removed in 8 to 10 days. A soft cotton and gauze dressing moistened with warm saline is used to maintain proper mobilization of auricular cartilage during the postoperative period. The dressing should routinely be changed in 24 hr and the auricular skin examined for its color and any evidence of hematoma formation. If, indeed, a hematoma has formed, the wound must be opened and the hematoma evacuated.

There are multiple other individualized approaches to otoplasty surgery but these two show the diversity possible and demonstrate the basic concepts. Readers are referred to the original works of those who have created these techniques, particularly Mustarde, Stenstrom, Furnas, Elliott, Davis, and Becker.

**Otoplasty Complications**

Otoplasty complications may be divided into postsurgical complications, and into a category of the unfavorable cosmetic results.
Surgical Complications

Infection

Postoperative wound infections in otoplasty are uncommon, and occur on the 3rd to 5th postoperative day as in other soft tissue infections. They are usually the sequel to an unrecognized or inadequately treated hematoma. The most significant symptom is pain, occurring as early as 12 hr postoperatively. Erythema, swelling, and purulent drainage may be noted on removal of the dressing. Removal of skin sutures should be considered to provide adequate drainage for necrotic debris, if present, and antibiotic therapy instituted on the basis of culture and sensitivity. Hospitalization for severe or progressive symptoms may be warranted. Delay in treatment or a rapidly progressive infection may lead to chondritis and deformity. Infections are relatively uncommon, probably because the auricle, like the face, is a relatively privileged area surgically with its rich blood supply and extensive anastomotic network.

Chondritis

The most significant sequela of infection is chondritis. Once started, it is difficult to eradicate, probably because cartilage is avascular and antibiotic therapy cannot penetrate to the locus of infection. Sequestra of dead infected cartilage form, leading to the necessity for potentially deforming debridements, and prolonged antibiotic treatment based on culture and sensitivities. Some progress has been made in the local delivery of antibiotics to problem areas such as infected bone or cartilage. Fibrin glue, biodegradable polymer beads, and other devices have been used as vehicles that release the antibiotic in a controlled fashion over time, and are implanted into the infected area at the time of surgical debridement.

Hypertrophic Scars and Keloid Formation

Unsatisfactory scars are uncommon on the ear. They can be prevented to some extent by adhering to the principle of tensionless skin closure, and, once formed, they are treated in standard fashion with intraleisonal steroids, and on occasion excision and reclosure. Recombinant human interferon-gamma injected intraleisonally has been used with success in keloids.

Suture Complications

Most suture complications may be avoided by the appropriate choice of suture material and by correct placement. A relatively unreactive material such as a nonabsorbable monofilament synthetic may be placed such that the knots are hidden under the thicker skin cover posteriorly. Most suture complications such as visible bridging, granulomas, and extrusion may be treated with removal of the offending suture at a time when the ear has healed into its new position and is unlikely to move, usually after 2 months.

Hypoesthesia

Decreased sensation over the auricle is a common postoperative complaint. Return of sensation usually occurs over a period of months as fine sensory nerve endings from the rich
circumaural network grow back in to replace those sectioned at surgery.

Unfavorable Cosmetic Result

Sharp Ridges

These may be produced by the cartilage cutting techniques; they are permanent and should be avoided. Stenstrom's work has shown that gentle abrasion of the anterior perichondrium alone is enough to create a tendency of the cartilage to curve toward the intact perichondrium of the posterior surface. This tendency can be enhanced and reinforced with Mustarde-Furnas-type sutures.

Deformities of the Antihelix-Helix Curvature

The placement of sutures is done in a radial fashion to recreate the normal antihelical curvature. Failure to create a cosmetically appropriate curve may lead to a "vertical post" deformity where the ear appears folded back about a vertical line. The sutures need to be tightened sequentially and equally to ensure that the setback of the new antihelix is uniform from superior to inferior. The new antihelix may be placed anywhere in the scapha by current surgical techniques, and thus the procedure should be designed such that the antihelix lies in a natural position with a scapha of the appropriate width. The actual roll of the antihelix may be made too wide or narrow depending on the amount of cartilage spanned by the sutures.

Position

Ideally the ear should lie approximately 18 mm from the mastoid as measured from the lateral surface of the helix; thus, an ear may be incompletely corrected, over-corrected ("pinned-back" appearance), or asymmetric from side to side.

Obliteration of the Postauricular Sulcus

Blunting of the sulcus is a result of excessive skin excision, producing tension on the wound closure and allowing tenting of the skin across the sulcus. Evidently, avoidance of the problem is achieved by excising only the skin that is in excess.

Telephone Ear and Reverse Telephone Ear

These are wonderfully descriptive terms that refer to an abnormally pinned-back-appearing concha in the first case, and an abnormally prominent-appearing concha in the second case. It usually occurs because of either over- or undercorrection of a deep conchal bowl.

Summary

Otoplasty is a challenging procedure. The techniques available - cartilage excision, sutures, and cartilage scoring or rasping - must be creatively combined to address the specific problems of each individual patient. The goals are a natural appearance and symmetry. Conservatism in cartilage excision is the rule and the operated look should be avoided.
Newborns with auricular deformities can be successfully treated with simple molds in the first few days of life.
Each plastic surgeon's skills are tested significantly when used to correct a congenitally malformed auricle. Although numerous publications have dealt with microtia by defining the condition's many grades, this chapter will concentrate on categories that frequently respond well to surgical reconstruction.

History and Classification

In 1959, Tanzer published the first article about using autogenous rib cartilage in auricular reconstruction for congenital microtia. Seven years later, Cronin began applying silastic as an implant material to correct the condition. Today, the world's foremost authority on auricular reconstruction is Brent, who began recording his work in 1974.

This chapter will classify microtia with a scheme described by Marx in 1926 and amended by Aguilar and Jahrsdoerfer in 1988. It divides microtia into three major grades. Grade I is a normal ear. In grade II, some of the auricular framework is present, but there are obvious deformities. Grade III describes the standard "peanut ear", which covers anotia - Marx's grade IV.

A clinical classification of auricular defects set forth by Tanzer in 1977 consists of five categories. These groups include:

1. anotia

2. complete hypoplasia (microtia):
   with atresia of the external auditory canal
   without atresia of the external auditory canal

3. hypoplasia of the middle third of the auricle

4. hypoplasia of the superior third of the auricle
   constricted ear (cup and lop)
   cryptotia
   hypoplasia of entire superior third

5. prominent ear.

Eleven years before Tanzer developed his classified description Rogers published a similar delineation that divided auricular defects into (a) macrotia, (b) lop ear, (c) cup ear, and (d) prominent ear. In 1988, Weerda compiled all these classifications into a concise document, presenting definitions that were proposed by Marx and Tanzer and revised by Rogers.
Weerda's system included surgical guidance for each stage. Aguilar summarized these in his review.

**First-Degree Dysplasia**

In this first level, most structures of a normal auricle are present. Only minor deformities exist. Normally, reconstruction does not require additional skin or cartilage. Conditions include:

- macrotia and protruding ears (prominent ears, bat ears)
- cryptotia (pocket ear, group IV B (Tanzer))
- absence of the upper helix
- small deformities (absence of the tragus, satyr ear, Darwin's tubercle, additional folds (Stahl's ear))
- colobomata (clefts, transverse coloboma)
- lobule deformities (pixed lobule, macrolobule, absence of the lobule, lobule colobomata (bifid lobule)).

Weerda's classification separates *cup ear deformities* into three classes, two of which are addressed under this category:

- **Type I** demonstrates a cupped upper portion of the helix, hypertrophic concha, and reduced height. Its synonyms include lidding helix, constricted helix, group IV A (Tanzer), lop ear, and minor (mild or moderate) cupping.

- **Type II** involves more severe lopping of the ear's upper pole. Here, rib cartilage is used as support when a short ear must be expanded, or the auricular cartilage is limp.

**Second-Degree Dysplasia**

Under this definition, only some normal auricular structures are recognizable. This stage is also called second-degree microtia (Marx). Additional skin and cartilage are employed to accomplish necessary partial reconstruction. *Mini-ear* falls under this category, as well.

Second-degree dysplasia describes Weerda's type III cup ear deformity as a condition in which the auricle is entirely malformed. Its synonyms are cochleshell ear, constricted helix, group IV (Tanzer), and snail shell ear.

**Third-Degree Dysplasia**

No normal auricular structures are present in third-degree dysplasia, requiring total reconstruction with skin and large quantities of cartilage. Its synonyms include complete hypoplasia group II, peanut ear, and third-degree microtia (Marx). Concomitant congenital atresia normally occurs in this phase.
Unilateral

In this circumstance, one ear is normal, and no middle ear reconstruction is performed on any child. Usually, auricle reconstruction does not become an alternative until a child reaches the age of 5 or 6.

Bilateral

Under this condition, a child is eligible for a bone-conducted hearing aid before his or her first birthday, middle ear surgery without transposition of the vestige at age 4, and bilateral auricle reconstruction at 5 or 6.

It is important to realize that the following recommendations are not absolute, and many unilateral cases have benefited from restoration of directional sound via atresia repair. For bilateral microtia, a bone-conducted hearing aid can be placed at birth. Also, even in bilateral cases, middle ear surgery can follow the first two stages of auricular reconstruction instead of being the first procedure.

Physician and Patient Information

When concomitant atresia is present in the treatment of congenital microtia, complete coordination between otologist and plastic surgeon is a must. The components of evaluation that should guide that coordination include

- the patient's age
- the grade of deformity
- the size of rib cartilages
- the presence of atresia
- the otological analysis
- photographs.

The management and treatment of microtia is accomplished in five stages:

Stage I - auricular reconstruction via the creation of a cartilaginous network derived from autogenous rib cartilage.

Stage II - lobule transposition.

Stage III - atresia repair by the otologist.

Stage IV -tragal construction.

Stage V - auricular elevation.

In this approach to treatment, the plastic surgeon's work should be done first, performing the procedure in a way that most efficiently reconstructs the microtia atresia complex.
Optimally, microtia correction should begin when the patient is 6 years old, especially in unilateral cases. At 6, the patient not only has sufficient cartilage to permit surgical reconstruction, but also is mature enough to manage the necessary postoperative care. Although bilateral microtia and atresia cases can be performed when the patient is younger than 6, the operation should not occur if insufficient cartilage exists to form a new ear.

Historically, material sources other than cartilage have proven failures. Neither irradiated cartilage nor silastic have been stable. Irradiate cartilage reabsorbs, whereas silastic tends to extrude over time. Furthermore, silastic implants are notorious for their inability to withstand trauma.

Diagnost Tests and Preoperative Preparation

Photographs of the patient are an integral element of preoperative planning, but the most important component is the proper preparation of the template. In unilateral cases, the template is based on the patient's contralateral ear. In bilateral cases, the mother's ear serves as the model. Measurement of the cartilage framework and its placement on the side of the head should be exact.

Radiologic examination should precede surgery, as well as a high-resolution computed tomography (CT) scan of the temporal bones. Although a CT scan is not necessary for microtia, it provides the plastic surgeon and otologist with information they can use to describe the entire reconstruction process to the patient's family before any surgical procedures occur.

Surgical Reconstruction: Congenital Microtia, Grade III

Stage I

The figures show auricular reconstruction undertaken during Stage I. Note that cartilage dissection is extraperichondrial, and there is no stripping of perichondrium at any point during the rib harvesting. The eighth rib is anchored to the sixth and seventh complex by stainless 5-0 wire, a technique popularized by Brent. When performed by trained surgeons, it produces very reliable results.

Stage II

The figures show lobule transposition that occurs in Stage II. An incision high on the back of the ear avoids protrusion of the lobule. The inferiorly based pedicle flap is very thin, so it should be handled with great care.

Stage III

It is after the first two procedures have been completed that the otologist performs atresia repair. Maintaining this sequence is important because the temporal bone remnant is in only one location, so the opening to the remnant can be made in only one place on the overlying skin. This makes it simple to line up the framework where the otologist has drilled the canal.
Complication rates increase substantially if the otologist drill the canal first, and it makes placing a cartilage framework around an external canal more difficult. In addition, the possibility of compromised blood flow and increased scarring make complications more difficult to avoid. The figures demonstrate the creation of the ear canal within the cartilage framework.

**Stage IV**

The trigal reconstruction is a delicate operation that requires ample presurgical planning.

As shown in the figures, a composite cartilage graft is normally acquired from the contralateral ear and placed in the proper location for the new tragus. A J-type incision is used to create a pocket for the new cartilage. The composite graft is then rotated 45° to face forward, covered by the anterior auricular skin.

**Stage V**

The figures show auricular elevation, and the reconstructed ear is shown. Elevating the auricle is the most dramatic stage of this procedure, because it brings the ear out from the side of the head, allowing it to assume a more natural appearance.

The split-thickness skin graft placed in the back of the ear should be reasonably thin to provide sufficient coverage and not expose the underlying cartilage, while still allowing proper soft tissue to nourish the skin graft.

The most important postoperative objective is to prevent the auricle from retracting significantly to the side of the head by encouraging the patient to move the ear forward and to continue flexing the auricular cartilage. A polyform splint may be fabricated to prevent retraction during the healing phase.

**Grade II Reconstruction**

Grade II auricular deformities involve those lesions that are not covered by descriptions of the "peanut ear" or those associated with Tanzer's grade IV. Therefore, reconstruction of grade II deformities first requires proper identification of the auricular imperfections. Is a part of the helix missing? To what degree? Is there a portion of the scapha missing? Are the lobule and tragus present? Does the conchal bowl exist?

"Lop ear" is a common description of any auricular abnormality that features a downward overgrowth of the helical rim. Insufficient scapha formation or the absence of the ear's antihelical bend can cause this irregularity. For this condition, I favor the Bard Cosman technique shown in the figures.

A Stahl ear deformity is just an extra fold, usually occurring in a posterior direction off the fossa triangularis. Treatment consists of removing the offending cartilage.
Correcting a lobule deficiency requires knowledge of the lobule's ability to reform. If one considers a Gavello flap technique an appropriate method, then the excess cartilage can be acquired from the contralateral ear and placed in the area along the lobule.

Perhaps the most common grade II operation is the correction of preauricular pits. It requires minimal plastic surgery experience. A direct excision is necessary, as well as following any anterior or posterior tracks. These procedures should be done before the patient's exposure to infection increases or a sebaceous cyst develops.

Complications

During surgical reconstruction, complications are possible, including

- skin necrosis overlying cartilage framework
- chondritis
- reabsorption
- malpositioning of the auricular implant
- tissue breakdown of the skin graft or of the ear's posterior aspect
- keloiding of the donor incision site or of the skin graft areas.

Placement of the cartilage graft severely strains overlying skin, allowing the potential for skin necrosis. Also, infection of cartilage can produce reabsorption, and the framework can be positioned improperly. Grafting procedures always possess the capacity for graft loss. The possibility of keloid formation increases when the skin graft is harvested from the abdomen or the buttock area.

These procedures can produce emergencies, too. They include

- Stage I - pleural tear, pneumothorax, pneumomediastinum
- Stage II - lobule necrosis
- Stage IV and V - chondritis.

Conclusion

There are two primary demands with which plastic surgeons should comply to provide appropriate care for patients who need surgical correction of congenital microtia. First, surgeons must sharpen these skills and keep them honed. Second, surgeons must practice the team approach with otologists as detailed in this chapter. Failure to offer this tandem approach to treatment results in a significant disservice to patients and their families. Combining the advantages of these disciplines invariably generates a level of care far superior to that created by independent work.
Vision

There are several ophthalmic terms that will be useful in our discussion. Visual acuity refers to the ability of the eye to resolve points in space as being separate. An eye with a visual acuity of 20/20 means that the eye can see the same standardized Snellen chart letter at 20 feet that a "normal" eye can see at 20 feet. Someone with a 20/400 visual acuity would need to be 20 feet away from a letter that could be seen from 400 feet by the "normal" eye. Acuity in the younger age groups is more difficult to determine accurately but a simple measurement is to see if the eye fixes on and follows a small moving object.

Refractive error is typically an inherent optical inability of the eye to focus the entering light rays into a clearly focused image on the retina. This can generally be corrected by glasses, contact lenses, surgery, and possibly laser surgery.

The word astigmatism suggests that the wavelengths of light entering the eye at 90° angles to one another are focused at two different distances or focal lengths in relation to the retina. This creates a blurred image on the retina and is a type of refractive error. Astigmatism can be induced by pressure on the eye because of a change in curvature induced on the optical surfaces of the eye. A typical example would be an orbital hemangioma.

Strabismus refers to a disturbance in ocular alignment. Typically one eye or the other will be intermittently or continuously misdirected either in (esotropia), out (exotropia), up (hypertropia), or down (hypotropia). This generally occurs on a congenital basis and can produce amblyopia (see below), or it may occur as a result of amblyopia due to loss of visual fixation.

Amblyopia is a real diminution of vision for which no appropriate structural cause in the afferent visual pathway exists. This typically occurs in one eye as a result of degradation or absence of a clear, focused image on the retina sometime during the first 10 years of life. Refractive error, strabismus, occlusion (ie, patching or eyelid swelling), or congenital or acquired ocular opacity such as a cataract or corneal scar are the usual etiologies of
amblyopia. This condition is often partially or totally reversible in children with appropriate management if it is delivered in a timely fashion. Amblyopia cannot occur after the visual pathways are fully developed, usually by the age of 10.

Four factors are required for any of us to have vision. A light stimulus from the visible spectrum of electromagnetic waves must create a sensation in the retina of the eye that must then be transmitted to and perceived by the brain. Absence of any of these results is no vision.

A newborn infant enters a visually sensitive period at or close at birth when the eye, brain, and their connections continue to develop. During this time any obstruction of the visual axis leading to deprivation of light and formed images will retard development of normal vision and produce amblyopia. Animal studies reveal in actual failure of neuronal development in the brain. The earlier in life the deprivation occurs and/or the greater its duration, the more visually devastating its effect will be. Visual development is most sensitive to disruption during the first 2 years of life. This sensitivity appears to decline markedly after age 5 or 6 but remains until the child is 8 to 10 years old.

In 1973, Awaya observed 15 infants whose eyes had been occluded for 1 week following eyelid surgery. They all had reduction of vision below 20/100 and developed marked strabismus. The amblyopia was most severe in the youngest patients. An occlusion of only 1 week produced this effect.

Hemangiomas occurring early in life that totally occlude the visual axis will produce a dense, possibly irretrievable amblyopia. Hemangiomas that do not obscure the visual axis can also lead to amblyopia, as was mentioned earlier, because of induced astigmatism and/or strabismus.

Any disease process that may be amblyogenic must be managed as early as possible. One must also be careful to avoid any iatrogenic interruption of the visual axis. A decrease in visual acuity and/or acquired strabismus should alert the surgeon that a possible threat to vision exists. A baseline ophthalmologic examination prior to surgery and follow-up examinations postoperatively should assure that any impending threat will be recognized early, so that appropriate management can be initiated immediately.

The Eye

Ophthalmic plastic surgery involves surgical procedures performed on structures near or immediately adjacent to the eye. Because of this, some understanding of its anatomy and function is paramount not only to follow along in this chapter but also to achieve a successful surgical result.

The most anterior structure is the clear, domed-shaped cornea. Its peripheral border is the limbus, which extends circumferentially 360° as the corneal junction with the sclera. The sclera is the leathery, white outer "coat" of the eye and is composed of dense, irregular connective tissue. The colored portion of the eye or iris is found just anterior to the lens. The space between the iris and the cornea is the anterior chamber, which is filled with clear fluid referred to as aqueous humor. The lens is suspended behind the pupil by the zonules. The
large cavity behind the lens is filled with a clear, jelly-like substance called the vitreous humor. The innermost layer of the eye is the retina and it is activated by the light rays that have been focused into an image by the lens after entering the pupil.

The rectus muscles are responsible for horizontal and vertical eye movements for the most part, and loss of function or restriction will produce diplopia in a person with two seeing eyes. The superior oblique not only depresses the eye but intorts it as well. This muscle runs through a "trochlea", which changes its vector of force. Damage in the area of the trochlea producing restriction of this muscle results in the Brown syndrome. The inferior oblique muscle elevates and extorts the eye. It is commonly seen during lower eyelid blepharoplasty. Injury here can also produce troublesome diplopia.

**The Eyelid**

The lower eyelid margin is at or just superior to the limbus inferiorly. Superiorly, the upper eyelid covers 1 to 2 mm of the iris centrally. The eyelid crease is generally 8 to 10 mm above the lid margin centrally. The lacrimal papillae, which denote the beginnings of the tear outflow system, are noted along with the caruncle. The palpebral fissure refers to the area between the eyelids and measures approximately 10 mm centrally. The area where the lids join nasally is referred to as the medial canthus, and the temporal union is the lateral canthus.

There are several things that should be noted. The lids can be split into an anterior and posterior lamella. The anterior lamella consists of skin, subcutaneous tissue, and orbicularis muscle. The posterior lamella includes the tarsus and conjunctiva. The levator palpebrae superioris is responsible for upper eyelid retraction and inserts on the anteroinferior one-third of the tarsus. In addition, it sends multiple strands anteriorly through the muscle to firmly attach the skin below the crease to the tarsus, thus forming the crease. The sympathetically innervated Müller's muscle arises beneath the levator and inserts at the superior aspect of the tarsus.

Retraction of the lower lid is accomplished by the inferior retractors, which are composed of Tenon's capsule, Müller's muscle, and the capsulopalpebral fascia. This fascia extends anteriorly from Lockwood's ligament, which is a condensation of fascia anterior to the inferior oblique muscle. This is connected posteriorly to the inferior rectus muscle so that when the eye looks down, the lower eyelid is retracted inferiorly to avoid obstruction of the visual axis during down-gaze.

Eyelids of appropriate dimension, apposition to the globe, and mobility are essential to the survival and normal function and comfort of the eye. In addition to protecting the eye from dust and assorted flying debris, the eyelids also move the tear film over the cornea to keep its epithelium moist and vision clear. The lids also serve as part of a "pump" to move the tears nasally into the lacrimal drainage system. The lids must have enough mobility to avoid obstruction of the visual axis and allow a full field of vision. Congenital or acquired abnormalities that alter the eyelids' ability to do the above can lead to discomfort, blurred vision, amblyopia, blindness, and even loss of the eye.

Eyelid abnormalities are most often easily diagnosed by direct examination. An ophthalmic slit lamp is useful to magnify certain structures such as small, misdirected lashes.
that are rubbing on the cornea. It is also useful for evaluating the eyeball. Fluorescein staining of the corneal epithelium due to an abrasion is easily seen with the slit lamp. A Wood's lamp and loupes are an adequate substitute.

Epithelial changes of the cornea often result from inadequate protection by the lids or abrasions due to abnormal position of the lashes or keratinized epithelium. These epithelial changes such as an abrasion cause pain, photophobia, red eye, tearing, and decreased vision, which in time can lead to amblyopia in children. In addition, this gives bacteria and fungi a foothold in the cornea stroma, which can lead to corneal ulceration, endophthalmitis (infection in the eye), blindness, and surgical removal of the eye. Although most eyelid abnormalities appear relatively benign at first, inappropriate management can lead to vision-threatening consequences.

Most eyelid surgery in adults is performed with local anesthetic in the office surgical suite. In children, however, a general anesthetic is required. No specific preoperative orders are necessary. Infiltration of the surgical field with 1% Xylocaine with 1:200,000 epinephrine with or without Wydase is recommended 10 min before incision. This will facilitate better hemostasis and therefore visualization. A 5/8-inch, 30-gauge needle is easy to control close to the eye and minimizes the chances of accidental perforation of the globe. An eye shield can be placed over the eyeball for protection. Postoperatively, elevation of the head and application of ice will minimize swelling and hemorrhage. No antibiotics are generally given. We have found 6-0 plain fast-absorbing gut suture allows for an excellent skin closure and avoids the trouble of removing sutures in a young patient. It is important to remember that any prolonged obstruction of the visual axis by patching, suture closure, or swelling is to be avoided. Amblyopia develops very quickly. Should this situation be unavoidable, an ophthalmologist should be consulted to manage the child's visual development.

The Lacrimal System

The lacrimal gland is located in the anterior portion of the superotemporal orbit. It is divided into two portions: the orbital lobe, which is posterior to the septum and therefore is intraorbital, and the palpebral lobe, which is outside the orbit. The two lobes are separated by the lateral horn of the levator aponeurosis but connected by six to eight ducts. The tear film is moved across the surface of the eye by the blinking action of the eyelids. The figure demonstrates this so-called lacrimal pump.

The lacrimal drainage system is illustrated in the figure. The tears enter the system at the upper or lower punctum. After a 2-mm vertical descent through the ampulla, the tears head toward the nose through the relatively horizontal canaliculi. In most cases these join to form a common canaliculus that empties into the lacrimal sac. The valve of Rosenmüller is located at this junction and with the assistance of several other "valves" prevents retrgrade flow. From the lacrimal sac, the tears enter the nasolacrimal duct, which opens into the inferior meatus beneath the inferior turbinate approximately 5 to 10 mm back from its anterior tip.

Children with lacrimal problems usually present with excess tearing and "goopy" eyes. Dry eye and tumors are very uncommon in children. In almost all instances, epiphora (tearing onto the cheek) is a result of blockage of the lacrimal excretory system. Diagnosis of these
problems then comes down to determining if an obstruction exists and, if so, where.

An external examination should include locating the puncta if present, ruling out corneal or conjunctival abnormalities, and palpating the lacrimal sac. If pus regurgitates from the puncta when pressure is placed on the sac, a diagnosis of lacrimal duct obstruction with dacryocystitis can be made. If not, a dye disappearance test is performed by instilling 2% fluorescein dye behind the lower lid into the inferior fornix and waiting 10 min. In cases of obstruction, the dye does not disappear. This is very dramatic with bilateral instillation of dye in a patient with unilateral obstruction.

Surgery for lacrimal abnormalities is discussed below. These procedures are usually done in the operating room under general anesthetic. There are no specific preoperative measures that need to be taken. Hemostasis during surgery can be a problem and is avoided by preoperative infiltration of 1% Xylocaine with 1:200,000 epinephrine into the lacrimal sac area. Intranasal 4% or 5% cocaine solution is also helpful for hemostasis and visualization in the nose. Postoperatively, the patient or the parents are warned about epistaxis and the possible need for packing. Instat (Johnson & Johnson) placed intranasally during the surgery will typically avoid any postoperative hemorrhage. If silicone is placed, its medial canthal position is pointed out to the parents and an effort is made to keep the child from pulling it laterally when wiping the eye. Antibiotics can be given intraoperatively for more involved intranasal surgeries, as we have seen an occasional case of preseptal cellulitis following lacrimal surgery.

The Orbit

The intricate anatomy of the orbit is beyond the scope of this chapter. The reader is referred to several excellent anatomical reference books. For purposes of this chapter, it is important to know that the orbit is bounded by bone in every direction except anteriorly, where it is bounded by the orbital septum. The soft tissue orbit is further divided into the peripheral surgical space outside the muscle cone and the central surgical space inside the muscle cone. The muscle cone is formed by the fascial connection of the four rectus muscles.

Orbital problems usually manifest themselves by proptosis (increasing anterior projection of the eye), decreasing vision, and diplopia or double vision secondary to motor nerve or extraocular muscle involvement.

In addition to the standard ophthalmic examination, the clinical examination should include evaluation of cranial nerves two through six, color vision, stereo vision, prism alternate cover testing for strabismus, retropulsion of the globe, manual palpation, exophthalmometry (anterior projection relative to lateral orbital rim), and auscultation. Further evaluation may include orbital ultrasound, computed tomography (CT) scanning, and magnetic resonance imaging (MRI). Although CT scanning remains the examination of choice for bony orbital abnormalities and soft tissue tumors that calcify, MRI utilizing an orbital surface coil is becoming more beneficial in soft tissue evaluation. Preoperative angiography is occasionally indicated to aid in diagnosis and in planning a surgical approach. Embolization of vascular tumors at this time can be done.
Pediatric orbital surgery requires general anesthesia and an operating room setting. The patient is placed in a supine, slight reverse Trendelenburg position. Hypotensive anesthesia, if available, can be useful in some cases where a vascular tumor is suspected or is located in a difficult to access area of the orbit.

Numerous approaches to the orbit are available, depending on the location of the tumor. The various approaches are illustrated. In general, the approach that allows the most direct access to the pathologic process is utilized. The superior orbitotomy is done in conjunction with a neurosurgeon. Postoperatively, the surgical site is generally drained for 1 to 2 days. Steroids can be given during the perioperative period if excess swelling is anticipated. Intravenous antibiotics are given intraoperatively if surgical time is prolonged or a sinus cavity is entered.

**Eyelid Abnormalities**

Congenital *coloboma* of the eyelid generally involves the upper lids but all four lids may be involved. The entire thickness of the lid is frequently absent. Treatment is not emergent unless there is corneal exposure. These lids can usually be closed with an end-to-end anastomosis. The edges of the coloboma are "freshened" by sharp removal of the epithelium and the lid is brought together using interrupted 5-0 Vicryl to approximate the tarsus (posterior lamella). The lid margin is approximated using three interrupted 6-0 silk sutures. One is placed at the mucocutaneous border, one at the gray line, and one in the lash line. The remainder of the skin edge is then closed.

*Epiblepharon* refers to a condition in which a horizontal fold of skin of either the upper or lower lid forces the lashes against the eye. This condition often resolves without surgery as the child grows. Rotational sutures can be used. An absorbable double-armed suture is placed deep in the fornix and brought out near the lash in a horizontal mattress-type fashion.

The term *congenital entropion*, on the other hand, suggests an actual inward rotation of the lid margin and eyelashes. This can result in corneal erosion, infection, permanent opacity with vision reduction, or loss of the eye due to infection. A debate exists concerning etiology but it appears that at least in some pediatric cases there is a defect in attachment of the inferior retractors to the tarsus and usually reestablishment of that attachment surgically will correct the problem.

*Ectropion* describes a condition in which the lid is out, away from the eye or down from its normal position near the limbus. It is almost always the lower eyelid that develops this problem. This can be congenital such as in association with Treacher Collins syndrome, or acquired secondary to scarring due to trauma, tumor, or previous surgery. Repair in children can be difficult and often requires full-thickness skin grafting. If the eye itself is tolerating the condition and not at risk due to exposure, surgery should probably be delayed as long as possible. Occasionally these eyes will improve on their own with time and digital upward massage.

*Epicanthus* refers to a relatively vertical fold of skin that is located between the medial canthus and the nose and may cover part or all of the inner canthus of the eye. Four separate
types have been described: epicanthus supraciliaris, palpebralis, tarsalis, and inversus. Repair of these entities typically requires correction of the associated telecanthus by medical canthal tendon shortening and a double Z-plasty. The Y-to-V and the Mustarde techniques have long been the standard.

Blepharophtosis or ptosis refers to a drooping of the upper eyelid margin relative to the pupil. There is no agreement on what constitutes a visually significant ptosis based on measurements of palpebral fissures or marginal reflex distance (distance from corneal light reflex to lid margin). In children, the ptotic lid can interfere with formed vision and thus produce amblyopia. In addition, there is a higher incidence of astigmatism in these patients, which also can produce amblyopia. Some children will use a chin-up head position to see, inducing a torticollis. These children need to be seen by an ophthalmologist and followed carefully for any evidence of visual decay.

Repair of these eyes is generally accomplished with a levator resection if the levator function is greater than 4 mm. If the function is 4 mm or less, this is considered poor, and a fascia lata frontalis sling is generally performed. The technique illustrated allows the lid to be raised using the vertical action of the frontalis muscle as it acts to raise the brow. Other techniques for placement of fascia or other sling materials have been described.

The combination of ptosis, epicanthus inversus, blepharophimosis (horizontal narrowing of the palpebral fissure), and telecanthus is referred to as blepharophimosis syndrome. This is transmitted as an autosomal dominant trait with 100% penetrance. The ptosis is usually associated with poor function and thus requires a frontalis sling. Improvement of the blepharophimosis generally occurs with repair of the epicanthal fold. Occasionally, a lateral cantholysis is also performed.

The large variety of eyelid tumors that can occur precludes their mention individually here; however, it is important to remember that any tumor that obstructs the visual axis or creates astigmatism can produce amblyopia, which, if not managed appropriately, could lead to permanent visual loss. A good example of this is the capillary hemangioma of the lid. These tumors can be a serious threat to visual development. Once it has been determined that the tumor is amblyogenic then treatment is instituted. In the case of a hemangioma, intralesional steroids will often significantly reduce or eliminate the tumor. A visually threatening tumor that is unresponsive to steroids can be surgically resected. Those hemangiomas that do not inhibit visual development are followed, as the vast majority will resolve on their own by age 7 or 8.

Two other tumors that are important are the angioma associated with Sturge-Weber syndrome and the plexiform neuroma of neurofibromatosis. Although these generally do not physically interrupt visual development, causing deprivational amblyopia, they can be associated with glaucoma. High intraocular pressure that is undetected and untreated can lead to severe visual loss and blindness due to glaucomatous optic nerve atrophy.

Lacrimal Abnormalities

Congenital cysts of the lacrimal gland are rare but dermoid cysts in the vicinity of the gland are more common. These will be discussed more in the section on orbital abnormalities.
Dermolipomas also occur in this area and are more solid in nature. CT scanning or ultrasound can be of great assistance in diagnosis if the clinical appearance is atypical. It is generally best to avoid surgery on this tumor because of postoperative scarring and restriction of motility, producing strabismus and therefore possible amblyopia in children under 10. "Beware the dermolipoma".

The figure depicts the external appearance of a lacrimal anlage duct. This congenital abnormality can take many forms. It may be continuous with the tear sac so that tears form on the cheek, or it may be a blind cul-de-sac. This is removed by first intubating the anlage duct and then surgically removing the epithelial lining and soft tissue surrounding the probe.

Atresia of the puncta produces epiphora. This is readily diagnosed on clinical examination as was mentioned earlier. Treatment is a conjunctivodacryocystorhinostomy (CDCR). This procedure will be discussed below.

Atresia of the nasolacrimal duct also produces epiphora and is more common. Guerry and Kendig found some impatency in 6% of random full-term infants. This obstruction is typically membranous at the valve of Hasner and usually opens spontaneously in the first few weeks of life. As in punctal atresia, these children have epiphora if the obstruction persists. Unlike punctal atresia, there is typically some associated infection/inflammation of the lacrimal sac (dacryocystitis) producing a mucopurulent retrograde drainage. If this drainage pathway is also blocked at the valve of Rosenmüller, a closed system develops that then behaves as an abscess, and a cutaneous draining fistula often is the result. This is generally associated with severe pain and preseptal cellulitis of variable severity.

Amniotoceles or congenital dacryoceles are usually manifest within the first days of life as a swollen, bluish-colored tumor in the area just beneath the medial canthal tendon. They represent a dilated lacrimal sac filled with amniotic fluid and mucopurulent material. They can be associated with obstruction of the ipsilateral nasal airway and produce some respiratory distress in these infants particularly if bilateral. Probing will generally resolve this condition but occasionally marsupialization of the dacryocele into the nose is necessary.

Treatment of nasolacrimal duct obstruction is somewhat controversial. Most authors would agree that conservative medical management is a reasonable initial step. This consists of topical antibiotic drops with or without a mild steroid. In addition, massage of the tear sac to promote drainage is encouraged. If this fails to relieve the obstruction and the patient is less than 1 year old and of manageable size, we feel a probing in the office using topical anesthetic is very successful with minimal morbidity. It also avoids the risks and expense of a general anesthetic.

If probing fails or the child presents at an older age, a probing with silicone intubation and infracture of the inferior turbinate under general anesthetic is performed. The silicone is left in place for at least 6 weeks and then removed in the office. In a small number of children, this too will fail and then a dacryocystorhinostomy (DCR) with silicone intubation is indicated.

The DCR is essentially a bypass procedure of the nasolacrimal duct. The lacrimal sac is marsupialized to the nasal cavity just anterior to the tip of the middle turbinate. A silicone
A stent is placed for 6 weeks. If the canalicular system is absent (ie, punctal atresia) or obliterated due to scarring, a pyrex Jones tube is placed to facilitate tear drainage.

Soft tissue trauma is discussed in the chapter by Farrior and Clark, but eyelid lacerations involving the canalicular of the lacrimal excretory system should be mentioned here. Canalicular laceration should be suspected in any laceration nasal to the puncta. Usually in children this requires an examination and repair under general anesthesia. If one or both canalici are lacerated, the system is intubated with silicone. The canalici are then approximated over the silicone stent with 7-0 Vicryl in the soft tissues nearby. The remainder of the laceration is closed in a standard fashion, being meticulous at the lid margin to avoid a notch. The silicone can be removed in 6 weeks.

Tumors of the lacrimal excretory system are rare and usually occur in adults. Neonates with swelling and bluish discoloration in the area of the lacrimal sac associated with some respiratory distress typically have an amniotocele or dacryocele. In this case the lacrimal sac is filled with amniotic fluid and may partially obstruct the ipsilateral nasal airway. This condition generally responds to probing but on rare occasion requires marsupialization of the cystic lacrimal sac into the nose. Meningoceles and meningoencephaloceles can also present in this area.

**Orbital Abnormalities**

There are many congenital abnormalities of the bony orbit, which are covered in the chapter by Marentette and Gorlin on craniofacial surgery. Our discussion here will focus on the more common pediatric orbital abnormalities listed in Table 1.

**Table 1. Common pediatric orbital abnormalities**

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Cellulitis</td>
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<tr>
<td>Nonspecific orbital inflammation (pseudotumor)</td>
</tr>
<tr>
<td>Dermoid</td>
</tr>
<tr>
<td>Hemangioma</td>
</tr>
<tr>
<td>Lymphangioma</td>
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<tr>
<td>Rhabdomyosarcoma</td>
</tr>
<tr>
<td>Glioma of optic nerve</td>
</tr>
<tr>
<td>Optic nerve sheath meningioma</td>
</tr>
<tr>
<td>Chloroma (leukemia)</td>
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<tr>
<td>Metastatic disease (neuroblastoma)</td>
</tr>
<tr>
<td>Retrobulbar hemorrhage.</td>
</tr>
</tbody>
</table>

Periocular cellulitis is usually the result of direct trauma or infection of adjacent structures such as the lacrimal sac, paranasal sinuses, nasopharyngeal cavity, blood vessels, or brain. Inflammation located in the soft tissue surrounding the eye in front of the orbital septum is referred to as *preseptal cellulitis*. The underlying eye is quiet with no evidence of involvement. Children will often present as somewhat toxic and be subdued. They should be admitted to the hospital and started on IV antibiotics such as ceftriaxone and nafcillin. Significant improvement should be noted within a few hours. If this is not the case or evidence of *orbital cellulitis* appears such as chemosis, proptosis, reduced vision, or impaired...
ocular movement, then an emergency CT scan should be done and appropriate surgical steps taken. This generally involves draining the paranasal sinuses and/or an orbital abscess. Orbital cellulitis is one of a few true ocular emergencies and if not treated properly can lead to total loss of vision and complete ophthalmoplegia.

Nonspecific orbital inflammation has been referred to as orbital pseudotumor in the past. This nomenclature is not helpful and can be confusing. Rootman has suggested a simple and useful classification for orbital inflammation. These syndromes share the common clinical findings of inflammation and histologically demonstrate polymorphous infiltrations of inflammatory cells. Based on location, the acute and subacute inflammations can be divided into anterior, diffuse, apical, myositic, and lacrimal.

The differential diagnosis for these tumors is long. In children, clinical findings of rhabdomyosarcomas, metastatic neuroblastomas, or leukemic infiltration can mimic those seen in orbital inflammation. In general, treatment with systemic prednisone is initiated. This usually produces a dramatic response and the steroids can be tapered over the next 2 to 3 months. Failure to respond to prednisone obligates the physician and patient to a biopsy to confirm a benign process. This being the case, steroids can then be restarted or orbital radiation can be considered. Cytotoxic medications have a role in those cases that are recalcitrant to more conventional therapy.

Orbital dermoids are congenital tumors composed of sebaceous material and hair follicles. They are generally well encapsulated and represent a "rest" of primitive ectoderm left in the area of a fetal cleft. This is tissue not normally present in this location and therefore is a choristoma. Dermoids are most commonly located in the anterior portion of the supratemporal orbit. These are generally easily removed via an anterior orbitotomy. Large, deep dermoids may require a transcranial superior orbitotomy. The tumor should be removed intact as the contents can create a severe inflammatory reaction. The tumor is generally removed while the child is young to avoid accidental rupture secondary to trauma.

Hemangiomas are included here, relating specifically to the orbital location and their effect on vision. These tumors can be either superficial, deep, or combined. If the visual axis is not occluded and no significant astigmatism has been induced, these tumors are followed closely and allowed to run their course. If the vision becomes threatened, a CT scan with contrast is performed to determine the extent of the tumor. Intralesional steroids can then be given under general anesthesia with monitoring of central retinal artery perfusion. This procedure often will result in marked shrinkage of the tumor. It can be repeated two to three times; however, it is important to obtain am cortisols 6 to 8 weeks after injection because growth retardation can occur even with local infiltration of steroids. As previously described, surgical resection can be performed but is best left until the tumor is out of the growth phase and stable for at least 6 months. We have found preoperative angiography and selective embolization helpful. This shrinks the tumor making it easier to remove and hemostasis is less of a problem.

Orbital lymphatic malformations (lymphangiomas) are composed of dilated, thin-walled vascular channels lined by flattened endothelium. The absence of pericytes and smooth muscle cells in the vessel walls helps differentiate this tumor from a capillary hemangioma. The channels are filled with a clear, proteinaceous and eosinophilic fluid material. Variable
amounts of lymphoid tissue are found in the tumor. Because of this, these tumors tend to enlarge in association with upper respiratory tract infections. Recurrent hemorrhages into the tumor are common and "chocolate cyst" formation is seen.

Lymphangiomas are probably congenital and generally progress from childhood until midadolescence. Unfortunately they do not regress like the capillary hemangiomas tend to. They invade the orbital soft tissues diffusely and complete surgical removal is impossible. Careful surgical debulking is the best treatment option to date. Radiation is not effective and sclerosing solutions add more complications and should be avoided.

*Rhabdomyosarcoma*, the most common pediatric orbital malignancy, is classified into four histologic types: pleomorphic, embryonal, alveolar, and botryoid. The embryonal and alveolar are the most commonly seen in the orbit. They are highly malignant and are characterized by a rapidly enlarging orbital soft tissue mass. Early biopsy to confirm the diagnosis followed by irradiation and chemotherapy allow a high rate of cure. Radical surgery is no longer indicated in the initial treatment of this tumor; however, conservative resection or debulking at the time of biopsy is reasonable. The brain, lungs, and cervical lymph nodes are the primary sites for metastasis.

Optic nerve gliomas or juvenile pilocytic astrocytomas are slow-growing tumors derived from astrocytes and oligodendrocytes. Pial connective tissue septa are scattered throughout the tumor. These "benign" astrocytomas generally produce fusiform enlargement of the optic nerve with loss of the central nerve shadow on CT. The nerve will often appear "kinked" in the area of the tumor. Growth posteriorly produces enlargement of the optic canal. Optic gliomas are more common in patients with neurofibromatosis.

Loss of vision generally occurs first but the tumor is not suspected by the parents until secondary strabismus or proptosis develop. The tumor expands in the central surgical space so the proptosis is straight out or axial. There is typically no associated pain unless the cornea is chronically exposed.

If the tumor is solitary and confined to the orbit, it can be surgically removed once vision is clearly and significantly compromised. Intracranial extension requires a craniotomy. Chiasmal involvement based on the visual field of the contralateral eye and neuroimaging precludes surgical removal. Use of chemotherapy and radiation therapy for this tumor is controversial.

Optic nerve sheath meningiomas are not very common in the pediatric age group but are mentioned here because they seem to be more aggressive in children than in adults. They can clinically present very much like optic nerve gliomas. In addition, fundus examination (retina, optic nerve, etc) classically reveals optic nerve atrophy with associated optociliary shunt vessels. CT examination is generally very helpful in making the diagnosis. The tumor has irregular borders and the nerve sheath is generally calcified, giving a "tram-track" appearance on coronal examination. The central nerve shadow is generally preserved.

A biopsy can be performed usually via a lateral orbitotomy. The biopsy specimen must include some optic nerve because the meningotheelial arachnoid hyperplasia seen in patients with gliomas can mimic the histopathology of a meningioma. Once the vision has deteriorated
or problems develop due to proptosis, the tumor can be resected. As mentioned, these tumors appear to be more aggressive in children and every effort should be made to resect the tumor prior to its gaining access to the intracranial cavity.

**Granulocytic cell sarcomas**, seen in association with myeloblastic leukemias, are green-colored tumors referred to as chloromas. This color is produced by a pigmented enzyme called myeloperoxidase. Management of this tumor generally entails appropriate diagnosis and treatment of the underlying systemic disease. Radiation or surgical drainage/resection can be used as indicated.

**Neuroblastoma** leads the list of metastatic tumors to the orbit in children. It arises from the adrenal medulla of infants and young children. Once in the orbit, it commonly produces periorbital hemorrhage, lid ecchymosis, and marked proptosis. These patients develop pallor, weight loss, and a palpable abdominal mass. Treatment is managed by a pediatric oncologist.

**Retrobulbar hemorrhage** is included here because it is a potentially blinding condition that, if managed appropriately, is little more than a nuisance. It can occur spontaneously, such as within a tumor, or as a result of trauma, or iatrogenically during a retrobulbar injection or after surgery.

The problem arises when an active arterial hemorrhage is confined to the orbital space by bone and the orbital septum. This causes a marked increase in orbital pressure, which in turn can interrupt perfusion of the globe via the central retinal artery.

Early detection and treatment of this condition by opening the orbital septum or lateral canthus, or, in some severe cases, bony orbital decompression will relieve the pressure and prevent loss of vision.
### Table 1. Specialities represented within a cleft team

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<thead>
<tr>
<th>Team concepts</th>
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<tbody>
<tr>
<td><strong>Personnel</strong></td>
</tr>
<tr>
<td>Specialities involved</td>
</tr>
<tr>
<td>Plastic surgery</td>
</tr>
<tr>
<td>Otolaryngology</td>
</tr>
<tr>
<td>Oral Surgery</td>
</tr>
<tr>
<td>Orthodontics</td>
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<tr>
<td>Speech pathology</td>
</tr>
<tr>
<td>Genetics</td>
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<tr>
<td>Pedodontics</td>
</tr>
<tr>
<td>Child psychiatry</td>
</tr>
<tr>
<td>Pediatrics</td>
</tr>
<tr>
<td>Nursing</td>
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<td>and a neurosurgeon</td>
</tr>
<tr>
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</tr>
<tr>
<td><strong>Support personnel</strong></td>
</tr>
<tr>
<td>Coordinator</td>
</tr>
<tr>
<td>Scheduling assistant</td>
</tr>
<tr>
<td><strong>Patient evaluation</strong></td>
</tr>
<tr>
<td>Individual evaluation versus</td>
</tr>
<tr>
<td>team visits</td>
</tr>
<tr>
<td>Coordination of surgery</td>
</tr>
<tr>
<td>&quot;Staffing&quot;</td>
</tr>
<tr>
<td><strong>Handouts</strong></td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
</tr>
<tr>
<td>&quot;In house&quot; versus coordination of local specialties</td>
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<tr>
<td><strong>Follow-up/outreach</strong></td>
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The pooling of patients from what would otherwise be several individual practices into a central cleft team also allows all members of the team to continue their education process and improve the care of each individual patient that comes to see them. This arrangement allows the team to become "patient centered" rather than "speciality centered", a concept first advocated by Koepp-Baker. This interaction between various specialties and subspecialty groups further provides an optimal training both in treatment techniques and interpersonal relationships for young surgeons and residents. In this setting they are exposed to a number of different viewpoints and protocols, which allows them to begin the filtering and molding process that is necessary as they develop their own concepts of cleft care. It is hoped that this educational process will foster the belief that the care of cleft patients must transcend egotistical and financial concerns of the individual team members. The purpose of this chapter is to generally outline a team system of cleft care with specific references to the care given by the cleft team at the University of Iowa.

Obviously, it is impossible to expect that one cleft team will be an exact copy of another. Based on an individual team's strengths and weaknesses, it will develop its own particular characteristics. Regardless of these variations, the premise of this chapter is that the team concept of care is an example of the whole being greater than the sum of its individual parts, thus allowing each group to best serve patients.

**Personnel**

**Involved Specialties**

**Surgical**

Ideally, the specialties represented within a cleft team would include those listed in Table 1, namely plastic surgery, otolaryngology, oral surgery, orthodontics, speech pathology, genetics, pedodontics, prosthetics, child psychiatry, pediatrics, nursing, and social work. At a minimum, surgical, nursing, dental, speech, and social work problems must be addressed and adequately covered. As stated previously, the surgical endeavors of many of these groups may overlap in part, but rather than being a liability within the cleft group, this can be used as a distinct advantage. First, regarding training, it allows residents from different services to have exposure to problems that are being both diagnosed and treated from different perspectives. This optimally provides for a free and open exchange of ideas between specialties and gives the trainees experience in different methods of handling complications. Another decided advantage of having several groups of people performing the same operation is increased convenience for the patient. No longer is the patient required to wait until an opening arises in a given surgeon's operating schedule. With multiple surgeons performing a given procedure, the patients can be scheduled at times that are more convenient for them and the cleft team no longer has to be idle during times of national conventions or individual vacations.

The credentialing of surgeons to perform given procedures has been and will remain a difficult problem. Prior to assuming a position as a staff member of the cleft team, an individual should have completed sufficient training that he or she is comfortable not only with the surgery itself, but also with the handling of complications. Preliminary training under a variety of teachers on a heterogeneous patient population is ideal, as this allows the
individual to develop his own approach using the best of his exposure from each source. This prevents overdependence on a narrow viewpoint of technique or philosophy, thus providing the training surgeon with an adequate variety of surgical options to allow any surgical problem encountered to be successfully overcome.

Everyone involved with the care of clefts realizes that the most critical measurement of success is long-term outcome. Thus, although no training program can provide the life-long follow-up necessary for true judgment of results, a good training program should provide adequate postoperative follow-up of at least 2 years so that some intelligent judgment can be made regarding the outcome of the techniques applied. This follow-up and observation of results over time, both good and bad, is perhaps the most critical aspect of any given surgical education.

Dental

Dental development and hygiene, particularly in the very early stages of tooth eruption and deciduous dentition, need qualified care and future planning. An early regimen of proper oral hygiene is essential to future surgical orthodontic and surgical endeavors. Obviously, expert dental intervention is necessary beginning as early as birth in some cases with application of orthopaedic positioning devices, and continues until final, permanent, stable occlusion is established and maintained. The orthodontist and surgeon must work in symbiosis as neither can achieve an optimal result alone. Often because of long distances and frequent visits that are necessary during active orthodontic care patients and families will elect to have some of their orthodontic care done locally. This is certainly acceptable as long as there is communication between the local orthodontist and the surgeon and orthodontist in the cleft team. Specifics of dental care are beyond the scope of this chapter and will be described by other authors.

Speech

Speech problems in the cleft patient are well documented and continue to be a source of difficulty in some patient's everyday interactions. Early evaluation, therapy, and careful follow-up by speech pathologists experienced in working with cleft patients is essential. Increasing use of fiberoptic evaluation requires close cooperation and interaction between the surgeon and speech pathologist. Speech pathologists play an extremely important role both pre- and postoperatively in surgical correction of speech problems. Indication for surgical intervention for the correction of cleft speech problems will be discussed by other authors.

Nursing

The nursing service provides an essential and often underappreciated role in the care of the cleft patients. A nurse who works consistently with the patients in clinic can provide tips on feeding and general day-to-day care that often may be overlooked by the medical staff. In reality these problems may be the most major frustrations the parents have in dealing with their new offspring. This same specialist should see the people in the cleft clinic, make rounds with the attending surgeons while the patient is in the hospital, and be available to take phone calls from the parents and coordinate the responses back to them. Again, along with the clinic coordinator, the cleft nurse should become a patient advocate and it is not unusual for a deep,
long-term bond to develop between the cleft nurse and the patient and their families. It is extremely important that cleft nurse specialists view their job as a stable, long-term commitment, and feel that they have the freedom to discuss any situation at any time with any team member.

Other Members of the Team

Other members of the team are often able to provide as much information and reassurance to the families during the initial visit and later as can those specialists already discussed. Specifically, genetics counseling is a vital and integral part of an initial cleft visit to any cleft team. Any guilt and accusations need to be dealt with, and factual information for further family planning must be given to the parents of the cleft child. Along with this genetics counseling, a general pediatric examination should likewise be carried out, the extent of which would depend upon the initial history and physical examination. Obviously, many children with clefts have additional special physical and psychological problems, and they will need to have these addressed every bit as expediently as their surgically correctable problems.

Although traditionally much emphasis is given to the medical and surgical members of the cleft team, anyone with experience in directing a large, integrated team caring for cleft patients realizes the truly indispensable contribution of a clinic coordinator, and reliable scheduling assistant. Every cleft team needs to have a dedicated full-time coordinator who ensures smooth patient flow during clinic days and maintains an ongoing line of communication between the patients’ families and the necessary specialists on the team. This person, of necessity, will become a patient advocate and often will provide profound insights into family dynamics and a patient's state of mind that are essential to a good outcome but would be otherwise impossible to elucidate. The wise surgeon will make good use of this information, giving it the consideration it deserves.

Because of the large number of surgeries scheduled and different surgeons involved, a centralized mechanism of scheduling must be implemented with a scheduling data base accessible by all members of the team. This prevents double bookings and individual unavailability, so that combined procedures can be smoothly scheduled and performed. Also, since nonsurgical members also have access to this schedule, optimal preoperative preparation and postoperative therapies can be performed.

Because of the magnitude of the anticipated procedures and related expenses, all families with newborn cleft children should have an initial visit with the social worker. At this point it is appropriate to identify potential family and financial problems that may surface under the stresses of the upcoming cleft care. Ideally this visit should occur at the initial visit to the cleft team and be followed up as deemed appropriate on an individual basis.

An important consideration is that as the completeness of the cleft team grows, so does the complexity and potential confusion for the cleft patients. It is not uncommon at the University of Iowa for a patient to have a scheduled return visit to our cleft clinic a month after the initial visit so that questions that have arisen since the initial visit can be answered. It is perfectly understandable that many parents are completely overwhelmed at the prospect of caring for a cleft child and are equally overwhelmed by the prospects of having to deal with so many new specialists who are attending to the care of their child. It is in this area that
the services of the clinic coordinator, the cleft nurse, and the scheduling secretary become most urgently needed.

**Patient Evaluation**

An idealized example of care for a patient with unilateral cleft lip and palate is presented. Visits to the clinic include both visits to individual specialists as well as team visits where multiple disciplines are seen. As much as possible, efforts are made to coordinate visits so that even if the entire team is not going to evaluate the patient, a number of individual specialists can evaluate the patient on the day of the visit.

Having a close working relationship and open communication, members of the team provide benefits when it comes to coordinating various surgical endeavors. We feel strongly that a number of procedures can be safely combined and performed under one anesthetic setting. We routinely perform myringotomy and tube placement along with other soft tissue surgeries and coordinate other dental and dental rehabilitation procedures in concert with soft tissue work. As long as the total time of the procedure is kept under 3 to 4 hr, we have not seen an increase in morbidity or surgical complications with this approach.

During each patient evaluation any of the members of the team may request that the patient be brought before the entire cleft team for a staffing. Occasionally, because of the extra time involved, patients may be reluctant to remain, but we have found that this staffing is essential for maintaining good communication between the involved specialists. Also we have found that this provides the ideal opportunity for the members of the team involved with scheduling to become aware of exactly what is being planned for this patient and to know who will need to be involved so that appropriate adjustments in surgical schedules can be made.

**Treatment**

Individual aspects of techniques and treatment will be covered elsewhere in the text. However, every specialist who practices at a referral institution must be aware that often coordination of an individual patient’s care with local specialists may be necessary. Increasing occurrence of two-career families, skyrocketing transportation costs, and expanding areas of coverage dictate that certain patients will not practically be able to come to a central cleft clinic for every single aspect of their care. Many times there are well-trained, experienced professionals available locally who are more than willing to provide ongoing care and follow-up. It is our feeling that these people should be maximally utilized and good relations with them must be maintained. This concept helps support the belief that referral to the university setting can be a two-way street. Each individual on the cleft team should expect to spend a sizable portion of his/her office time on the phone to local specialists each week. This close communication with local referring professionals is essential not only for the good care of an individual patient, but for the ongoing viability of the cleft clinic itself.
This chapter will discuss the basic surgical management of a patient with a unilateral cleft lip. There are many factors that influence the management of the patient with this disorder. They depend on the extent of the cleft deformity, the presence or absence of any other associated abnormalities, and the techniques favored by the surgeon performing the repair, which are all important variables in determining the final management of the cleft patient.

It must be emphasized that the importance of having a cleft team is vital and benefits the management of every patient.

Anatomy

In the normal patient, the orbicularis oris is a complete muscular sphincter of the opening of the oral cavity. This muscle is not covered by fascia and consequently contacts the skin anteriorly, and on the oral surface contacts the labial mucosa.

In the unilateral cleft deformity, the extent of the muscular deformity is dependent on the degree of the cleft deformity. When there is a minimal cleft deformity, the majority of the orbicularis oris muscle is intact and does form a sphincter with only the lowest portions of the orbicularis oris muscle divided at the cleft margins. When the cleft itself involves a significant portion of the lip, there is complete disruption of the orbicularis oris muscle (Fig 1). The fibers are directed upward along the margins of the deformity and may not connect with fibers of the opposite side of the cleft deformity. In a near-total or any complete cleft of the lip, these muscle fibers are directed superiorly with fibers inserting into the area of the base of the ala on the cleft side on the lateral segment and the medial segment fibers insert near the base of the columella. Thus, these muscle fibers have both an abnormal direction and insertion.

The vascular supply is also abnormal in the cleft lip patient (Fig 2). The normal arterial supply of the upper lip is obtained from the superior labial artery. This is a branch of the facial artery. These arteries arise near the corner of the mouth and are directed close to the free border of the upper lip. In the normal patient, the arteries from each side have several anastomoses in the area of the midline of the lip. During its passage through the upper lip, the superior labial artery has several branches that course upward, including the lateral nasal artery toward the area of the nasal alar base, branches that course superiorly to the midportion of the lip, and the ascending septal branch is directed toward the columella, which anastomoses with its fellow of the opposite side. In the unilateral cleft, the superior labial artery follows along the margins of the cleft when there is a significant degree of clefting and does not anastomose with its fellow of the opposite side. On the medial side of the cleft defect, the superior labial artery is usually much smaller in the area of the nasal columella,
which results in a diminished blood supply to this area.

Patients with an incomplete cleft and a complete cleft of the lip are illustrated in Figs 3 and 4.

**Timing of Lip Repair**

In the USA the majority of lip repairs are performed in accordance with the "rule of tens". Basically, this rule is designed to insure that the infant is of adequate size and allows a proper period of time for complete evaluation of the infant for other abnormalities before the lip repair is performed. In the "rule of tens" the infant should be at least 10 weeks of age, have a hemoglobin of 10 g per 100 mL, and weigh at least 10 pounds.

A few surgeons in the USA and in other countries operate on infants at a much earlier age. This varies from a few hours after birth (usually performed under local anesthesia) to a few weeks of age. The advantages of a much earlier lip repair are alleged by its proponents, who believe that maximal healing potential is present at this time, it avoids an additional separate hospitalization, allows the parents to take home a more normal-appearing child, and it may provide earlier, balanced facial musculature to facilitate feeding.

**Goals of Primary Lip Repair**

The primary goal of lip repair is to reconstruct a functional lip that has a normal cosmetic appearance. In order to achieve these goals, there should be reconstruction of the lip with correct alignment of the orbicularis oris muscle, a Cupid's bow, symmetric reconstruction of the vermillion, the creation of a nasal floor and sill, and symmetrical placement of the base of the ala and columella. In addition, many surgeons perform a reconstruction of the nasal deformity of the lower portion of the nose at this time. The goal of this reconstruction is to lengthen the columella on the side of the cleft, correct the deformity of the lateral crura of the lower cartilage, and correctly position the alar base. These goals should be attempted to be attained without the excessive sacrifice of tissue in the area of the cleft and in as nontraumatic a manner as possible. One must always remember that animal studies have shown that unilateral lip repair does influence facial growth. This consideration is vital and must be remembered whenever a unilateral cleft lip repair is performed.

**Lip Adhesion**

In some centers lip adhesion may be used as a preliminary step in the management of complete unilateral clefts of the lip. The procedure is a surgical technique that uses tissue that is usually discarded at the time of primary lip repair to convert the complete unilateral cleft into an incomplete unilateral cleft. This allows the lip to act as an orthodontic appliance, which can improve the alignment of the maxillary arches and may allow an enhanced result from the repair of the lip at the time of the definitive lip repair.

There are basically two types of lip adhesion used. The original description of the use of medial and laterally based rectangular flaps of tissue normally discarded located in the superior portion of the cleft was described by Randall (Fig 5). Walker et al used inferiorly
based flaps at the vermilion border to construct the lip adhesion (Fig 6). In these techniques, lip adhesion is performed on patients from 1 to 4 weeks of age with the definitive lip repair usually performed 3 to 6 months later.

The primary advantages of lip adhesion or the conversion of a complete cleft to an incomplete cleft is the improvement in the alignment of the maxillary arches and a subsequent narrowing of the width of the cleft defect itself. Disadvantages include the extra operation required, the possible increase in the amount of tissue that must be excised at the time of the definitive lip repair, and the possible dehiscence of the adhesion itself. In addition, not all cases result in improved alignment of the alveolar arches, as occasionally the smaller arch segment will collapse inward rather than improve its alignment.

The alignment of the alveolar arches may be markedly improved by the use of the nonsurgical technique of presurgical orthopedics by a maxillofacial prosthodontist. This technique involves the use of the prosthetic appliance to improve the position and alignment of the maxillary arches prior to the surgical procedure. When successful, this will allow repositioning of the abnormally placed maxillary segments and the subsequent decrease in the width of the cleft defect. This also facilitates the definitive lip repair and eliminates excessive tension in wide clefts of the lip.

In this method of therapy, treatment by a prosthodontist usually begins at approximately 2 weeks of age with the use of a functional appliance that is fitted to the nipple of the feeding bottle and pacifier (Fig 7). The functional appliance is constructed from an impression taken during the second week of the cleft infant's life. An appliance is designed so that pressure is applied in the designated area to move the maxillary segments into a more favorable anatomical position. This appliance obviously functions only when a child sucks and must be readapted every 2 weeks as the segments move and facial growth occurs. The readaptation of the appliance usually involves obtaining a new dental impression and redesign of this appliance. Successful use of this procedure usually requires 3 to 6 months with the vast majority of patients ready for the definitive lip repair at 4 months of age.

The advantages of a presurgical orthodontic therapy include the improvement in the alignment of the maxillary arches, which facilitates improved surgical results with better symmetry in the underlying skeleton and nose, and the completion of a repair with less tension. It eliminates the necessity of a surgical procedure such as the one used in lip adhesion to attempt to correct this problem and allows a more controlled correction of the underlying skeletal position than does the adhesion. Disadvantages include the necessity of an experienced prosthodontist in order to construct this appliance and the requirement of multiple visits and modifications of the appliance itself to obtain the desired results. In addition, this therapy may not be covered by the patient's health insurance.

It would appear that lip adhesion at this time in the USA is used less frequently than it was 10 to 15 years ago. However, in selected cases it has the potential to yield improved results when presurgical orthopedics are not available or cannot be used because of the multiple visits required prior to performing this definitive lip repair.
Definitive Lip Repair

There have been numerous methods of lip repair described in the medical literature. The two most commonly used techniques in this country are the rotation-advancement method and the interdigitation of triangular flaps; they will be discussed and described in detail.

The rotation-advancement method was originally described by Millard and has undergone some changes, but the basic concept still remains. In this technique the medial flap is rotated downward while the lateral flap is advanced as illustrated in Fig 8. This technique is probably the current method of choice for the repair of most clefts of the lip. The advantages of this technique include minimal discarding of tissue; a suture line that is superior to all others as it recreates the philtrum on the cleft side; easy access; and the facilitation of primary reconstruction of the columella, nasal tip, lower lateral cartilage, and alar base, and the creation of a nasal sill during its performance. Most importantly, it is an extremely flexible technique that enables the surgeon to adjust the repair as it proceeds. The disadvantages of this technique include the occasional difficulty in closing an extremely wide cleft without extensive undermining of the lip and the tissue overlying the face of the maxilla, which may result in an excessive tension on the lip closure, and the potential for creating a small nostril on the side of the cleft, which in the author's experience is nearly impossible to adequately correct. In addition, it is often difficult for an inexperienced surgeon to obtain optimal results as it requires a significant amount of surgical judgment at the time of the procedure, rather than being based on exact, measured methods.

The second most common method of lip repair involves the use of a triangular flap to reconstruct the cleft defect. This technique was described by Tennison and popularized by Randall. In this technique a single inferiorly based triangular flap on the cleft side of the defect is inserted into an incision on the noncleft side in the middle portion of the lip (Fig 9). The main advantages of this technique are that it is based on exact and careful measurements using defined landmarks, which are easily taught, and it can be performed by the inexperienced or occasional operator. In addition, it can be easily used on wide clefts and it maximally preserves tissue. Disadvantages of this technique include a failure to restore the philtral column, the creation of a scar that is occasionally not cosmetically pleasing, a greater degree of difficulty in performing a primary nasal repair at the time of the lip repair, and the inability to modify or adjust the repair once it is initiated by the surgeon.

Surgical Technique - General

Lip repair is performed under general anesthesia using a noncuffed endotracheal tube. The anesthesiologist must secure this endotracheal tube in the midline without distorting the lower lip or commissure. At our institution this is usually obtained by taping the tube to the chin. Once the tube is secured in place, a hypopharyngeal pack is placed to prevent aspiration of blood during the procedure. Some authors also inject a small amount of a local anesthetic agent with a vasoconstrictor to decrease bleeding. However, blood loss is usually minimal (less than 30 cc when general anesthesia is used without the injection of any vasoconstricting agent). If a vasoconstricting agent is injected, a minimal amount must be used to prevent significant tissue distortion that would affect the final result. After the infant has had his skin prepped and is draped, important anatomical landmarks are marked using gentian violegt applied with a fine-pointed, wooden, cotton-tip applicator used as a marking pen. It has been
my experience that most commercial marking pens do not have a fine enough point to be adequately used for this purpose (Fig 10). All of these landmarks can be visually identified and these include the peak of Cupid’s bow on the noncleft side (3 on Fig 10), the midpoint of the central portion of the lip (4), the base of the columella on the noncleft side (2) and on the cleft side (8), the base of the ala on the noncleft side (6) and on the cleft side (7), the commissure on the noncleft side (12) and on the cleft side (13), and the midpoint of columella (1). Once these points have been determined, the remaining points may be located. The peak of Cupid’s bow on the medial side of the cleft (5) is determined by measuring the distance between the peak of Cupid’s bow on the noncleft side and the midpoint of the lip (the distance from 3 to 4). The peak of Cupid’s bow on the medial side of the cleft is an equal distance from the midpoint of the lip and this determines point 5. The peak of Cupid’s bow on the lateral portion of the cleft side is determined by measuring the distance from the commissure to the peak of Cupid’s bow on the noncleft side (the distance from 12 to 3) and is an equal distance from the commissure on the side of the cleft (5 equals the distance from 12 to 3 to 13 to 5’). This is usually located where the vermillion portion of the lip begins to attenuate on the side of the cleft. This may frequently be further lateral than the measured location of points in the actual surgical repair. This more lateral point should be the location of point 5’. Prior to initiating the operative repair itself, the high point of Cupid’s bow on the medial and lateral cleft segments should be permanently marked so that it does not become obliterated during the procedure. This is usually performed by dipping a 25-gauge needle in gentian violet and tattooing these points on the lip by inserting the needle through the skin at points 5 and 5’.

**Surgical Technique - Rotation Advancement**

In the rotation advancement technique the same anatomical landmarks described in Fig 10 are utilized. The medially based rotation flap is outlined beginning at the high point of Cupid’s bow on the medial cleft segment and follows the vermillion cutaneous junction initially in a superior direction and then curves obliquely upward toward the base of the columella to the midportion of the columella (Fig 11). The superior aspect of the incision is no longer at the vermillion cutaneous junction but is medial to this. This allows the creation of the c-flap when the vermillion cutaneous junction incision is continued superiorly into the floor of the nose. When undermined, this flap may be sutured on itself to lengthen the columella on the cleft side (Fig 12).

The laterally based advanced flap is developed by following the vermillion cutaneous junction from the high point of Cupid’s bow on the lateral cleft segment (5’ on Fig 10) into the nose, as is done on the medial segment. A horizontal incision at the junction of the lower margin of the ala with the lip on the lateral segment is then made following along the margin of the ala laterally and then superiorly. This will allow the lateral flap to be advanced medially into the defect created by the downward rotation of the medial cleft segment. It also allows the alar margin to be freed totally from the underlying maxilla, and provides wide exposure to the lateral portion of the cleft nose.

When performing the incisions on the medial segment, the incision along the curvilinear line from 5 to 1 is through the skin only (Fig 12). The incision from point 5 along the vermillion cutaneous junction to the floor of the nose is through the skin and underlying subcutaneous tissue all the way down to but not through the mucosa on the oral side of the
medial cleft segment. This will allow the creation of a turnover flap of additional oral mucosa based on the mucosal surface of the medial segment of the flap. It can be used to augment the mucosal closure of the cleft defect if necessary.

On the lateral cleft segment the incisions are through-and-through directly downward through the previously drawn lines from points 5' to 1' and extending into the floor of the nose. This allows the creation of a vermilion flap laterally based on the vermilion of the lateral cleft segment, which can be used to augment vermilion fullness when the vermilion is reconstructed in the final stage. This is frequently necessary in bilateral clefts of the lip.

The skin is then undermined from the underlying muscle for a distance of approximately 1 cm on the medial and lateral segments of the lip. The underlying mucosa is also dissected from the overlying muscle for a similar distance. This provides direct exposure of the abnormally inserted orbicularis oris muscle underlying the skin. The muscle attachments are transected from its insertion at the base of the columnella medially and at the base of the ala laterally. Then a back cut is performed as illustrated in Fig 13. This allows the creation of medially and laterally based flaps of orbicularis oris muscle, which then can be rotated downwards and sutured together to reconstruct the orbicularis oris muscle later in the procedure.

Next, attention is directed toward the nose itself. The skin is elevated off of the entire columella and dissection is continued superiorly over the most medial portion of the dome of the lower cartilage on the cleft side (see fig 16) and over the entire surface of the lateral crura on the medial side of the cleft (Fig 14). The base of the ala is completely separated and freed up from the underlying maxilla and the skin is elevated off of the lateral crura of the lower cartilage from the lateral aspect, as illustrated in Fig 15. Then, with great care, the nasal mucosa is elevated from the undersurface of the lateral crura primarily from a lateral approach. This will allow the repositioning of the deformed alar cartilage on the side of the cleft and the maintenance of its position by bolsters later in the procedure, to re-drape the mucosa and skin in an appropriate position over this cartilage.

At this time a skin hook is placed at the junction of the skin of the upper portion of the columella and the lateral crura. When the skin hook is elevated superiorly, it causes the c-flap to fold on itself (Fig 12). This may now be sutured to itself with interrupted 6-0 nylon to lengthen the skin of the columella on the side of the cleft. At this time the medial flap is rotated downward to determine if the medial cleft segment has obtained adequate lengthening (Fig 16). If not, the incision beneath the base of the columella may be extended to allow greater rotation to provide additional length to the medial cleft segment. The lateral cleft segment can then be advanced until adequate lip length is obtained in the lateral cleft segment.

The previously constructed muscle flaps are then rotated downward and approximated together with two or three sutures of 4-0 Dexon (Fig 13). Following this, a 5-0 nylon stitch is placed between the high points of Cupid's bow on the medial and lateral cleft segment and tied, and it can be used as a traction suture to aid in the suturing of the lip.

A permanent deep suture of 5-0 nylon can then be placed from the most advanced of the lateral advancement flap (1' on Fig 16) to the area of the anterior nasal spine. This is a
stabilizing suture and should, at this time, provide a lip which is of adequate length and symmetry.

Once adequate lip length and symmetry is obtained, the skin is then closed with interrupted 6-0 nylon sutures. It should be noted that prior to placing the stay suture, the floor of the nose is closed with interrupted 5-0 chromic stitches from the mucosal flaps, which were obtained from the incision along the vermilion cutaneous junction extended into the floor of the nose. This reconstructs the anterior portion of the floor of the nose.

Care must be taken not to have any distortion or excessive narrowing of the nostril on the side of the cleft. If this occurs an elliptical excision of skin only is performed around the alar margin to correctly position the base of the ala (Fig 17). By excising an ellipse of only skin here, it allows anterior positioning of the usually retroplaced alar base of the lateral cleft segment. These incisions are then closed with interrupted 6-0 nylon (Fig 18).

The vermilion is then closed. If this is attenuated it may be augmented by using a primary Z-plasty or the vermilion flap still attached to the lateral segment may be de-epithelialized and inserted into a submucosal pocket created in the medial vermilion segment. The external surface of the reconstructed vermilion is closed with interrupted 6-0 nylon. The inner surface and the mucosal layer is closed with interrupted 5-0 chromic.

At the conclusion of the procedure bolsters are placed to stabilize the repositioned skin and mucosa over the lower portion of the nose and the side of the cleft. Usually two bolsters are used with one bolster going through the lateral nasal mucosa and overlying skin and through the lower lateral cartilage. A medial bolster is usually placed from the area of the apex of the inner surface at the junction of the lateral crura and the columella and extending superiorly and somewhat laterally toward the other side of the nose to correctly re-drape and reposition this reconstructed portion of the nose.

The lip repair is now complete. It should be noted that some surgeons prefer not to do any significant dissection in the lower portion of the nose and do not perform the steps of the nasal tip reconstruction at the time of the primary lip repair, other than the repositioning of the alar base. Figures 19 and 20 show the pre- and postoperative photographs of a patient who had a complete unilateral cleft repaired by the use of the rotation advancement technique.

**Surgical Technique - Triangular Flap Method of Reconstruction**

When performing the triangular flap technique of lip reconstruction the same previously described anatomic landmarks (Fig 10) are utilized. In addition, initially the distance the lip must be lengthened is determined by the difference in length between the base of the columella and the high point of Cupid's bow on the nonleft side (3-2) compared with the cleft side (5-8). The difference in these two distances is the number of millimeters that the lip must be lengthened. To compensate for scar contracture, the author always adds 2 mm to this number. When this distance, including the 2 mm added, is greater than 6 mm, the author uses a two-triangular flap technique with the inferior flep based laterally and a small superior triangular flap based medially. In the vast majority of cases, however, only the inferior triangular flap is used (Fig 21).
When the size of the triangle is 6 mm or less, the incisions are outlined with gentian violet as illustrated in Fig 21. The triangle on the lower portion of the lip is based on the lateral lip segment. It is drawn starting at the high point of Cupid's bow on the lateral segment and continued upward on the vermilion cutaneous junction for the predetermined distance (the number of millimeters the lip needs to be lengthened), and an equilateral triangle is constructed according to points 5', 10', and 11 as shown in Fig 21. Thus, the distances from 5' to 10', 10' to 11, and 11 to 5' are equal. This triangle will be inserted into a defect created by an incision of equal length from the high point of Cupid's bow on the medial cleft segment that is perpendicular to the vermilion cutaneous junction (5-10). This incision is, again, the same length as the previously determined distance necessary to adequately lengthen the lip.

If an upper triangle or second triangle is necessary, an upper equilateral triangular flap is based on the medial cleft segment near the columella. This incision starts toward the base of the columella and meets the vermilion cutaneous junction with any additional length that is required greater than 6 mm. The equilateral triangle is then constructed based on this line (points 8, 9, and 12). This triangle will be inserted into a defect created by an incision of equal length on the lateral segment located in the crease at the junction of the nasal ala with the lip (8' to 9'). Following this, the high points of Cupid's bow on the medial and lateral cleft segments (5 and 5') are tattooed with a 25-gauge needle dipped in gentian violet.

The actual incisions are then performed once the drawing of the repair is completed (Fig 22). On the medial cleft segment the incision along the vermilion cutaneous junction from 5 to 9 and extending into the floor of the nose are through the skin and underlying subcutaneous tissue and to, but not through, the underlying oral mucosa on the medial cleft segment. When an incision from point 5 through the vermilion is performed, this will create a turnover flap of mucosa based on the oral mucosal service of the medial cleft segment, which can be used for additional mucosa, if necessary, in the reconstruction of the defect. The incision from the high point of Cupid's bow (5) on the medial cleft segment is through skin only. If an upper triangular flap is necessary this incision is also through skin only.

On the lateral cleft segment a through-and-through incision along the vermilion cutaneous junction from 5' to 8', which extends into the floor of the nose, is then performed. This again creates a flap of vermilion based on the vermilion of the lateral cleft segment, which can be used to augment the vermilion of the central portion of the lip if this is deficient. The incision of the lower triangular flap from 5' to 10' to 11 is through skin only. The curvilinear incision from point 11 to the vermilion cutaneous junction is also through skin only. There is a small triangular piece of skin here that is then excised and discarded.

The skin and underlying mucosa on both the medial and lateral cleft segments are then dissected free from the underlying orbicularis oris muscle for a distance of approximately 1 cm. Following this, the attachment of the orbicularis oris muscle to the base of the columella medially and to the base of the ala laterally are then excised. As illustrated in Fig 23, the flaps of the orbicularis oris muscle are dissected free, which can then be used to reconstruct the muscular sphincter of the orbicularis muscle.

The base of the nasal ala can then be dissected free from the underlying facial skeleton on the side of the cleft. This will facilitate the correct positioning of the ala during the reconstruction, if desired by the surgeon.
The flaps to create the floor of the nose are then elevated from the lower anterior septum and from the lower anterior portion of the lateral nasal wall. The reconstructed floor of the nose is closed with interrupted 4-0 or 5-0 chromic sutures. A permanent 5-0 nylon or absorbable 4-0 Dexon suture may then be used to place the nasal ala in the desired position prior to closure of the lip. It is deeply placed from beneath the base of the ala to the area of the anterior nasal spine and tightened until the ala base is in the proper location.

The muscle flaps are then rotated inferiorly, approximated, and closed with two or three sutures of 4-0 Dexon (Fig 23).

A 5-0 nylon suture is then used to approximate the high points of Cupid's bow on the medial and lateral cleft segments and is maintained as a traction stitch. The triangular skin flaps are then transposed and inserted and held in place by a Gillies corner stitch of 6-0 nylon. The skin is then closed with multiple interrupted 6-0 nylon sutures.

Again, the vermilion is managed the same way as in the rotation advancement method of repair to create an adequate and symmetrical vermilion. The external vermilion sutures are 6-0 nylon, and 5-0 chromic is used to close the inner vermilion sutures and the mucus membrane on the undersurface of the repaired lip (Fig 24). A patient with a cleft deformity that was reconstructed by the use of the triangular flap technique is illustrated in Figs 25 and 26.

**Postoperative Care**

Postoperative care is as vital as the technique used in performing the procedure itself. Without adequate postoperative care, optimal results cannot be attained.

The incision must be kept free of crusting to prevent separation when the sutures are removed. The incision should be cleaned with 3% hydrogen peroxide, and an antibiotic ointment applied using a cotton-tip applicator twice a day. This should keep the incision itself free of crusting.

Arm restraints are used so that the infant cannot bend his arms at the elbow and are kept in place for 2 to 3 weeks following the surgical procedure. During this period the infant should be fed by the use of an ear bulb syringe rather than nursing. During the initial postoperative period the arm restraints are kept pinned to the mattress or an infant seat. However, it is not feasible to do this for the entire 3-week period. All sutures are removed at 6 to 8 days. At this time Steri-Strips may be applied to the lip for the next 6 weeks. This is done in an attempt to decrease the postoperative scarring by reducing the tension on the incision itself.

Systemic antibiotic coverage with penicillin is used intraoperatively and for a 5-day period following the procedure.

**Complications**

Severe complications are not common following primary lip repair. The most common complication is the presence of secondary lip and nasal deformities. These may include
inadequate or excessive length to the lip, notching or asymmetry of the vermilion, a short or deviated columella, a horizontal orientation of the nares, and an abnormality in size of nostril and alar base location.

Other complications include infection, wound dehiscence, and scar contracture or hypertrophy. These are uncommon and rarely seen. If hypertrophy of the scar occurs, the use of intrascar injections of steroids along with massage have eliminated this problem.

The author always advises the parents that an additional procedure may be necessary in the future to achieve the desired cosmetic result when performing any cleft lip repair. Although the majority of these patients do have an additional procedure performed at an older age, it is hoped this can be of a minor nature rather than a major complete reconstruction.

**Summary**

The techniques of repairing the unilateral cleft lip have been discussed and the two most common techniques have been presented in detail. It will appear that at times each is the optimal technique to use. Selection of the technique may well depend on the surgeon's experience and comfort in performing the procedure. In the future, careful studies are needed to attempt to delineate the parameters that will improve the selection of the proper technique in each individual patient. This would be a major benefit in the reconstruction of the unilateral cleft lip deformity.
The patient with a congenital cleft lip is one of the most challenging for the facial plastic surgeon. It is generally recognized that bilateral clefts are among the most difficult in which to achieve pleasing aesthetic and functional results; it has been said that their repair is twice as difficult as unilateral clefts and the results are half as satisfactory. For decades the specific surgical technique employed by most surgeons was the unilateral lip procedure they were using (rotation advancement, triangular flap, etc) applied to the bilateral cleft in two separate staged procedures. This approach seemed to negate one of the few advantages in many of these problem patients; that is, the deformity is frequently a symmetrical one so that performing staged repairs creates asymmetry, at least temporarily. A watershed in the treatment of bilateral clefts was Millard's presentation of his one-stage definitive repair at the international conference in Toronto in 1977 and the publication that year of his encyclopedic treatment of these deformities, Cleft Craft, The Evolution of Its Surgery. II, Bilateral and Rare Deformities. This chapter will explore this author's approach to the primary repair of the bilateral cleft, including lip, premaxilla, and nose, over the past 15 years. General principles of management, especially the difficult problem of the premaxilla, and fine points of surgical technique will be emphasized.

Treatment of Bilateral Cleft Lip - General Principles

The approach to any bilateral cleft lip will depend upon the type and spectrum of anatomic deformity, that is, complete/incomplete, symmetrical/asymmetrical, wide/narrow, and especially the status of the primary palate components (premaxilla and prolabium). The soft tissue prolabium may be diminutive, inadequate for definitive cleft closure, or large enough to provide ample forked flaps for lateral columella lengthening as well as a generous philtral segment. The premaxilla in a complete bilateral cleft, attached only to the nasal septum, may be found, as a result of the lack of normal lateral restraining forces, a centimeter or more anterior to the maxillary arch and deviated both sagittally and coronally as well. Since the premaxilla provides one of the most challenging problems of the bilateral cleft lip, it will be considered first.

The Premaxilla

Historically, the protruding premaxilla has been managed by a variety of approaches, which may be summarized as follows:

1. No direct treatment - lip adhesion or definitive repair only.
2. Presurgical orthopedics by external devices or internal prostheses, which may be active or passive.
3. Premaxillary surgical procedures involving excision or setback.
With few exceptions, our approach has been to use indirect surgical procedures, single or staged lip adhesions with the difficult premaxilla/prolabium. These procedures are described below. The advantage of this approach is that a small prolabium is enlarged, and the definitive one-stage procedure may be done under less tension. No direct trauma to bone is involved, and this conservatism should eliminate or minimize any secondary maxillary growth inhibition effects. Disadvantages include a lack of significant premaxillary retroposition with inability to close large nasolabial fistulae and the risk of additional surgical procedures. Also, there is a possibility of wound dehiscence, especially with unilateral lip adhesions.

External presurgical orthopedics using an elasting band over the premaxilla and taped to the infant's cheeks or attached to a head cap has been recommended. Problems with parent compliance and skin breakdown limit its effectiveness.

Passive prosthetic devices (HOtz) avoid direct manipulation of bony segments but require a skilled prosthodontist frequently adjusting the device. Active prostheses (Georgiade and Latham) produce rapid positioning of premaxillary/maxillary segments narrowing the alveolar ridge cleft and permitting soft tissue closure at either definitive repair or lip adhesion (Millard). These devices, however, are more complicated to use and require even more sophisticated dental expertise. They must be affixed to bone with risk to growth centers and dentition. Although preliminary longitudinal studies appear to indicate no final unexpected growth problem, further data and comparative studies would seem to be warranted.

The reservations just expressed for active prostheses apply with greater force to direct surgical attack on the premaxilla. Complete excision is mentioned, only to be condemned. The resultant severe midface retrusion ("dishface") is completely unacceptable by modern standards. Surgical repositioning requiring osteotomies of the nasal septum may be combined with bone grafting to the alveolar clefts at age 10 to 11 years, or just prior to eruption of the permanent canine teeth. Earlier surgery at or just before entering school should be regarded with caution until more long-term follow-up is available. In summary, in a severe bilateral cleft lip, a conservative, long-term approach for maxillofacial growth should be to employ soft tissue techniques such as lip adhesion or definitive repair for their orthopedic effect on the premaxilla. Secondary problems such as malocclusion and fistulae are then corrected after significant facial growth has occurred. A more aggressive plan would be to reposition the displaced segments in early infancy to childhood, providing more normal function earlier with the possibility that final facial morphology may be compromised.

**Surgical Procedures - Lip Adhesion - Bilateral**

**Indications**

Single-stage bilateral adhesion may be helpful in closing most severe bilateral clefts whenever excessive tension or the closure may compromise a one-stage definitive repair. An ideal case is shown in the Figure, where the symmetrical, complete cleft includes a premaxilla extending 1 cm beyond the maxillary arches. Other factors are the amount of tissue in and mobility of the lateral lip segments. Thinner and more restricted lips are more likely to benefit by an adhesion. Likewise, the unilateral adhesion may be used as an orthopedic technique to move a deviated premaxilla into better alignment with the maxillary arch or to achieve better
soft tissue symmetry. Occasionally, staged lip adhesions are indicated.

**Technique of Bilateral Lip Adhesion**

The bilateral lip adhesion employs the same basic principles as unilateral adhesion.

1. Landmarks for definitive repair are determined and avoided.
2. Cleft edge flaps medially and laterally are incised and elevated.
3. Intercartilaginous incisions in the nose free the ala for medial advancement.
4. Supraperiosteal dissection of the lateral lip is done only enough to allow closure.
5. Retention suture, 3-0 or 4-0 nylon, passes between prolabium and premaxilla, grasping both lateral segments at the junction of lip and nose.

6. Layered closure muscle, deep soft tissue, and skin with all deep sutures are placed prior to tying any. The result is a nasal sill/superior lip adhesion with the retention suture relieving most of the distracting forces on the closure at its area of greatest stress.

**Postoperative Care - Airway Management**

Tongue-tip suture is placed for continuous traction of the tongue, if necessary, postextubation. It is removed when the infant is fully awake and with control of his airway, usually in the recovery room.

**Feedings**

Asepto syringe feedings are begun the morning after the operation and continue for 10 days.

**Wound Protection and Care**

Arm restraints are worn for a total of 10 days and the infant is kept in a supine or side position. Antibiotic ointment is applied to the sutures two to three times a day and suture removal is at 7 to 10 days under sedation in the outpatient clinic.

**Complications**

The most serious complications directly related to the procedure is wound dehiscence. This is almost always related to excessive tension on the suture line. The incisions simply pull apart gradually. Dehiscence is much more common following unilateral lip adhesion, about 8%. In fact, this has not occurred in our series of bilateral lip adhesion using the technique described. Fortunately, if landmarks for definitive repair have not been disturbed, the deformity following dehiscence is essentially the same as the original deformity. Other minor problems associated with bilateral adhesion are some increased scar tissue between prolabium and premaxilla and increased bleeding from this area at the definitive operation. An over
wound infection, although always a possibility, has not occurred in any lip adhesion procedure in this series.

One-Stage Bilateral Cleft Lip Repair

The Figure shows a technique as described by Millard for bilateral cleft repair. This technique has a number of advantages: one-stage symmetrical lip and nasal reconstruction with orbicularis oris muscle continuity, mucosal line labial sulcus, symmetrical Cupid's bow, midline lip tubercle, philtral dimple, and bank flaps for columella lengthening at a later stage. The Figure shows a typical bilateral complete symmetrical cleft lip with landmarks and incisions indicated. The height of the lip is determined by the height of the prolabium, not by the vertical length of the lateral lip segments. If there is a marked difference in these dimensions, adjustment is made by excising excess skin in a half-crescent fashion beneath the nasal ala. The philtral flap should be made as narrow as feasible because of its tendency to stretch and widen with healing. Due to the excellent blood supply the superior extent of the philtral incisions may lie medial to the columella edge. These excisions then diverge slightly as they reach the vermillion border. The small quadrilateral shaded flap just below the vermillion cutaneous junction may be used to posteriorly reinforce the vermillion tubercle. Both lateral lip elements are handled in the same way with the cleft edge incisions, matching the prolabial height. The small flaps (a and b) of lengths 10-12 and 11-13 consist of vermillion/mucosa, submucosa, and a very small amount of muscle. They will be brought beneath the prolabial flap to reconstruct the midline vermillion. Lip and nose are freed by circumalar incisions, 6-8 and 7-9, and the lip is dissected in a supraperiosteal plane from the maxilla as necessary to allow medial advancement without excessive tension. The nasal ala may be freed by an incision in the intercartilaginous area of the nose for medial advancement. The fork flaps (c and d), consisting of skin and subcutaneous tissue, will be saved to bank later in the floor of the nose. All incisions have been made in the Figure. The vermillion flap (e) remains attached to the inferior/posterior aspect of the prolabial flap (a) with care taken not to compromise the blood supply to the flap that has been thinned posteriorly. Note that incision 10-12 and 11-13 are placed at the vermillion cutaneous junction or, at most, 0.5 mm above that. Including more skin in this flap results in an often conspicuous scar horizontally across the philtrum, especially with vertical growth of the lip. The Figure shows the prolabial flap retracted superiorly. The tips of the nasal ala have been brought medially and sutured to the periosteum in the region of the anterior nasal spine. In order to achieve the best symmetry, suturing each flap separately may be required. Excess vermillion from the prolabium is trimmed and sutured onto the anterior surface of the premaxilla to provide medial labial sulcus lining. The Figure shows the superior portions of the lateral lip flaps have been secured medially in the same fashion as the alar flaps. Again, separate suturing may be indicated for symmetry. Orbicularis oris muscles are then approximated as shown in the Figure. Skin edges are trimmed and adjusted as necessary and a philtral dimpling stitch is placed on the undersurface of the prolabial flap to the reconstructed orbicularis oris muscle. Final suturing with subcutaneous and skin stitches is applied. When there is a tendency for midline vermillion deficiency (whistle deformity), the flap (e) attached to the prolabium may be tucked beneath the vermillion flaps (a and b) to help create a midline tubercle.
Postoperative Care

A tongue stitch is applied to help control the airway until the patient is fully awake. Asepto syringe feedings are begun the morning following surgery and continued for 10 days. Elbow restraints are also used for this period of time. The patient is discharged when taking feedings well and the parents are comfortable with feeding and incision care. Antibiotic ointment is applied to the sutures 2 to 3 times each day. Sutures are removed after 6 to 7 days in the outpatient clinic under sedation.

Columella Lengthening and Bilateral Cleft Lip

One of the most obvious deformities associated with bilateral cleft lip is a shortened columella. In general, the more severe the cleft lip, the more deficient is the columella, 1 to 2 mm in length in these cases. A relatively mild bilateral cleft may have only a slightly deficient columella, thus not requiring a secondary lengthening procedure. An advantage of Millard's single-stage lip closure is the provision of prolabial tissue in the form of forked flaps that are saved ("banked") for columella lengthening. These flaps are placed just below the reconstructed nasal sill in the so-called whisker position or in the medial portion of the sill itself in the "praying hands" position. When the flaps are elevated and sutured together to form a new columella, the lip scars are not disturbed unless it is decided to revise poor philtral scars or correct secondary deformities such as a markedly wide philtrum. Then the forked flaps are taken from the philtral sides, narrowing the philtrum and creating fresh scars. Timing of this procedure may range from when the patient is 2.5 years old to just before he or she enters school. Before describing the technique in detail several other methods have been described: (a) a single, midline philtral flap (Burdach), which produces a midline vertical scar and is contraindicated when the upper lip is already tight; (b) medial rotation of the nasal sills (Cronin), useful if the sill is quite wide; and (c) medial rotation of the superior nostril margins by a gull wing-type incision of the nasal tip, which produces a more obvious scar but may be advantageous in the Asican cleft nose.

Procedure for Columella Lengthening by Banked Forked Flaps

At the primary bilateral cheiloplasty, full-thickness flaps are created from the philtral sides by narrowing the philtrum to appropriate dimensions (see above). These flaps consist of skin and subcutaneous tissue only since there is no muscle in the prolabium of complete bilateral clefts. A narrow strip of vermilion may be included with the flaps, but as this produces a more obvious scar, it is usually not done. The forked flaps are rotated 90° so that they line the medial portion of the nasal sill. At 2 to 4 years of age the patient is ready for columella lengthening. An endotracheal tube is secured in the midline without disturbing the lip. No local anesthetic is used. The banked flaps are outlined with gentian violet. Medially the incisions meet in a peak at the midcolumella slightly above its junction with the lip. The inner incision is carried medially and then superiorly along the shortened membranous septum to the nasal dome and a 2 to 3 mm back cut is marked parallel to the dorsum of the nose. The external excision is made first to insure symmetry. Thick, forked flaps are elevated by sharp dissection to the columella. Bleeding is brisk but is easily controlled with electrocautery. The membranous septum incisions are made carefully on each side, and with scissors the columella with attached flaps are elevated to the nasal tip. Blunt scissors dissection over the alar domes is done next, aided by the small back cut at the domes.
Forked flaps are then sutured together beginning at the apex with 6-0 or 7-0 nylon in the skin. Any discrepancy in length may be adjusted by trimming at this point. Exceptionally thick flaps may be carefully thinned posteriorly and two or three subcutaneous absorbable sutures are placed to strengthen the new columella. The columella tip is placed at the anterior nasal spine area of the premaxilla and a 5-0 or 6-0 chromic suture is placed from the dome incision to a point as high as possible along the membranous side of the lengthened columella. Suturing continues inferiorly, alternating from side to side to maintain symmetrical midline position of the columella. At the tip, a deep suture (4-0 or 5-0 Vicryl) is placed to the anterior nasal spine so that the lip columella angle is fixed, usually best when made just slightly over 90°. Failure to place this suture correctly results in a widened, overly obtuse columella angle. Care must be taken also not to compromise the columella blood supply by compressing the flaps too much in this closure. It may be necessary to remove sutures from the membranous septal incision to relieve tension.

After setting the nasal labial angle, there is a gaping defect between lip and nose. The alar bases may be freed by circumalar and intercartilaginous incision if necessary and sutured more medially to the columella base/nasal spine. This produces a difference in length between lip and nasal incision margins that may be compensated for by placing deep sutures by a halving technique. First the midline is approximated, and the remaining lateral defects closed with deep 3-0 or 4-0 Vicryl. Finally, skin is sutured with nylon. The resulting convexity of the upper lip smooths out with healing.

**Postoperative Care**

Routine suture care is used as described for lip repair. In general, arm restraints are not required in the older patients. A regular diet is begun the day after operation. More time is necessary for healing, so sutures are left in place for 10 to 12 days and removed either under heavy sedation in the clinic or by light general anesthetic in the operating room.

**Complications**

Superficial infection, especially where columellar incision and infranasal incisions meet, is common, probably due to bathing the area in nasal secretions in young children. Invasive wound infections, however, are rare, even in the relatively poorly vascular forked flaps. Loss of columella length with healing is variable and unpredictable. Some decrease in projection is inevitable and shrinkage of one-third is fairly common. "Migration" of the columella inferiorly to produce an excessively obtuse nasolabial angle occurs only if the columellar base has not been firmly sutured to the premaxillary periosteum at the desired angle.

Final correction at the nasal tip deformity is carried out in adolescence by an open rhinoplasty technique employing cartilage strut grafts to achieve full tip projection.
Pediatric Facial Plastic and Reconstructive Surgery

James D Smith, Robert M Bumsted

Chapter 12: Cleft Palate

Craig W Senders and Jonathan M Sykes

Historical Perspectives

A French dentist named Le Monnier Rouen was the first to attempt closure of a congenital cleft palate in the mid-18th century. The technique involved cauterization of the edges of the cleft and holding the edges together with suture. Presumably this was carried out on cleft of the soft palate only.

Attempts at closing the hard palate were not successful until the 19th century when the importance of reducing tension was discovered. In 1928, Dieffenbach recommended separating the oral mucosa from the bone of the hard palate as a method of closing the palate. Additionally, he used lateral osteotomies to aid in the closure. The importance of utilizing relaxing mucosal incisions to reduce tension, became popularized in the mid-19th century by Pancoast (1843), Warren (1828, 1843) and Fergusson (1845).

The 20th century has brought about refinements in techniques, with attention to improving function. In 1937, Wardill and Kilner independently described a technique to lengthen the soft palate involving V-to-Y advancement of palatal mucosa. This procedure is limited by the difficulty of providing a similar advancement of the nasal layer. Additionally, this technique requires leaving large areas of exposed palatal bone, which may inhibit future palatal growth. Attempts at increasing the length of the soft palate without inducing scarring in the hard palate resulted in numerous modifications, from using buccal or nasal flaps to skin grafts. A definitive improvement in velopharyngeal function has not been found.

In the 1930s, Veau was the first to focus upon the anatomy in clefts. Later, Kriens developed the intravelar veloplasty. This technique releases the attachments of the malpositioned tensor veli palatini and levator veli palatini muscles from the posterior portions of the hard palate and realigns these muscles. Retrospective studies based upon historical controls have shown improved velopharyngeal function. The only prospective randomized study found no improvement in function. Although realigning the malpositioned palatal muscles has a strong theoretical advantage, the scarring induced by isolation of the muscle may limit palatal function.

In a further attempt to improve function of the soft palate after repair, Furlow developed the double reversing Z-plasty palatoplasty. This technique utilizes lengthening of the palate by a Z-plasty with realignment of the palatal musculature. There is less muscle dissection than in the intravelar veloplasty. This may result in less scarring and improved function. Unfortunately, the only published studies are retrospective and based upon historical controls.
Another approach popularized by Schweckendiek is a two-stage repair of the palatal defect. In the first stage, only the soft palate is closed. The hard palate cleft narrows after soft palate repair. Typically, the hard palate is then closed at 5 to 6 years of age. This allows the benefit of early soft palate movement and function and minimizes the detrimental effects on facial growth of the hard palate repair. It is paramount to this technique to use a palatal obturator in the interval prior to hard palate repair. This requires a talented, interested, and dedicated prosthodontist. At most centers this is not practical.

At the present time, the most popular repairs are single-stage repairs, although there has been a resurgence of interest in the Schweckendiek-type procedure in a number of centers across the country. The Wardill-Kilner V-to-Y advancement is used at a number of centers, although studies have not shown this to improve velopharyngeal function. Efforts at reestablishing the muscular continuity via the intravelar veloplasty or the Furlow palatoplasty have gained significant popularity, although improvement of velopharyngeal function has not been absolutely determined.

As we approach the 21st century there are four basic principles of palatoplasty: (a) multiple layer closure, (b) elimination of tension at the suture line, (c) reconstruction to allow improved velopharyngeal function, and (d) minimization of scarring of the hard palate to limit inhibition of midfacial growth.

Anatomy

The hard palate is divided embryologically into the primary and secondary palate. The primary palate forms at the time of lip formation between the 4th and 6th week of gestation. The secondary palate forms between 7th and 12th week. The medial and lateral incisor teeth develop from the primary palate, the remainder of the teeth develop from the maxilla.

A cleft of the lip can result in a cleft alveolus and a cleft between the primary and secondary palate. Additionally, this cleft between the primary and secondary palate can inhibit the fusion of the palatal shelves, from anterior to posterior. Thereby a cleft of the lip can result in a complete cleft of the lip and palate.

In a complete bilateral cleft lip, the primary palate (often referred to as the premaxilla) is not fused with the secondary palate and is attached to the vomer. In severe cases the premaxilla is displaced anteriorly and the lateral alveolar segments are collapsed. In this situation the vomer is not attached to either palatal shelf. However, variations can occur where the vomer is attached to one or the other palatal shelf.

A cleft of the secondary palate occurs between the 7th and 12th week of gestation. This is a different embryological malformation than in a cleft lip or a cleft lip with a cleft palate. The expression is variable. A submucous cleft palate is a mild expression. In other cases the cleft will involve the entire secondary palate. In addition, this cleft can be bilateral, with the vomer unattached to either palatal shelf. Typically, the vomer is attached to one or the other palatal shelf.

The major blood supply of the hard palate is from the greater palatine artery. This artery passes through the greater palatine foramen, which is located next to the second molar
tooth. Additional sources of the blood supply of the hard palate come from the incisive artery as well as the lesser palatine artery and the ascending pharyngeal artery.

The soft palate is composed of five paired muscles: tensor veli palatini muscle (TVPM), levator veli palatini muscle (LVPM), palatoglossus muscle, palatopharyngeus muscle, and musculus uvulae. Both the TVPM and LVPM arise from the base of the skull and a portion from the auditory tube. A portion of the TVPM arises from the lateral portion of the auditory tube. It is thought that the TVPM opens the eustachian tube. The TVPM passes anterior to the hamulus of the pterygoid plate where it makes a 90° turn to become the palatal aponeurosis. The LVPM arises from the medial aspect of the base of skull and attaches to the medial aspect of the auditory tube. It travels anteriorly into the soft palate and interdigitates with the LVPM of the opposite side. This muscle is important in elevating the soft palate and providing velopharyngeal closure. It is thought that the LVPM and the salpingopharyngeus muscles have no effect on auditory tube opening, because of their anatomic origin.

The palatoglossus forms the anterior tonsillar pillar and inserts into the posterior portion of the soft palate. The palatopharyngeus is a large muscle that forms from the lateral portion of the pharynx and makes up the posterior tonsillar pillar. In the soft palate it forms two fasciculi, divided by the LVPM. The anterior fasciculus inserts into the palatal aponeurosis. The posterior fasciculus interdigitates with the opposite posterior fasciculus, posterior to the LVPM. A few fibers of the posterior fasciculus pass nasally over the LVPM to join the anterior fasciculus. The musculus uvulae is a small muscle that attaches to the posterior nasal spine and inserts into the uvula of the soft palate. It can play an important role in velopharyngeal closure.

A cleft palate distorts the musculature of the soft palate. Instead of the LVPM and the palatopharyngeus muscle interdigitating with the opposite muscle, these fibers tend to assume a more anterior projection and insert along the margin of the cleft and into the posterior portion of the hard palate. In a palate repair that does not realign the muscles, it may be difficult for these muscles to function normally.

**Indications for Surgery**

The most important reason to repair a cleft palate is to allow the patient to develop normal speech. Without repair of this large oronasal fistula, the patient is condemned to a life of poor communication. Unfortunately, hypernasal speech is less well tolerated than hyponasal speech.

Surgical repair of a cleft palate also serves to improve swallowing. However, by the time most patients have their palatal surgery, adaptation to the cleft has occurred and daily nourishment is no longer an issue. In carefully selected cases, a palatoplasty is performed primarily to improve swallowing in a child with developmental disabilities.

Auditory tube dysfunction with resultant chronic otitis media is highly prevalent in patients with cleft palate. To date, studies have not demonstrated that repair of the cleft palate results in improved auditory tube function. It is unknown whether this is due to an abnormal origin of the muscle in relation to the auditory tube or an inability to improve the function.
of the muscle.

Timing of the Surgical Repair

Palatoplasty is performed on patients as young as 3 months of age and as old as 2 years of age. At the majority of centers the palatal repair occurs between 9 and 18 months of age. Those who perform the repair on 3-month-old patients generally utilize a two-stage repair. The soft palate is repaired first, and the hard palate is repaired at a second stage.

The theoretical benefit of repairing the palate early is improved muscular function and ultimately improved speech. Many believe that having an intact soft palate during swallowing, prior to the development of speech, will improve function during speech. However, palatal surgery is more difficult on younger patients and the risks of a blood transfusion increase.

There are no prospective randomized studies to compare early and late palatal closure. However, there are several retrospective studies that suggest there may be a benefit to closing the palate prior to 18 months of age. Overall there is a trend toward earlier palatal closure.

In over 150 palatoplasties that have been performed at the University of California (UC)-Davis, no patient has required a transfusion. An added benefit is that the older child does not require feeding by bottle, so the postoperative repair will not suffer additional trauma.

Preoperative Evaluation and Preparation

The greatest concern at the preoperative evaluation is whether the mandible has grown to an adequate size to minimize the risk for postoperative airway obstruction. In the UC Davis experience, approximately 1% to 2% of the patients have required reintubation for postoperative airway obstruction. In a patient with Pierre Robin sequence, if the mandible does not demonstrate catch-up growth, delaying surgery until 24 months of age is appropriate.

Many centers allow bottle feeding immediately following palatoplasty. The UC Davis craniofacial anomalies team has been slow to follow this trend as randomized studies have not been completed.

A child that is not above the 5th percentile for weight and height can be anticipated to have a complicated postoperative course. Typically these patients are neurologically delayed and a delay in the surgery will not further compromise their speech development. However, chronic airway obstruction and cardiac anomalies must be ruled out in these failure-to-thrive patients.

Surgical Technique

Von Langenbeck Procedure

The von Langenbeck procedure is a relatively popular procedure that was introduced in 1859. The procedure is optimally suited for clefts of the secondary palate only, but can be performed on complete clefts if the anterior portion of the anterior palate is not closed. The
The technique involves a releasing incision laterally, leaving a bipedicle flap to provide blood supply to the palatal segments. This procedure does not add any length to the soft palate.

The edges of the cleft are incised down to bone over the hard palate and extend into the soft palate all the way to the tip of the uvula. Relieving incisions are made just medial to the alveolus and extend around the maxillary tuberosity just lateral to the hamulus. Large mucoperiosteal flaps are elevated on the hard palate. Care is taken not to damage the greater palatine artery as it exits the greater palatine foramen. The soft palate muscular attachments to the hard palate are separated and blunt dissection is utilized along the posterior portion of the releasing incision. The mucoperiosteum is elevated off of the nasal layer. The incision is closed in three layers with the nasal layer being closed from anterior to posterior using 4-0 chromic suture. The palatal musculature is closed with 3-0 chromic suture or 4-0 synthetic absorbable suture. The hard palate is closed with 3-0 chromic suture or a 4-0 synthetic absorbable suture in a vertical mattress fashion. To eliminate the dead space over the hard palate, the nasal layer can be included in two of the oral mucosal sutures.

Three-Flap Palatoplasty

This is the preferred technique of the UC Davis craniofacial anomalies team for closure of clefts of the secondary palate. The three-flap palatoplasty can be utilized to lengthen the soft palate using a V-to-Y technique (Wardill-Kilner operation). However, the V-to-Y technique leaves more bone exposed. Subsequent scarring of the hard palate will cause more problems with midfacial growth and is not worth the unproven benefit of lengthening the soft palate.

This procedure involves an incision along the cleft, extending down to the bone over the hard palate and dividing the border of the cleft of the soft palate up to the base of the uvula, the sides of which are excised. Both incision must join each other anteriorly. The lateral incision is very similar to the von Langenbeck lateral releasing incision. An incision is made just lateral to the hamulus around the maxillary tuberosity, medial to the alveolus, to approximately the level of the canine tooth. An incision is then made from the apex of the cleft to the canine tooth. Incision and complete elevation is performed on one side prior to the opposite to facilitate better hemostasis.

Using a periosteal elevator the mucoperiosteum is elevated, mobilizing the large palatal mucoperiosteal flap. Care is taken not to damage the greater palatine artery as it exists its foramen. Kitners are utilized to provide blunt dissection on both sides of the greater palatine artery and improve the mobilization. This often better defines the posterior palatal shelf. Using Padgett elevators, the muscle attachments to the posterior palatal shelf are dissected free and the nasal mucosa is elevated. Blunt dissection around the hamulus with a Kitner as well as complete dissection around the vascular pedicle usually will allow the flaps to meet easily in the midline. However, if there is tension, the greater palatine vessels can be sharply dissected from the mucoperiosteal flap using a right-angle Beaver blade for several millimeters. Rarely, it is necessary to dissect the tendon of the tensor veli palatini muscle from around the hamulus or fracture the hamulus to provide greater flap mobility. Closure of the nasal layer is performed in an anterior-to-posterior direction with 4-0 chromic suture.
The muscular layer is then closed with 3-0 chromic or 4-0 synthetic absorbable suture with two or three large bites. The oral mucosa is then closed using interrupted vertical mattress sutures of 3-0 chromic or 4-0 synthetic absorbable suture from posterior to anterior. The dead space over the hard palate can be eliminated by including the nasal layer in two of the oral mucosal sutures. Typically the lateral incisions can also be closed unless the cleft is quite large. The saloon door effect as described by Bardach and Nosal explains this surgical mystery. Because the palatal vault typically has a significant vertical height, this adds increased width to the palatal flaps. At the time of closure the flaps assume a more horizontal orientation and effectively leave very little exposed bone. If bone is left exposed laterally it should be covered with an absorbable hemostatic agent.

**Two-Flap Palatoplasty**

A two-flap palatoplasty is utilized for unilateral complete clefts of the lip and palate. This technique is very similar to the three-flap palatoplasty technique with modified palatal incisions. Along the edge of the cleft, an incision to bone is made over the hard palate and extending into the soft palate at the midpoint between the oral and nasal layers. This incision is carried to the base of the uvula, the sides of which are excised. A lateral incision is made just lateral to the hamulus, passing around the maxillary tuberosity. The incision is made from posterior to anterior just medial to the alveolus until it joins the previously made cleft incision. On the nonleft side, the lateral incision is carried past the canine tooth, to the opposite canine where it joins the cleft incision. Mucoperiosteal flaps are elevated as in the three-flap palatoplasty. Again, attention is paid to eliminating tension on the closure line and allowing a three-layer closure over the soft palate and a two-layer closure over the hard palate. Care is taken to eliminate the dead space over the hard palate by including the nasal layer with the deep portion of the vertical mattress suture. Any large areas of exposed bone are covered with an absorbable hemostatic agent.

**Intravelar Veloplasty**

The technique of intravelar veloplasty was described by Kriens in 1969 and 1970. This technique gained popularity in the 1970s and 1980s. Dreyer and Trier in a retrospective study using historical controls, showed the need for a pharyngeal flap dropping from 35% to 9% when using the intravelar veloplasty. However, Marsh et al in 1986 reported in a randomized prospective study that there was no statistical difference between the two groups. This suggests that the increased scarring caused by dissection and plication of the levator veli palatini muscle inhibits the function, despite its new anatomical relationship.

The technique of intravelar veloplasty can be combined with the three-flap palatoplasty, two-flap palatoplasty, or von Langenbeck procedure. All of the mucosal incisions and elevations are the same, except that in the intravelar veloplasty attention is taken to completely mobilize the levator veli palatini muscle as well as the aponeurosis of the tensor veli palatini muscle. This involves sharp dissection for approximately 1 cm from the cleft margin. Care is then taken to approximate or plicate the muscles of the soft palate to provide better function.
Double Reversing Z-Plasty

The double reversing Z-plasty was described by Millard in 1978 at the annual meeting of the Southeastern Society of Plastic and Reconstructive Surgeons. However, it was not until 1986 that studies utilizing this technique were published. This technique utilizes a Z-plasty to lengthen the soft palate, and it realigns the levator veli palatini muscle. Because the muscle is dissected on one surface only, there should be less fibrosis than in the intravelar veloplasty.

As originally described, the procedure combines the palatal lengthening of a Z-plasty and realignment of the palatal muscles with a closure of the hard palate under tension. Thus, this procedure has taken two steps forward and one step backward. The procedure is excellent for clefts of the soft palate.

The procedure involves making a Z-plasty on both the oral and nasal surface in a reversed pattern from one another. Because of the difficulty in elevating the levator veli palatini muscle from the nasal mucosa, right- and left-handed surgeons would perform the operation in mirror image. The illustrations describe the procedure for a right-handed surgeon.

An incision is made along the margin of the cleft to the base of the uvula, the sides of which are excised. A lateral incision is then made from the apex of the cleft to the hamulus on the left side. This incision is carried down to the nasal mucosa. Care is taken to try and include all of the soft palate musculature and most of the palatal aponeurosis in this flap. Using a right-angled Beaver (R) blade, the nasal mucosa is separated from the palatal musculature. Laterally, blunt dissection using scissors is most optimal. Using right-angled scissors an incision is made in the nasal mucosa from the base of the uvula to within 1 cm of the hamulus.

On the right side an incision is made through the oral mucosa from the base of the uvula toward the hamulus. This incision is carried through the epithelium and the fibrous glandular tissue overlying the palatal musculature. This flap is relatively thick as the musculature is intimately associated with the nasal mucosa. This flap is elevated sharply to the hard palate, although care is taken not to damage the greater palatine vessels. Using a Padgett elevator, the palatal musculature is dissected from the posterior border of the palatal shelves. Using right-angled scissors or a #15 blade, an incision is made from just posterior to the hamulus to the apex of the cleft. Care is taken to save 1 or 2 mm of nasal mucosa anteriorly to allow suturing. This incision separates the right palatal musculature from the hard palate. The Z-plasty limbs are transposed and the nasal layer is closed with a 4-0 chromic suture. If there is significant tension, the lateral portion of these incisions can be closed upon itself rather than stretching the Z-plasty flaps to the apex. The left palatal musculature on the left is sutured to the right lateral soft tissue (palatopharyngeus muscle) with a 3-0 chromic suture. The Z-plasty flaps on the oral layer are then transposed. Again, the palatal musculature is sutured to the palatopharyngeus of the opposite side with a deep suture. The oral layer is closed with a 3-0 chromic or 4-0 synthetic absorbable suture in an interrupted fashion. If there is tension laterally, the incision is closed upon itself rather than pulling the tip of the flap into the apex. Care must be taken at the original apex of the cleft to insure that there is a good two-layer tension-free closure.
Postoperative Care

Typical postoperative care involves a pureed diet, per cup only, for 3 weeks. All patients are placed in arm restraints. A mist tent rather than a mist hood is utilized to markedly increase oral hydration. This is particularly important during the first day as clots in the pharynx can become dry and result in upper airway obstruction. The mist tent is discontinued when oral intake becomes regular. Oxygen saturation is routinely monitored. The patients are discharged from the hospital when they are taking 90% of the calculated maintenance fluid for their size.

Complications

Bleeding

Blood loss can be minimized by the careful use of a vasoconstrictive agent and allowing time for the vasoconstriction agent to take effect. If significant bleeding is encountered during elevation of flaps, pressure is applied for several minutes. After the flaps have been elevated, it is important to search carefully for bleeding points, as an uncontrolled bleeder resulting in only 2 cc of blood loss per minute would result in 240 cc of blood loss over a 2-hour period. For most children this would require a blood transfusion.

Postoperative bleeding of any significance requires a return to the operating room to determine the source.

Airway Obstruction

Some degree of airway obstruction following palatoplasty is relatively common. Significant airway compromise may be due to tongue swelling, laryngeal edema, pharyngeal secretions, or prolonged action of anesthetic agents. Tongue swelling has been thought to be due to ischemia induced by the mouth gag. Therefore, it is prudent to release the mouth gag periodically during the procedure. Additionally, continuous suspension of the mouth gag from the Mayo stand is not recommended.

Significant airway obstruction requires treatment. In past years a suture was placed in the tongue and a resident or nurse held the tongue out to improve the airway in troublesome cases. However, in this day and age patients with significant airway obstruction should be reintubated.

Palatal Dehiscence

A palatal dehiscence due to poor blood supply infection, poor blood supply, and/or poor suturing technique is probably best treated by observation and reoperation when scarring becomes mature. In fact, looking at the palatal incision in the postoperative period, has been termed the "evil eye". This is because there is little benefit in diagnosing a dehiscence. Looking with a tongue blade in an uncooperative patient may damage the repair. A small dehiscence often will heal on its own without sequelae. A slight separation of the uvula is seen relatively frequently and has no functional consequences. If a dehiscence is caused by a trauma, such as falling onto a hard object, then immediate repair may be beneficial.
Oronasal Fistulae

Fortunately, with proper care, oronasal fistulae are quite unusual. Additionally, many fistulae are quite small and are of no functional significance. Often, on a complete cleft of the lip and palate, there is a small oronasal fistula just posterior to the alveolar cleft. In essence, this is a continuation of the alveolar cleft. This should be repaired at the time of alveolar bone grafting, which is typically between 5 and 11 years of age.

A soft palate fistula typically can be excised and closed in three layers. However, a fistula of the hard palate requires a larger surgery. A fistula here is best treated with relatively large mucoperiosteal flaps and closed in two layers. Often it is necessary to almost repeat the two- or three-flap palatoplasty to allow a tension-free closure. Closures that involve lesser procedures often fail in the long run.
One of the major goals of cleft palate repair is to create a velopharyngeal mechanism that is capable of separating the oral and nasal cavities during speech. When symptoms of velopharyngeal inadequacy are observed, most surgeons view this as a surgical failure requiring secondary surgical management. This chapter presents a broader view of the problem and illustrates that not all symptoms of velopharyngeal inadequacy are the result of the absence of adequate tissue and length. The need for differential diagnosis is emphasized as the first and most critical step in management planning. Diagnostic evaluation methods are discussed and suggestions are presented for determining which intervention strategy is appropriate, ie, speech therapy, prosthetic management, or surgical intervention. Several surgical approaches are discussed along with the issues of patient selection, surgical technique, complications, and postoperative care.

Most otolaryngologists-head and neck surgeons traditionally have seen their involvement with the evaluation and management of the velopharynx in children as limited to those surgeons who were directly involved in the care of patients with cleft palate. On the contrary, head and neck surgeons who see children in routine clinical practice will be asked several times in their careers for an opinion concerning the competence of the velopharynx for speech. The opinion given (whether right or wrong) is likely to have profound ramifications for each patient.

Velopharyngeal symptoms may have a variety of etiologies including over cleft palate, submucous cleft, neuromotor impairment, interference from other vocal tract structures (particularly the palatine tonsils), and mislearning. It is particularly important for the clinician to know that velopharyngeal symptoms do not occur solely as the result of overt or submucous cleft palate. For example, velopharyngeal inadequacy that may appear to occur in isolation of any obvious orofacial anomaly, may in fact be the single presenting symptom that can point to a multiple malformation syndrome. For example, Jones studied 428 patients who presented to a large cleft program at Children's Hospital and Health Center, San Diego. She showed that of the 428 patients, 75% of those who presented with velopharyngeal inadequacy as the primary symptom were later shown to have multiple malformation syndromes. These data suggest that the head and neck surgeon should be particularly thoughtful and thorough in the evaluation of a patient who presents with velopharyngeal symptoms. Although the concept that velopharyngeal symptoms may point to a more global disorder may seem academic for the physician in routine clinical practice, it is not. For example, velocardio-facial syndrome (VCF) is a recently delineated congenital malformation syndrome first identified by Shprintzen et al. It can be associated with clefting, cardiac anomalies, characteristic facies, learning disabilities, and speech disorders. Frequently, these children first present in pediatric or otolaryngology offices with complaints of velopharyngeal symptoms. Positive identification of this syndrome is particularly crucial for any surgical considerations. Specifically, many of these children have medially displaced internal carotid
arteries, which can be an obvious contraindication for many surgical procedures and therefore of great importance and interest for surgeons, especially head and neck surgeons who may be called upon to perform tonsillectomy, adenoidectomy, or velopharyngeal surgery to remediate the presenting symptoms.

Although the focus of this chapter is on the evaluation and management of the velopharynx, it must be understood from the outset that the velopharynx does not function in isolation and cannot be evaluated separately from the entire vocal tract and communication process. Rather, disturbances in anatomy and function that have the potential for affecting velopharyngeal function are far more complex and interrelated than has been acknowledged historically.

The Velopharynx Revisited

For clinical purposes, the speech production mechanism may be thought of as a large air-filled container always closed at the bottom with two openings to the atmosphere, the lips and the nares. Within the container, there are several valves that can be opened or closed to varying degrees, thus changing the shape of the container and the resistance to airflow. All of these valves must move in a highly coordinated manner creating a series of rapidly changing air pressures and airflows, which we ultimately perceive as the sounds of speech.

As viewed quite simplistically, the role of the velopharynx is to separate the oral and nasal cavities during speech and swallowing. For speech, the velopharynx directs air from the lungs and larynx through the mouth for oral sounds and through the nose for nasal sounds. When this valving is disturbed, speech can be affected in a number of ways including hypernasal resonance, nasal air emission, hyponasal resonance, nasal substitutions, compensatory articulation, sibilant distortion, and increased risk for disorders of phonation, ie, laryngeal voice disorders. Table 1 presents definitions of several speech symptoms associated with cleft palate and/or velopharyngeal dysfunction.

Table 1. Definitions of resonance, articulation, and phonation disorders frequently associated with cleft palate and/or velopharyngeal dysfunction

Hypernasality: The perception of inordinate nasal resonance during the production of vowels. This results from inappropriate coupling of the oral and nasal cavities. (The term inordinate is used to allude to the fact that low vowels and vowels in nasal consonant contexts are normally somewhat nasalized.)

Nasal emission: Nasal air escape associated with production of consonants requiring high oral pressure. It occurs when air is forced through an incompletely closed velopharyngeal port. Nasal emission may be audible or not. (Note: Hypernasality and nasal emission are not synonymous although they often occur together and are both symptoms of velopharyngeal dysfunction.)

Hyponasality: A reduction in normal nasal resonance usually resulting from blockage or partial blockage of the nasal airway by any number of causes including upper respiratory
infection, hypertrophied turbinate or hypertrophied adenoids, a wide, obstructing pharyngeal flap.

**Hyper-hyponasality:** The simultaneous occurrence of hypernasality and hyponasality in the same speaker usually as the result of incomplete velopharyngeal closure in the presence of high nasal resistance, which is not sufficient to block nasal resonance completely.

**Cul-de-sac resonance:** A variation of hyponasality usually associated with tight anterior nasal constriction often resulting in a "muffled" quality.

**Nasal substitution:** The articulators are placed appropriately for an intended oral consonant. However, incomplete velopharyngeal closure causes the sound to be produced as a nasal consonant. For example, "b" becomes "m" and "d" becomes "n". Such substitutions frequently are called "homorganic nasals".

**Compensatory articulation:** The articulators are placed inappropriately so as to enable creation of the plosive or fricative characteristics of the sounds they replace. For example, if a patient cannot build up oral pressure for the fricatives (eg, "s") or plosives (eg, "p") because of velopharyngeal dysfunction, they may create those pressures below the level of the velopharyngeal port. Such substitutes include glottal stops, pharyngeal stops, and pharyngeal fricatives among others.

**Sibilant distortion:** Inappropriate tongue placement for the sounds "s" and "z".

**Laryngeal/voice symptoms:** A variety of phonation disorders may accompany velopharyngeal dysfunction including hoarseness, low speaking volume, strained or strangled voice quality, and unusual pitch alterations. The most recent theory for the co-occurrence of velopharyngeal and laryngeal symptoms is that speakers with velopharyngeal dysfunction may attempt to compensate for the inability to achieve complete closure and maintain adequate speech pressures by compensatory activity at the level of the larynx.

Historically, the velopharynx has been viewed as a simple binary valve with two possible positions, open or closed. However, research and clinical observation over time have shown that the velopharynx is a complex three-dimensional valve with a variety of shapes and patterns of activity that vary between speakers. Just as the lips and tongue are considered speech articulators, so too is the velopharynx, as it takes different positions or shapes for different sounds. Furthermore, it is not enough that the velopharynx be capable of achieving complete closure of the valve, but it must do so in a tightly controlled time domain in coordination with other articulators.

**Terminology: More Than a "Mere" Semantic Difference**

Traditionally, when symptoms have been present that suggested that the velopharynx was not functioning correctly, we referred to the problem as "velopharyngeal incompetence" (VPI). However, hearing hypernasality or nasal emission (ie, symptoms associated with VPI) does not necessarily indicate that the velopharynx cannot work. These symptoms merely indicate that the velopharynx is not functioning at that time. Wendel Johnson, one of the
fathers of modern speech pathology and a great semanticist, explored the relationship between language and science. He taught us that the way we talk about a topic influence the way we think about it. Therefore, when a clinician hears hypernasality or nasal emission, i.e., symptoms of VPI, and in turn makes a diagnosis of "VPI" the semantic label he or she has used can have far-reaching implications. For example, the diagnosis of VPI suggests that the velopharyngeal mechanism cannot achieve closure. For most clinicians this implies that only physical management (either surgical or prosthetic) will correct the problem. Additionally, such a label may bias the inexperienced speech pathologist to discontinue speech therapy on the grounds that additional therapy will be of no value until physical management is completed. Furthermore, if the diagnosis is incorrect, any surgical or prosthetic attempts to manage the problem will be of little value. In this instance the clinician has used imprecise diagnostic labels, which can unintentionally lead to erroneous assumptions and inappropriate actions.

Previous authors have made recommendations for standardizing the nomenclature. For example, Loney and Bloem reviewed the literature and found neither consensus nor precise definitions of the terms velopharyngeal incompetence, velopharyngeal inadequacy, or velopharyngeal insufficiency. They found that authors frequently used all three terms interchangeably or one term to describe all types of velopharyngeal malfunction.

In a response to Loney and Bloem, Trost-Cardamone suggested a taxonomy for velopharyngeal disorders based on etiology. She states:

Impaired velopharyngeal closure for speech can result from a variety of etiologies. Moreover, there are perceptual speech characteristics that are pathognomonic of velopharyngeal impairment and that can distinguish among certain subtypes of velopharyngeal function problems. In both diagnosis and treatment, it is necessary for the clinician to have a taxonomic system for reference, which should serve to relate etiology to deviant velopharyngeal and speech production patterns. This is especially important because the nature of the velopharyngeal function disorder allows for certain treatment alternatives and excludes others.

Trost-Cardamone suggests the following taxonomy:

Velopharyngeal inadequacy: the generic term used to denote any type of abnormal velopharyngeal function. Within the broad group of inadequacies, there are subgroups of structural, neurogenic, and mislearning or functional origins.

Under the broad classification of velopharyngeal inadequacy she delineates three etiologic categories: velopharyngeal insufficiency, velopharyngeal incompetence, and velopharyngeal mislearning. Each of these is defined as follows:

Velopharyngeal insufficiency: Includes any structural defect of the velum or pharyngeal wall at the level of the nasopharynx; there is not enough tissue to
accomplish closure, or there is some type of mechanical interference to closure. Most often these problems are congenital.

*Velopharyngeal incompetence:* Includes neurogenic etiologies that result in impaired motor control or impaired motor programming of the velopharynx... Motor control disorders can cause partial or total paresis of the soft palate and pharyngeal walls. Depending upon the nature, level, and focus of the nervous system lesion, velopharyngeal incompetence often disturbs velopharyngeal closure for protective and reflexive acts of gagging and swallowing, as well as for speech.

*Velopharyngeal mislearning:* Includes etiologies that are not caused by structural defects or by neuromotor pathologies of the velopharyngeal complex.

In this latter category of velopharyngeal mislearning, Trost-Cardamone includes soundspecific nasal emission and velopharyngeal symptoms associated with deafness or hearing impairment.

The use of standardized, precisely defined terms is one of the prerequisites to scientific investigation. However, the use of clearly defined nomenclature is also a critical factor in clinical practice where unclear terminology may lead to inappropriate assumptions and inappropriate management. Loney and Bloem and Trost-Cardamone should be congratulated for their discussion of the lack of standardized terminology and for their attempt to define terms. However, in clinical practice the delineation they propose often may be too cumbersome for daily use and may not accomplish their goal of "improved interprofessional communication". Trost-Cardamone's system offers an important, meaningful classification of velopharyngeal inadequacies based on etiology, which is missing in the literature. It is sound and useful, especially once differential diagnosis is known. However, accurate use of the levels of definition frequently requires diagnostic information that may not be available at the first presentation of velopharyngeal symptoms.

Therefore, a simpler and more direct approach for routine, clinical practice is to use the generic and all-encompassing term *velopharyngeal dysfunction*. This term does not assume, or rule out, any possible cause of the perceived speech symptoms or any management approach. As described by Netsell, velopharyngeal dysfunction during speech may be the result of structural deficits, neurological disorders, faulty learning, or a combination of sources. Dalston uses the term *velopharyngeal dysfunction* and defines it as follows:

... any impairment of the velopharyngeal complex. It may result from a lack of sufficient tissue to enable contact to be effected between the soft palate and the posterior pharyngeal wall ("velopharyngeal insufficiency"), a lack of neuromuscular competency in moving velopharyngeal structures into contact with one another ("velopharyngeal incompetency") or both. Finally, it may be due to maladaptive articulatory habits that do not reflect physical or neuromuscular impairment (eg, phoneme-specific nasal emission).

By not implying the source of the symptoms, the term *dysfunction* acknowledges that some features of the velopharyngeal valving mechanisms are not functioning appropriately,
but no cause or treatment approach is implied or suggested until appropriate diagnostic testing can be conducted.

One more note on terminology seems appropriate in the discussion of velopharyngeal function. In the book *Cleft Palate Speech* McWilliams et al point out that individuals with velopharyngeal incompetence are typically described as displaying "hypernasal voice quality". However, the word *voice* more accurately refers to problems associated with phonation. Therefore, they suggest the use of the term *phonation disorders* to refer to problems that occur at the level of the larynx and the term *resonance disorders* to refer to hypernasality and other disturbances that occur supraglottally. Once again, the use of clearly defined, physiologically based terminology leads to a more accurate and precise nomenclature for both clinical and research applications.

**Differential Diagnosis**

In the evaluation and treatment of velopharyngeal dysfunction, the clinician must be aware that speech symptoms often attributed to velopharyngeal incompetence may come from a variety of sources or combination of sources. For example, hypernasality and/or nasal emission may be the result of the lack of sufficient tissue to allow closure of the velopharyngeal port; neuromotor impairment involving innervation of muscles of the velopharyngeal port as in many congenital anomalies, neurologic diseases, or head injury; mislearning or other behavioral factors as in instances of phoneme-specific nasal emission or lack of oral/nasal discrimination; or from other structural involvement, such as a palatal fistula, that may allow nasal air escape or enlarged tonsils, which have been shown to result in impaired velopharyngeal function in some instances.

The process of differential diagnosis can be quite difficult since speech symptoms that may appear to be quite similar and indistinct to the casual listener or inexperienced observer or in some instances even the experienced listener may in fact be quite diverse in both etiology and, therefore, appropriate management. For example, in the cleft population, it is not uncommon to observe hypernasality and nasal emission in a patient with a repaired cleft and a residual oronasal fistula. Casual perceptual observation cannot determine the source of these symptoms. Therefore, it would be inappropriate to label the phenomenon as velopharyngeal incompetence without investigating the symptoms further. In some instances, the symptoms may be solely attributable to air escape through the fistula. In this case the diagnosis would be hypernasality and nasal emission due to a patent oronasal fistula and may be unrelated to the velopharyngeal mechanism. In this diagnostic situation, the appropriate management would be repair or obturation of the fistula. In other cases, the symptoms may appear to be attributable to a combination of air escape through the fistula and a lack of proper velopharyngeal function. In these cases, diagnosis and management planning become far more complicated.

Another illustration of the need for differential diagnosis is the phenomena of "phoneme-specific nasal emission". This is the perception of nasal emission isolated to specific pressure consonants such as /s/ and /z/, as opposed to the consistent nasal emission generally seen when the velopharyngeal valve is incapable of closure. The inexperienced clinician (and until recent years, even some experienced clinicians) have mistaken this articulation-based error for true velopharyngeal incompetence. Many patients have been
referred for or have received inappropriate physical management for what is basically an articulation error that requires behavioral therapy for effective remediation.

As these examples illustrate quite clearly, the evaluation of velopharyngeal function for speech must be carried out by a speech pathologist with expertise in this area. Although some cases may be straightforward, most are not. It is a detailed description of symptoms, their frequency and severity, and their response to behavioral probes that defines the problem and leads to appropriate management suggestions. This description or differential diagnosis is made by addressing a number of diagnostic questions using a variety of evaluation methods. For example, the speech pathologist might ask:

1. Can the patient achieve complete velopharyngeal closure?

2. If the patient can achieve closure, what are the speech symptoms that are resulting in the perception of a speech disorder?

3. If the patient cannot achieve closure, what are the possible reasons why not?

4. If closure is intermittent, which speech environments facilitate closure and which contexts are detrimental?

5. How much do symptoms of velopharyngeal dysfunction contribute to the overall communication handicap?

6. Has previous behavioral management for the symptoms in question been attempted? If so, what was the outcome?

7. Is management indicated? If so, is behavioral management, ie, speech therapy, indicated in isolation or in conjunction with physical management.

Ultimately, the speech pathologist must interpret the findings of the comprehensive evaluation of velopharyngeal function along with the patient's history and physical findings and arrive at a provisional differential diagnosis. This information should then be discussed with members of a multidisciplinary team and interpreted along with other pertinent physical and social information regarding the patient. Only then can an appropriate, individualized management and follow-up plan be established.

The Rationale for Multilevel Multimethod Evaluation

Speech production is one of the most intricate of all human behaviors. A block diagram of the speech production process as conceptualized by Netsell: CNS organization --> Nerve impulses --> Muscular events and structural movements --> Air pressures and air flows --> Acoustic waveform. The process originates in the central nervous system governed by various linguistic processes. Nerve impulses travel to various muscles and the muscular events result in structural movements. As many as 100 muscles may be involved in this process. These muscle and structural movements then result in air pressure and airflow events, which result in the acoustic output that ultimately is perceived as speech. Although this process occurs over a number of levels, generally only the latter two or three levels are available for
observation and measurement. For many years we were limited to observations at one level of this complex speech production process, ie, the auditory perception of the acoustic output, or listener judgments of speech. With the increased application of instrumentation in clinical practice we have moved more routinely into observation of aerodynamic events and structural movements.

This view of the speech production process reminds us that in clinical practice, the events we most frequently observe as "speech" represent the most peripheral level of observation in the speech production process. This fact provides the rationale for the use of instrumental methods that allow us to make observations of the speech production process at earlier levels. Information from instrumental assessments at various levels of this complex process not only provide relevant clinical information about speech production but also provide insight into speech motor control processes.

Which Method is Best?

Many clinicians and researchers who are experienced in the use of instrumental methods, frequently are asked which method is best. In this era of shrinking budgets and ever-reducing reimbursement, this question might at first appear quite reasonable. However, the answer is not simple. Instrumental methods can differ from one another on a number of features including the level of the speech production process accessed, whether the method provides direct or indirect observation, clinical practicality, invasiveness, cost-to-benefit ratio, validity, and reliability.

There is increasing interest in the use of instrumental methods for assessing velopharyngeal function for speech. It has been suggested that instrumental methods are more reliable and informative than listener judgments alone, and therefore lead to a more accurate diagnosis and improved treatment planning. Critics of the trend toward instrumental assessment often cite a study by McWilliams et al to show the poor correlation among instrumental methods, particularly aerodynamic methods. In this study, McWilliams and her coauthors compared listener judgments of velopharyngeal function (specifically hypernasality) with nasal manometry, pressure-flow, and multiview videofluoroscopy. They showed complete agreement among methods for only three of the 35 speakers studied.

Although these data have been interpreted to point out the poor validity of some instrumental methods, an alternative interpretation has been proposed. D'Antonio and her colleagues have suggested that the important issue is not how often the methods agree completely or even which method is best. Rather, the user must ask:

1. What unique and what overlapping information does each method provide?
2. When is it appropriate to use a given method or combination of methods? That is, what information is being sought? And is this the most appropriate method and observers to provide that information?
3. When the information from multiple methods does not appear to agree, what information does this provide and how might this alter our diagnostic and management decisions?
Results from two studies reported by D'Antonio and colleagues suggest that the reliability and validity of evaluation methods may be affected to a greater degree than we have acknowledged by variables such as an individual user's training, background experience, and visual and auditory perceptual strengths. The authors suggest also that in clinical practice, it is likely that the effects of these and other variables that may result in instances of "apparent disagreement" may be diminished by operation factors such as the combined use of multiple evaluation methods and the use of an interdisciplinary team approach for the collection and interpretation of evaluation data.

Another important question that must be addressed in clinical practice is whether a methodology is being employed correctly. As with all areas of medicine, there is great potential for misuse and damage when instrumentation is used incorrectly. With the increased utilization of instrumental methods for the evaluation of velopharyngeal function comes the distressing reality that in several instances the advantages and limitations of a method and the recommendations for its appropriate use have been ignored. For example, D'Antonio et al conducted a national survey concerning methods used for the evaluation of velopharyngeal function and showed 90% of cleft palate teams now have nasendoscopic evaluation capabilities. This is a significant increase in availability compared with studies conducted in 1980 and 1984, which showed only 8% availability. Unfortunately, the results also showed that increased availability did not necessarily assure optimal use.

For example, the principal advantage of direct visualization techniques such as nasendoscopy is the assessment of velopharyngeal function during dynamic speech activity. However, three cleft palate teams reported the use of general anesthesia or heavy sedation during endoscopic examination. Utilization such as this clearly demonstrates the potential for significant misuse when the clinicians involved have not clearly identified the questions being asked and the most appropriate method to answer those questions. Additionally, responses to the same survey were disappointing with regard to the interpretation of results. In the majority of centers, endoscopic evaluations of velopharyngeal function were performed by a physician alone. This finding is of particular concern since most physicians are not likely to be familiar with many of the behavioral variables that can affect velopharyngeal function or with how to interpret physiologic findings in conjunction with the overall speech production processes and communication skills present in a given patient. Results such as these indicate that the advantages instrumental methods can and often do have can be overshadowed by inappropriate use. Once again, clinicians must insure that the most suitable method is being utilized by the most appropriate user in order to answer the question under investigation. In this way, we can help to insure that the method being employed is "the best method".

**Evaluation Methods**

Numerous methods exist for evaluating velopharyngeal function at different levels of observation within the speech production process. Frequently reported methods include:

- Listener judgments
- Intraoral examination
- Acoustic analysis
  - Spectrograph
  - TONAR
Accelerometer
Aerodynamic measures
Observation of structural movement
  Multiview video fluoroscopy
  High-speed x-ray
  Ultrasound
  Photodetector
  Nasendoscopy
  Electromyography.

It can be noted to some extent, that the least invasive procedures allow observation at the most peripheral stages of the speech production process and the relative invasiveness of the methods increase as we move up to higher, or earlier, stages.

Obviously, not all, or even most available evaluation methods can or should be employed in a given patient or patient population. As discussed previously, each method has inherent strengths and weaknesses and can provide both unique and overlapping information pertaining to velopharyngeal function.

The rationale for selection of assessment methods appropriate to a given population or question has already been discussed in the section Which Method is Best? However, one topic that was not covered is the strategy for selecting a combination of methods for clinical use that complement each other's unique contributions and limitations. In this way, the outcome can be one where "the whole is greater than the sum of its parts". For example, we have seen that nasendoscopy provides direct visualization of the velopharyngeal port but relies on subjective, visual interpretation, usually by a single observer. On the other hand, aerodynamic assessment is an example of a method that may provide quantitative data regarding the effects of velopharyngeal closure, but in this case the data about velopharyngeal closure are indirect. As illustrated below, these two methods can compliment one another and if utilized together, can provide direct and quantitative information about velopharyngeal function.

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<th>Nasendoscopy</th>
<th>Aerodynamic Measures</th>
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<td>indirect</td>
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This examples illustrates once again that the selection and combination of instrumental evaluation methods should be given great consideration prior to their inclusion in the diagnostic armamentarium.

In current practice, the most commonly employed methods for clinical evaluation of velopharyngeal function are perceptual assessment by a trained speech pathologist, aerodynamic measures, nasendoscopy, and multiview videofluoroscopy.

**Peroral and Head and Neck Examination**

A thorough peroral and head and neck examination is essential for any child who presents with known or suspected velopharyngeal dysfunction. The oropharynx is visualized
and palpated, and the length of the soft palate gauged with respect to the depth of the nasopharynx, both at rest and during phonation. Symmetry of palatal excursion during phonation is noted, keeping in mind that normal palatal motion with swallowing and gag often is present in children even with severe velopharyngeal dysfunction and does not relate to, or predict, velopharyngeal activity during speech. Any signs of palatal clefting are noted: bifurcation or notching of the uvula, notched or U-shaped posterior border of the hard palate, and any midline mucosal color change (zona pellucida) or muscle bulge indicating muscle diastasis.

The general head and neck examination should place special emphasis on three areas: the upper airway; the ears, including audiological evaluation; and any structural anomalies that may indicate the presence of a recognized pattern of human malformation or syndrome of which cleft palate is a known feature. The upper airway (including the septum, turbinates, adenoid and palatine tonsils, and tongue base) is examined for obstruction that may affect speech quality or speech articulation and for any potential obstruction that might further compromise the airway if any surgery is performed to improve velopharyngeal function. The relationship between the nasal airway and breathing and speech is far more complicated in patients with cleft palate than has been understood historically. An excellent review of current clinical data of importance for the head and neck surgeon involved in the care of patients with cleft palate can be found in the work of Warren et al.

The ears are examined for middle ear effusions, and audiologic testing is completed, if needed. General craniofacial configuration is examined; any anomalies such as synostoses, telecanthus, maxillary or malar hypoplasia, abnormal pinnae, abnormal mandibular shape and excursion, or malocclusion may be reason for referral to a pediatric geneticist.

Perceptual Speech Evaluation

Evaluation by an experienced speech pathologist is essential and should include a comprehensive evaluation of communication skills, including symptoms of velopharyngeal dysfunction. Particular attention should be paid to assessment or resonance, nasal emission, articulation, the presence of compensatory articulations (such as glottal stops and pharyngeal fricatives), the presence of facial grimacing, and overall speech quality and intelligibility. The head and neck surgeon should be capable of performing simple and brief listener judgments of these variables so that he or she is aware of when to refer for a speech language evaluation or to an expert in the evaluation of velopharyngeal function. However, under no circumstances should the head and neck surgeon attempt to perform a speech evaluation for the purposes of differential diagnosis of velopharyngeal dysfunction, just as a speech pathologist would never perform surgery.

The advantage of the listener evaluation is that it has high "face validity", that is, the human auditory perceptual system is excellent at determining when there is a speech disorder, particularly impaired velopharyngeal function. It is the way that speech sounds that is the "gold standard" for whether treatment is indicated. In most instances, regardless of the results of more sophisticated instrumental measures, it is the contribution that an impairment in velopharyngeal function causes to the overall communication competence of an individual that will determine the severity of the disorder and whether management of any kind is indicated. Unfortunately, in spite of the fact that it is "how the speech sounds" that is the critical
variable, there are numerous criticisms that indicate that subjective, listener evaluations, by a single clinician (without known inter- and intrarater reliability) are frequently unreliable. Listeners vary in tolerance for various speech parameters and differ greatly in personal biases of what may constitute a communication impairment. Therefore, although listener judgments of speech are essential, they should always be accompanied by instrumental assessments that may help to validate the listener's perceptions. Shprintzen and Golding-Kushner summarize this point quite well:

The correct approach to treatment of VPI is dependent upon adequate diagnostic information, and potentially dangerous or fatal complications may occur when VPI is treated inappropriately. Therefore, it is imperative that clinicians apply state-of-the-art diagnostic procedures to the patient with VPI. This is of particular importance when one considers that hypernasality is one of the few speech disorders that is often treated with some form of physical management, including a large number of surgical approaches.

Aerodynamic Measures

As described previously, the human vocal tract is an air-filled container closed at the bottom by the lungs. The lungs generate air pressure that is valved by the vocal tract at a number of levels including the larynx, velopharynx, nasal cavity, and lips. A number of aerodynamic methods have been employed such as active posterior rhinomanometry to assess the function of the velopharynx and nasal cavities during speech. Warren and DuBois introduced the most commonly applied methodology for the measurement of oral and nasal pressure differential and nasal flow, which can be placed into a hydrokinetic formula to estimate the area of velopharyngeal opening at the moment of peak pressure of a stop consonant. Other methods exist that are an adaptation. Although this methodology requires sophisticated equipment and significant operator competence, its major advantage is that it is a noninvasive method that can be used with small children to provide quantitative data concerning velopharyngeal function. Studies have shown these methods (when used and interpreted properly) provide valid and reliable information regarding velopharyngeal function. These methods are particularly valuable because they can subtract out the contribution of the nasal passages to the impedance of airflow from the velopharynx. This is especially useful in the common situation of the patient with velopharyngeal incompetence in the presence of hypertrophied turbinates or a structurally occluded nasal airway. In these instances, aerodynamic measures can provide critical information that the human ear simply cannot sort out. Aerodynamic data also provide important information about the timing of closure of the velopharyngeal port that is of great value in patients with neurologic contributions to their velopharyngeal symptoms.

Although aerodynamic methods have numerous advantages and important applications for the cleft and noncleft populations, they have not gained widespread clinical popularity because of the expense involved in instrumentation and the amount of training necessary for accurate interpretation of test data. Additionally, although aerodynamic measures are quantitative, valid, and reliable, they tell the surgeon nothing about the anatomy and structure of the mechanism he or she may be about to perform surgery on.
Nasendoscopy

Movement of the velopharyngeal mechanism during speech is complex and includes musculature contractions of not only the soft palate, but also of the lateral and posterior pharyngeal walls. This complexity is reflected in the fact that the patterns of closure of the velopharyngeal port are highly variable among both normal speakers and patients with velopharyngeal dysfunction. Every child with known or suspected velopharyngeal dysfunction should undergo nasendoscopy, especially if a surgical procedure is anticipated. The examination should be performed with both the speech pathologist and surgeon in attendance, with a permanent record of the examination recorded on videotape. Permanent documentation on videotape is important for reviewing results of the evaluation after the stress of the actual procedure is over. It allows for multidisciplinary review of the results and provides a means of monitoring magnitude and direction of change. This last advantage cannot be emphasized enough in today's medical-legal climate. Additionally, if surgical intervention is recommended, a videotape record of the evaluation allows the surgeon to view the tape just prior to the operation to recall the specifics of the pattern of closure and the function of the mechanism to be operated on.

Most children 3 years of age and older can be examined successfully with the nasopharyngoscope. D’Antonio et al found that 87% of children between 3 and 6 years of age can be examined. The diameter of the scope can be a limiting factor. The 3.6-mm scope gives a superior video image, and can be used in younger children. The 3.0- to 3.2-mm scope results in a video image of slightly less quality, but can be used in younger children. Nasal mucosal anesthesia is accomplished by means of a hand-held atomizer, which is used to gently spray 2% cocaine solution into the anterior portion of the nasal cavity. If this does not provide adequate anesthetization, a tightly rolled cocaine-soaked cotton pledget may be gently introduced into the nasal cavity and passed posteriorly to further anesthetize the most sensitive, posterior areas of the nasal cavity.

Following removal of the pledget, the nasopharyngoscope is passed above the inferior turbinates, opposite the middle meatus, into the nasopharynx. The scope is positioned so that a complete circumferential view of the nasopharynx is obtained. The child is then asked to produce a sample of words and short sentences that comprise an adequate speech sample. The speech pathologist should be available to conduct the speech evaluation during the endoscopic examination. Behavioral, diagnostic probes should be used to ascertain the child's performance limits. If possible, attempts toward velopharyngeal closure should be assessed during correctly articulated speech sound productions only. Finally, function should be observed in single sounds, single syllable words, short phrases and sentences, and in increasing levels of complexity. The speech sample and evaluation should be designed to place minimum and maximum demands on the velopharyngeal mechanism during speech production. Consistency of closure and patterns of variability are critical findings for the speech pathologist to help establish a differential diagnosis.

Direct visualization of velopharyngeal movement in this manner allows a mapping of the defect that is recorded on videotape. The degree and symmetry of soft palate motion, lateral pharyngeal wall motion, and the pattern of velopharyngeal closure, or attempted
closure, in the coronal and sagittal planes are noted. In addition to the permanent videotaped record, movement of the soft palate, lateral pharyngeal walls, and posterior pharyngeal walls may be rated on a numerical scale and recorded on a diagram. The vertical level of most prominent lateral wall motion is estimated with respect to its distance from the rostrum of the torus tubarius.

It is important to pass the scope into the inferior nasopharynx, as the vertical level of most prominent lateral wall motion may be obscured by movement of the soft palate superiorly. If large palatine tonsils have been noted previously during the peroral examination, special attention is afforded them during videonasendoscopy. Some children with mild velopharyngeal symptoms do not exhibit adequate lateral seal between the velum and the lateral pharyngeal walls because the superior pole of one or both palatine tonsils projects into the nasopharynx.

Following evaluation of the velopharyngeal mechanism, the head and neck surgeon should evaluate laryngeal and vocal fold function during rest and phonation. Over 40% of patients with VPI will exhibit abnormal voice characteristics, as well as pathologic vocal fold changes such as nodules, thickening, edema, and various patterns of incomplete glottic closure. This information can be important for directing behavioral speech therapy in selected cases.

Videonasendoscopy should be repeated approximately 3 months to 6 months postoperatively. This is necessary to confirm the success of the surgical procedure in those children who have achieved postoperative velopharyngeal competency. In patients who demonstrate persistent velopharyngeal dysfunction postoperatively, videonasendoscopy allows the surgeon and the speech pathologist to visualize the mechanism during speech and to determine whether the residual symptoms are physical or behavioral. This allows proper planning of a revision procedure if necessary or for additional speech therapy.

The advantages of nasendoscopy are obvious. It allows for direct observation of the velopharyngeal mechanism during dynamic speech activity. It is relatively noninvasive (under appropriate circumstances) and gives clinicians important information about the physical structure to be modified if physical management is necessary. Another advantage that has not been discussed is the potential for nasendoscopy to be used for diagnostic therapy or for biofeedback therapy.

In spite of the significant advantages of the use of nasendoscopy in the evaluation of velopharyngeal dysfunction, the method does have some limitations that should be remembered. At the present time it is still a subjective, visual-perceptual evaluation. Quantification methods do not exist for this procedure. There are numerous problems with image distortion, scope placement, repeatability etc. Additionally, as discussed previously, since most otolaryngologists are familiar with the procedure and scopes are readily available in almost all otolaryngology offices, there is the danger that many head and neck surgeons feel qualified to perform the complex differential diagnosis of velopharyngeal dysfunction without the assistance of a qualified speech pathologist.
Multiview Videofluoroscopy

This method is a radiologic technique that facilitates evaluation of the velopharyngeal mechanism in several planes during dynamic speech activity. In most settings, high-density barium is placed into one or both nostrils with a syringe and the patient is instructed to "sniff" the contrast material into the nasopharynx. The patient is positioned and as many views as necessary (and practical) are conducted in order to obtain an adequate representation of velopharyngeal function. The most common views employed are the lateral view, frontal view, Towne's view, and base view. (In some institutions, the Towne view will be used as a substitute for the base view and the Waters projection will be used as an alternative for the frontal view.)

The lateral view usually is taken first, and is the most common. It provides information about the relative length of the soft palate, its thickness, and the depth of the pharynx. Tonsillar and adenoid tissue are also noted. The frontal view provides important information about the vertical location of most active movements of the lateral pharyngeal walls. The combination of the lateral and frontal views supplement one another to give a more accurate impression of the velopharyngeal mechanism. However, in some cases important information is still missing and the base (or Towne or Waters view) is necessary to clarify the three-dimensional nature of velopharyngeal movements.

Many of the advantages and cautions associated with videofluoroscopy are similar to those discussed for nasendoscopy. For instance it should always be performed by the radiologist in cooperation with the speech pathologist. Interpretation of results should never be attempted without sound accompanying the fluoroscopic images or in isolation of a more comprehensive speech and velopharyngeal evaluation. Additionally, lateral, static, two-dimensional x-rays are not a substitute for providing information regarding the dynamic activity of the velopharyngeal mechanism.

Earlier multiview cineradiography allowed frame-by-frame analysis of articulatory movements over time (not only of the velopharynx but of the tongue, lips, and jaw as well). Videofluoroscopy continues to allow the assessment of the velopharynx in relation to other articulators but does not lend itself well to accurate quantitative analysis over time. However, this limitation is overridden by the reduction in radiation necessary for cineradiographic studies compared with videofluoroscopic examinations. Nevertheless, even in the latter case the amount of radiation exposure is the principal disadvantage of this method, especially in young children for whom repeat studies may be necessary.

Issues Related to Management Decisions

Once a diagnosis has been established, attention turns toward the question of whether management of any kind is indicated. McWilliams et al address the interdisciplinary roles in diagnosis and management:

We want to emphasize our conviction that it is the role of the speech pathologist to judge the adequacy of velopharyngeal function for speech production. This is done in collaboration with other professionals who also provide valuable information about the mechanism and its function and suggest
alternatives for management. If physical management is chosen as the preferred method of treatment, the surgeon or the dentist is legally and ethically accountable to the patient and family for proper treatment and must always have the last word about whether or not to perform the surgery or construct the prosthesis. However, it is the speech pathologist who decides whether or not the proposed physical management is indicated for speech improvement.

No form of management is without its own costs. Although surgical management may be the most permanent and carry the most physical risks, prosthetic management and speech therapy also require significant compliance, time commitment, and often emotional commitment. Therefore, no management alternative should be undertaken casually.

It is incumbent upon the speech pathologist to interpret the available evaluation data and to determine whether management of any form will improve speech intelligibility, speech quality, or quality of life. Unfortunately, there is no formula for making such decisions easily. For example, in one instance, a patient may present with a significant communication impairment of which velopharyngeal dysfunction is only a small source of the overall decreased communication competence. In such a case a cost-benefit analysis might lead to the decision not to offer physical management, especially surgical management. Another patient might present with mild velopharyngeal dysfunction with no other communication impairment. Although the symptoms may be slight the patient and/or the patient's family may feel that even a small observable stigmata will result in detrimental effects and reduced life options. Again a cost-benefit analysis may suggest that physical management will be likely to result in only a minimal improvement in speech quality. However, in this case even small improvement may be enough to translate into significant improvements in quality of life. In this situation, it may be appropriate to offer physical management, even surgical management.

This discussion illustrates the point that in spite of our ability to collect detailed scientific data concerning the source and magnitude of velopharyngeal symptoms, in the final analysis decisions regarding management are as much a part of the art of cleft care as the surgical act itself.

As discussed previously, there are three broad categories of management for velopharyngeal dysfunction - speech therapy, prosthetic management, and surgical management - which will be discussed in the following sections. All three of these management categories are familiar to clinicians involved in cleft care. Most readers will have personal biases about the particular effectiveness and value of each method. However, in reality, the reader should realize there has been little prospective research on treatment outcome for any of these methods. Much of what we, as clinicians, believe to be true about each method's effectiveness usually is based on personal experience.

McWilliams et al quite accurately point to the need for prospective research concerning treatment effectiveness. They remind us that the same methods available for the evaluation of velopharyngeal function

... are also appropriate for assessing changes in speech and velopharyngeal function in response to therapy and to surgical and dental treatments. Prospective research predictive of treatment outcome is needed. It is difficult
to accomplish because many variables in addition to treatment influence the results. For example, patients with different characteristics respond differently to a given treatment, and measurement error is not unknown. Two clinicians may differ in the skill with which they use a particular technique. These confounding variables are difficult to control experimentally, but the development of well-founded treatments depends on sound treatment research.

**Speech Therapy**

The role of the speech-language pathologist and the field of speech pathology in general often appears to be an enigma to most medical specialists. There are a variety of potential speech-language pathology interventions for patients with craniofacial anomalies that may come under the broad label of "speech therapy". These include parent counseling, speech and language stimulation, joint parent-child communication therapy (individual or group), articulation therapy, phonological processes therapy, resonance therapy, voice therapy, velopharyngeal function therapy, and language therapy.

In discussing the role of behavioral therapy in the management of velopharyngeal dysfunction Van Demark and Hardin provide an excellent definition of the role of speech intervention:

The goal of speech therapy for any child is to establish age-appropriate speech production patterns through behavioral modification. In addition to addressing any developmental articulation deficits that the child may demonstrate, therapy for the child with cleft lip and palate is typically directed toward the objectives of eliminating or reducing inappropriate patterns of nasalization, oral distortions, and "compensatory" articulation gestures.

In discussing the role of speech therapy in the management of velopharyngeal dysfunction, it must be understood that in most cases the velopharyngeal symptoms cannot easily be separated out from other speech production disorders. Therefore, therapy usually must focus on more than one source of speech errors and clinicians may take diverse approaches to staging therapy goals and/or managing different aspects of the global disorder. Just as there are numerous approaches to the surgical correction of cleft lip and palate, there are a variety of opinions and approaches to the treatment of speech impairments associated with clefting.

**Diagnostic Speech Therapy**

A common surprise for most nonspeech pathologists is that a child with velopharyngeal dysfunction who currently is receiving speech therapy or has received many years of therapy, may never have engaged in training focused specifically on improved velopharyngeal function. Unfortunately, without direct knowledge of a patient's therapy program or appropriate documentation of programming, the mere fact that a child has received speech therapy is no insurance that velopharyngeal function itself has ever been a target of therapy.


Therefore, when velopharyngeal function is borderline or variable, it is appropriate to
determine whether it can be improved. A careful review of previous therapy may suggest that
suitable efforts have been made to stimulate improved closure. In such cases, further
diagnostic attempts may not be necessary. However, frequently such a history cannot be
documented and a period of "diagnostic therapy" may be warranted.

When velopharyngeal function is variable, as it often is, particularly in young children
with cleft palate, an important source of diagnostic information is "stimulability testing". A
cornerstone of modern speech therapy is that a child's ability to be stimulated for improved
speech production through auditory and visual (and in some instances tactile) models and cues
is a good prognostic indicator of potential for long-term improvement. Such stimulability
testing is particularly useful in the child with variable velopharyngeal function. It can provide
valuable information about whether behavioral management is likely to remediate the
velopharyngeal symptoms or if such amelioration of symptoms appears unlikely.

Morris suggests there are two major subgroups of patients with marginal
velopharyngeal dysfunction and proposes that the two groups can be distinguished by the
response to short-term therapeutic intervention.

The first group is the almost-but-not-quite (ABNQ) subgroup. This group tends to
present with mild and consistent nasalization of speech, which is highly consistent among and
within tasks. Morris suggests:

speech training is not successful with the ABNQ group for the purpose of
improving velopharyngeal function, that is, because the patient apparently has
already extended the mechanism to the physiologic limits and does so
consistently. If trial training for that purpose is provided to confirm the
diagnosis of ABNQ marginal incompetence, it should be discontinued after 6
hours of treatment if no improvement in velopharyngeal function is observed.

In this case, the lack of response to highly focused therapy suggests that physical
management is the treatment of choice if the impairment is significant enough to warrant
intervention.

The second diagnostic group of marginal velopharyngeal function described by Morris
is the sometimes-but-not-always (SBNA) subgroup. Patients in this group generally show
marked inconsistency in velopharyngeal function. Morris indicates that some patients in this
category will show improvement with training and some will not. He believes that the major
diagnostic determinant in this group is the lack of positive response to directed speech therapy
focused on improving velopharyngeal function. Morris suggests that patients in this group
usually demonstrate that they are capable of increasing oral productions within their speech
repertoire at the single sound or single word level. However, they often are unable to
generalize this pattern into connected, conversational speech. Morris explains the importance
of diagnostic therapy in this group:

Diagnosis of these patients is frequently controversial. Inexperienced speech
pathologists fail to interpret correctly the lack of improvement from training.
Parents are falsely optimistic about the outcome because they observe the
variance in speech production that is typical of the group. Surgeons and dentists who work often with cleft patients overinterpret the observation that the SBNA patient can perform well on single word tasks or highly specific speech activities. As a consequence, it is vital that observations about response to speech training (or rather, lack of it) be included as part of the diagnostic findings for these patients.

Morris suggests that a child in the SBNA subgroup who will be capable of achieving complete, consistent velopharyngeal closure should do so after approximately 10 hr of intensive, focused training. On the other hand, he believes that if no improvement is observed in connected speech within 10 to 20 hr of training, the diagnosis of SBNA should be made.

Unfortunately, it is these patients who present the greatest dilemma for the speech pathologist and other team members in attempting to establish an appropriate management plan. Oftentimes these patients' inability to achieve consistent closure is a complex, interrelated problem with several contributing variables. For instance it is likely that many of these patients demonstrate poor timing of velopharyngeal movements and poor coordination with other articulators and vocal tract components, which may result in the observed variability. Since the velopharyngeal symptoms in this group are quite complex in origin and manifestation, it is this group of patients that often shows limited improvement following surgical intervention. Therefore, the decision to manage these patients surgically should be undertaken with caution. Detailed counseling of the patient and the patient's family concerning realistic expectations from surgical intervention should be a high priority.

**Articulation Therapy**

All speech sounds can be characterized by the *place of articulation* (ie, the position of the lips and tongue for a given sound), the *manner of articulation* (ie, the way the vocal tract is altered to constrict the flow of air), and whether the sound is accompanied by laryngeal vibration (ie, voiced or voiceless).

Articulation therapy is the process of training an individual to produce a sound in the correct place of articulation and to direct the airflow in the correct manner with appropriate laryngeal voicing. Articulation therapy for individuals with cleft palate uses similar principles and techniques as articulation therapy for other clinical populations. However, the types of errors, the severity of the disorder, and the staging of therapy may be complicated by the presence or possible presence of physical impairments. Physical contributors that may impair articulation development or articulation skills include past or present velopharyngeal dysfunction, palatal fistulae, abnormalities in the skeletal relationship between the maxilla and mandible, dental abnormalities and nasal airway deviations.

Not all children with cleft lip and palate necessarily develop misarticulations. However, when articulation errors occur, they traditionally have been categorized as omissions, distortions, and substitutions (including compensatory articulations). In general, for most children with cleft palate, tongue placement is shifted posteriorly in an attempt to valve the airstream before it escapes or is diminished through an open velopharyngeal port or through an open cleft. Most commonly, the correct place of articulation is sacrificed while the correct manner of articulation is preserved.
There are several opinions in the literature regarding the relationship between velopharyngeal dysfunction and articulation disorders. Peterson-Falzone sums up the controversy:

The relationship between velopharyngeal closure and speech is rarely as clear-cut as both the clinician and the research worker would like. Studies relating specific speech problems (and the severity of those problems) to measurements of velopharyngeal closure have often yielded contradictory and confusing results because of problems in measuring both speech output and the function of the velopharyngeal system.

In spite of the research data necessary to confirm a causal relationship between velopharyngeal insufficiency and resulting articulation errors, most clinicians would agree that many of the misarticulation patterns found in patients with cleft palate are the result of past or present velopharyngeal insufficiency. There is no disagreement that these articulation errors should be addressed in therapy. However, there are several opinions in the literature regarding the timing of articulation therapy in general and, in particular, its value (if any) in managing velopharyngeal dysfunction.

No one would argue that articulation therapy cannot eliminate a large velopharyngeal gap due to the absence of adequate tissue. However, Van Demark and Hardin cite several studies that show decreased perception of hypernasality and/or audible nasal emission (ie, symptoms of velopharyngeal dysfunction) in some children following intensive articulation therapy. They suggest, "This improvement, along with concomitant improvement in articulation, may facilitate speech intelligibility for select children and minimize the need for surgical management".

Hoch et al point out that traditionally it was believed that articulation therapy for children with documented velopharyngeal insufficiency should be deferred until physical management of the velopharyngeal mechanism was accomplished. One argument for this approach is that inappropriate therapy (especially for young children) may lead to posterior tongue posturing to facilitate velopharyngeal closure, which can then result in undesirable compensatory articulation patterns that may be difficult to remediate after surgery. Hoch et al and Shprintzen et al cite evidence to suggest that articulation therapy can improve velopharyngeal function prior to physical management. These authors assert that the changes in velopharyngeal function associated with the improved articulation may alter subsequent surgical planning. Based on their experience with articulation therapy prior to surgical intervention. Hoch et al recommend the following:

To summarize, when VPI is observed in association with compensatory articulation errors (most specifically glottal stop substitutions), we believe that surgical correction of the VPI should be deferred. This approach is contrary to the widely accepted practice of first physically managing the VPI. We suggest that it is more appropriate to eliminate the compensatory articulation errors first with a well-planned speech-therapy program. The rationale is that improvement of oral articulation may have the benefit of improving the valving of the velopharyngeal sphincter. Furthermore, in at least a small percentage of cases, correction of the oral articulation disorder will eliminate
the need for surgery altogether. In addition, when oral articulation has been normalized the effects of surgery can be immediately appreciated and assessed.

The controversy between these two approaches continues to exist. However, with the use of direct visualization techniques, there is increasing clinical evidence that marked improvements in velopharyngeal function can be observed over time with speech therapy. It is unclear whether these changes are truly the result of "articulation therapy" alone or other behavioral therapeutic techniques included during the process of articulation therapy. However, these numerous and frequent clinical observations support the approach espoused by Shprintzen and Hoch et al and suggest that at a minimum a trial of intensive articulation therapy is warranted for some patients prior to secondary surgical management for velopharyngeal insufficiency. Certainly, well-controlled experimental studies are needed to confirm these observations and to establish a databased approach to treatment planning.

Behavioral Therapy for Velopharyngeal Dysfunction

As discussed in the section on speech therapy as a diagnostic tool behavioral therapy is often appropriate for a limited duration for patients with inconsistent velopharyngeal closure. Van Demark and Hardin suggest that the goals of behavioral management in these cases are:

1. Maximize the range of velar or pharyngeal wall movement when small portal openings are evident.

2. Generalize oral responses obtained during simple speech tasks (single-word production) to more complex tasks (connected speech) when inconsistent closure is demonstrated.

Proponents of behavioral therapy have argued that a patient who evidences marginal closure should be able to enforce purposeful movement of the velopharyngeal structures. Opponents have pointed out, however, that despite clinical case reports of success, limited data are available to support the notion that velopharyngeal competence can be taught.

Speech pathologists who may read this chapter and wish more information regarding behavioral management techniques for velopharyngeal dysfunction should refer to the writings of McWilliams et al, Van Demark and Hardin, and Hoch et al.

In addition to traditional behavioral techniques, a variety of biofeedback methods have been described. However, little experimental data are available concerning the effectiveness of instrumental methods in managing velopharyngeal dysfunction. Until such data are available, biofeedback therapy should be considered experimental. Nevertheless, individual reports from several centers and investigators suggest that instrumental biofeedback techniques may have significant promise as treatment modalities and should be investigated further.

The goal of all of these methods - articulation therapy, behavioral therapy, and biofeedback alike - is to facilitate consistent velopharyngeal closure first at the single-sound level, then expanded to all sounds in all sound contexts, single words, short phrases, modeled
connected speech, and, ultimately, spontaneous conversational speech. Therapy should begin at the lowest level at which the child can achieve success and progress at a rate that challenges the child but allows continued success. If progress is consistently observed it is reasonable to continue with behavioral management. However, although behavioral management for children with marginal, variable velopharyngeal dysfunction can be quite successful and rewarding in some cases, therapy should be continued only as long as progress can be documented. It is just as unethical to relegate a child to prolonged speech therapy with no success as it is to refer a child with behavioral velopharyngeal symptoms to surgery. The critical variable in engaging in behavioral therapy for these patients with variable, marginal velopharyngeal dysfunction is to know when "enough is enough". In many cases, surgical intervention is far more humane than the cruelty of continued, prolonged behavioral therapy that is doomed to failure. In these cases, the potential for reduced self-esteem and a personal sense of failure outweighs any commitment to the ideology of behavioral management strategies.

**Postsurgical Speech Therapy**

Many patients undergo surgical intervention for velopharyngeal dysfunction with or without the benefit of a comprehensive presurgical evaluation. Some of these patients will present postoperatively with continued symptoms of velopharyngeal dysfunction. In these cases, comprehensive evaluation is even more critical than in the preoperative situation. Depending on the approach to presurgical speech therapy, many, perhaps even most, cleft patients will require continued therapy for remediation of developmental errors, placement and manner errors, compensatory articulation errors, or phonation disorders that could not be remediated presurgically. However, in many cases with persistent velopharyngeal dysfunction, where comprehensive presurgical evaluation was not conducted, it is possible that the etiology of the velopharyngeal symptoms was not properly symptoms may be diminished but residual articulation errors and learned habits persist and must be addressed through behavioral therapy. In other cases surgery may have been the appropriate management choice but failed to provide an adequate closure mechanism. And, in some cases, surgical intervention may not have been the appropriate course of management and therefore, no noticeable change in velopharyngeal function will be evident.

It is often necessary and appropriate to conduct a period of postsurgical evaluation and possibly more diagnostic therapy to understand the "apparent surgical failure". However, it should be understood that such cases will be less likely to occur if adequate differential diagnostic information was obtained and utilized by the surgeon prior to surgery. Said another way, appropriate differential diagnosis and presurgical diagnostic speech therapy should result in a higher percentage of surgical success. This leads to happier patients, satisfied families, and much happier surgeons.

However, if postsurgical therapy appears indicated, it should be based on the same comprehensive evaluation process suggested for the new, unoperated patient. *It should never be assumed "a priori" that the patient has been given an adequate velopharyngeal mechanism that he/she now simply needs to learn to use effectively.* Without data to support such a claim, this assumption is simplistic and dangerous. And, in many cases, postsurgical speech therapy will be of little value. Once again, it is unfair to the patient, the family, and the speech pathologist to relegate the patient to a prolonged period of therapy that has little chance of
effecting improvement or diminishing symptoms. This is a far too common occurrence after an "apparent surgical failure". Too often, the surgeon seems to act as if he/she has run out of options. So, the patient is turned over to the speech pathologist to "fix the remaining problem". This set of circumstances is a setup for failure for both the patient and the therapist.

**Prosthetic Management**

Prosthetic treatment of velopharyngeal dysfunction has been available for many decades. LaVelle and Hardy define satisfactory prosthetic management as follows:

1. An *optimum result* occurs when the prosthesis results in palatopharyngeal port closure during speech production except in association with production of nasal consonants; that is, the resulting pattern of closure would be essentially normal.

2. The result is considered *successful* when there is palatopharyngeal closure throughout speech production.

3. The result is considered only *desirable* when the palatopharyngeal port area is reduced so that incompetence is a relatively minor speech physiology problem.

Improvement in speech is expected, but the multiple speech physiology deficits of many of the patients for whom this management procedure is appropriate frequently negate the goal of normal speech production.

There are two basic types of speech prostheses: speech bulbs (or obturators) and palatal lifts. The first option is used when there is inadequate tissue to achieve velopharyngeal closure, resulting in a gap that can be filled with the speech bulb. A palatal lift, on the other hand, generally is reserved for cases where there is adequate tissue, but the control, coordination, or timing of movements is impaired.

Traditionally and frequently, a speech bulb or obturator is prescribed for a patient when there is clear velopharyngeal insufficiency but there is some contraindication for surgical intervention. Although historically this has been the most common use of a speech prosthesis, prosthodontic appliances can and are being employed in at least two diagnostic situations.

In the first circumstance the patient has been evaluated and shown to have incomplete velopharyngeal closure, either mild or severe. However, for any number of reasons it is unclear to the speech pathologist and other team members whether management of the velopharynx alone will provide a noticeable or significant improvement in speech intelligibility or speech quality. In these cases a trial period of prosthetic management usually will provide the diagnostic information needed to establish a management plan. Essentially then, this use of a prosthesis is a "reversible" test of the effect of surgical management of the velopharynx.
A second role for prosthetic management in the diagnostic process is to employ a prosthesis to determine whether dynamic velopharyngeal activity can be stimulated or improved. Some reports have shown that prosthetic management may improve or facilitate motion of the velum, posterior pharyngeal wall, or lateral walls in some patients. Although these findings are not universal and remain controversial, a trial of prosthetic management may be appropriate in some patients for whom little or no velopharyngeal movement is noted, and surgical intervention, therefore, would require near or complete obstruction of the nasopharyngeal airway. If improved muscle or structural function can be demonstrated, this information might be useful in altering an existing diagnosis or management plan.

Additionally, in cases of patients with dysarthria (either congenital or acquired), it is possible in some cases for management of the velopharynx with a palatal lift to free other vocal tract components to begin to function more normally.

As has been discussed earlier in this chapter, the velopharynx is only one valve in a series of interrelated valves that make up the vocal tract. For some patients with complex, multifactorial speech production disorders, as in cerebral palsy or other conditions that include paralysis or paresis of the vocal tract, management of the velopharynx may stimulate activity of other vocal tract components as well as activity of the actual velopharyngeal mechanism itself. In selected cases such as these, a trial period of prosthetic management will provide critical diagnostic information about the continued course of management.

Although there have been advances in materials and techniques for manufacturing the actual prosthetic appliances, the dental specialist historically has been dependent primarily on perceptual judgments of speech to determine the function and fit of a prosthesis in a given patient. Oftentimes this was an arduous process, requiring multiple visits to both the prosthodontist and the speech pathologist. With the advent of more objective, instrumental methods, several reports emerged describing the benefits of instrumental methods for facilitating prosthesis manufacture and assessing prosthesis fit.

Perhaps the greatest advance, which apparently has increased both interest in and utilization of speech prostheses, is the increasing availability of fiberoptic nasendoscopy in the management of patients with velopharyngeal dysfunction. Direct visualization of the velopharynx in conjunction with auditory perceptual ratings allows more precise evaluation, fabrication, and tailoring of prostheses for palatal management. Flexible fiberoptic nasendoscopy is the method of choice for direct observation of the velopharynx during dynamic speech activity. Endoscopic evaluation prior to fabrication of the initial prosthesis defines asymmetric or unusual closure patterns. This information is used by the prosthodontist in the fabrication of the prosthesis to provide the best possible fit. Video documentation of the endoscopic evaluations also allows review by any specialists involved in the patient's care prior to decisions regarding management. Additionally, if the endoscopic evaluations are videotaped, there is permanent documentation, and successive evaluations allow for accurate assessment of change over time. This approach allows the prosthodontist and speech pathologist to work together closely to provide a highly individualized and tailored prosthesis for the patient.

D'Antonio et al analyzed retrospective data comparing prosthesis fitting with and without the benefit of nasendoscopy used in a team approach with a speech pathologist and
prosthodontist for fitting palatal prostheses. The authors interpreted their data to show that the number of patient visits and the time required to achieve optimal fit were reduced when using direct visualization in a team approach compared with the traditional trial and error approach by clinicians working in isolation. The authors suggested that the reduction of time between initial evaluation and adequate fit benefited the patient and the clinician in time, cost, and particularly, in patient compliance.

The procedure of speech bulb reduction has gained popularity recently. It was first described by Blakely in the 1960s when he suggested that for a small number of patients gradual reduction of the bulb portion of a pharyngeal bulb obturator can result in increased velopharyngeal movements sufficient enough over time that the prosthesis may become unnecessary in some cases. McGrath and Anderson have become the main proponents of this method. Investigators from several centers have reported cases where speech bulb reduction has been effective in improving velopharyngeal function prior to surgical intervention or completely eliminating all symptoms of velopharyngeal dysfunction so that the speech appliance could be discontinued. This method appears to have great clinical potential. Unfortunately, there is still a paucity of experimental and/or prospective data to support many of the contentions made by the proponents of speech bulb reduction as a routine clinical practice. Necessary information is needed regarding the patients for whom this procedure is most likely to be of benefit, objective documentation of change in structural movements, and the long-term stability of improvements in function.

Of most appeal is the claim that prostheses can be used in patients with little or no lateral wall motion to prepare them for pharyngeal flap surgery such that the prescribed flap will not necessarily need to be as wide and potentially obstructive if lateral wall activity is not present. Although this concept has great intuitive appeal and may in the future prove to be correct, there are no prospective data to support or negate this hypothesis at the present time. In spite of the controversy surrounding some of the claims of prosthetic management of velopharyngeal dysfunction, this continues to be an underutilized form of management and is an active and exciting area for future research.

**Surgical Management**

The obvious first goal of any surgical procedure designed to correct velopharyngeal dysfunction is obtaining velopharyngeal competency. The not-so-obvious second goal should be the creation of velopharyngeal competency with minimal obstruction of the airway. The pharyngeal flap and pharyngoplasty operations are antiphysiologic in terms of their effect on nasal airway physiology; that is, some degree of nasal airway obstruction will result. Depending on individual patient differences in etiology of the velopharyngeal inadequacy, anatomy, type of operation performed, and postoperative healing, the degree of nasal airway obstruction will vary from imperceptible to significant.

The maximum resultant airway effect may be obstructive sleep apnea. The head and neck surgeon should correct any mechanical airway obstruction evident on physical examination prior to performance of a pharyngeal flap or pharyngoplasty, so as to lessen the detrimental airway effects of the procedure as much as possible. This may include conservative nasal septal reconstruction, turbinoplasty, and nasal valve reconstruction, especially in the cleft child. Children with large "kissing" palatine tonsils should undergo
tonsillectomy prior to performance of a pharyngeal flap or pharyngoplasty. Prior removal of tonsils improves the technical performance of the pharyngeal flap or pharyngoplasty procedure, as well as avoids obstruction of the ports by large tonsils following healing. Reassessment of speech should follow the tonsillectomy once the oropharynx has completely healed (3 to 6 months), as some children may show significant improvement of hypernasality and nasal emission and may not require further operative therapy. If further surgical therapy is indicated to improve speech following complete healing of the oropharynx after tonsillectomy, instrumental methods may be used to reassess the velopharyngeal defect, allowing "improved tailoring" of the pharyngeal flap or pharyngoplasty.

A rare child with clinical evidence of adenoiditis and recurrent upper respiratory infection may undergo adenoidectomy approximately 3 months prior to the pharyngeal flap or pharyngoplasty (performance of the adenoidectomy at the same time as a pharyngeal flap would not allow access to the nasopharynx in the case of postoperative bleeding). Rarely a child will have hypertrophied lingual tonsils and require laser lingual tonsillectomy.

Following a pharyngoplasty or pharyngeal flap, children who have significant nasal obstruction or obstructive sleep apnea should undergo videonasendoscopy to assess the size of the port (pharyngoplasty) or ports (pharyngeal flap), as well as be reexamined for obstruction at another site that was not appreciated preoperatively.

Numerous methods of surgical therapy have been utilized for the correction of velopharyngeal inadequacy. Five basic categories of operations may be utilized: palatine tonsillectomy, veloplasty with redirection of velar musculature, the double-opposing Z-plasty of Furlow, the superiorly based pharyngeal flap, and pharyngoplasty. The choice of operation is largely based on direct observation of attempted velopharyngeal closure by videonasendoscopy for each individual patient.

Can surgeons consistently obtain adequate velopharyngeal function and maximal nasal airway function by tailoring the surgical procedure to the pattern of velopharyngeal closure for each patient? The surgical techniques described in this chapter attempt to create a tailor-made speech operation to fit each patient's individual velopharyngeal defect. Individual patient differences in wound healing, tissue contraction, and ultimate scar formation are not subject to control by the surgeon and may therefore make the techniques described less precise than one might wish. The final answer to the question will come only from prospective studies involving preoperative and postoperative measurements of speech and velopharyngeal function employing perceptual and instrumental evaluation methods.

Palatine Tonsillectomy

If videonasendoscopy shows inadequate seal between the velum and the lateral pharyngeal walls because of projecting superior poles of the palatine tonsils, tonsillectomy should be considered. This should be a "class act" tonsillectomy, with fine dissection of the tonsil, including the capsule, and without damage to the underlying palatoglossus, palatopharyngeus, and constrictor musculature. Dissection by unipolar cautery is avoided. Bleeding points are controlled with gentle packing and bipolar cautery. Failure of tonsillectomy to correct velopharyngeal dysfunction does not preclude the use of another operation at a later date.
Veloplasty with Redirection of Velar Musculature

The generally accepted definition of submucous cleft palate consists of the triad of bifid uvula, notching of the posterior border of the bony palate with loss of the posterior nasal spine, and diastasis of the soft palate musculature. These findings are usually obvious on physical examination. Congenital palatopharyngeal disproportion has been used to explain the presence of velopharyngeal dysfunction in a child without obvious evidence of clefting, either over or submucous. In this condition, palatal function is inadequate either because the soft palate is too short or the anteroposterior dimension of the nasopharynx is too deep. However, substantial evidence exists to demonstrate that the majority of so-called short palates (congenital palatopharyngeal disproportion) represent unrecognized submucous cleft palate, or at least a microform of over cleft palate: (a) muscle diastasis is visible on the nasal surface of the soft palate during videoendoscopy, (b) muscle diastasis or abnormal insertion of velar musculature is directly visible at the time of the pharyngeal flap operation when the soft palate is transected and dissected, (c) the frequency of middle ear disease approaches that for overt cleft, and (d) the occurrence of short palate in family lines where overt cleft is present.

If a diastasis of velar musculature is present (such as for the case of a classic submucous cleft palate), videonasendoscopy will show either a midline furrow on the nasal surface of the soft palate, or a lack of musculus uvulae bulge, or both. This anatomic defect, coupled with good palatal motion in general and good lateral wall motion, will result in a small midline defect in velopharyngeal closure. These patients can be corrected with transection of the soft palate and redirection of the velar musculature. The operation begins with a midline through-and-through incision of the soft palate, bisecting the bifid uvula if present. The velar musculature is dissected from its attachments to the posterior edge of the hard palate, as well as the overlying nasal and oral mucosa. The dissection should be accomplished with fine forceps and scissors, utilizing adequate illumination and loupe magnification. The velar musculature is thus redirected from an anteroposterior direction to a horizontal direction, and sutured in the midline. The nasal mucosa is closed prior to the suturing of the muscle. Final closure involves uvuloplasty if needed, and closure of the oral mucosa. This technique is best utilized for correction of velopharyngeal dysfunction secondary to the unoperated submucous cleft palate. For the case of a previously repaired cleft palate, another option should be chosen.

Furlow Double-Opposing Z-plasty

In 1978, Furlow introduced a technique of cleft palate repair by double-opposing Z-plasty, which he described in detail. As noted by other authors who have utilized this method, speech results have improved when compared with other palatoplasty techniques.

The surgical technique is not described in detail here, as the surgical steps are thoroughly explained in Furlow's classic paper. Although probably not the best method for correction of velopharyngeal inadequacy following cleft palate repair, Furlow's method may be ideal for surgical repair of the unoperated submucous cleft palate, due to several theoretical advantages. When compared with veloplasty with redirection of the velar musculature, the double-opposing Z-plasty method reorients the musculature and simultaneously lengthens the soft palate. Although palatal length is an inconsistent determinate of velopharyngeal adequacy, healing via Z-plasty may be superior to healing via a straight-line closure that may tether the
soft palate and result in less mobility. Another possible advantage involves the fact that the bulk of velar musculature is relatively small in many children with a submucous cleft palate. Furlow's method accomplished redirection of the velar muscles via separation of the overlying mucosa on only one versus both sides, which may result in less surgical trauma, less postoperative scarring with healing, and thus improve palatal mobility.

**Superiorly Based Pharyngeal Flap**

If videonasendoscopy reveals moderate (approximately one-third the distance to the midsagittal plane) to excellent (two-thirds the distance to the midsagittal plane) lateral pharyngeal wall motion, construction of a pharyngeal flap is probably the best option for surgical management. Patients with minimal lateral wall motion may be candidates, but are at risk for hyponasality and airway obstruction. The degree of competency obtained from a pharyngeal flap will depend on the size of the constructed lateral port, tailored to the degree of lateral pharyngeal wall motion. The goal is to perform an operation that will create openings between the nasopharynx and the oropharynx just small enough to allow competency, yet large enough to minimize the amount of nasal obstruction. Three elements are essential to the creation of a flap that, it is hoped, will achieve this: flap width, height or level of the flap base, and control of the lateral port size.

Based on the knowledge of the defect in velopharyngeal closure (and the degree of lateral wall motion) observed by videonasendoscopy for the particular patient, the width of the flap is constructed relatively thin (for excellent or better lateral pharyngeal wall motion), of moderate width (for moderate pharyngeal wall motion), or relatively wide (for less than one-third the distance to the midsagittal plane lateral pharyngeal wall motion). Based on the preoperative knowledge of the vertical level of maximal lateral wall closure, the length of the flap is determined. The base of the flap is estimated such that the flap will leave the posterior pharyngeal wall at a right angle to join the palate without tension, and at the same time be opposite the position of maximum lateral wall movement. The markings (flap width and length) are completed prior to the incisions.

The soft palate is sharply incised in the midline, from the posterior edge of the hard palate through to and bisecting the uvula. The oral mucosa is incised to a point approximately 2 mm from the posterior edge of the bony palate, whereas the nasal mucosa is incised approximately 0.5 cm from the edge of the posterior hard palate, in order to facilitate later placement of the pharyngeal flap. The tissue contents of the soft palate are carefully inspected at this time, and the presence of muscle fibers are noted, as well as their direction. Children with cleft palates who have undergone previous primary palatoplasty with intravelar veloplasty, will be noted to have muscle fibers that run in a transverse direction as a result of the previously created levator sling. This levator sling will be carefully reapproximated at the conclusion of the procedure. Children with submucous cleft palate will be noted to have the classic misdirection of the levator muscle fibers, curving cephalad into the posterior edge of the hard palate. These muscles are dissected at a later time in the procedure and an intravelar veloplasty is performed.

The previously marked lateral borders of the pharyngeal flap are incised through mucosa. The divided palate allows easy access into the nasopharynx for extension of the incisions in a cephalad direction to define the level of the flap base. By scissor dissection, the
incisions are extended deep through the underlying superior constrictor muscle to the plane of the alar layer of prevertebral fascia. Blunt dissection is completed in the plane between the superior constrictor muscles and the alar layer of prevertebral fascia that connects the two lateral incisions. Good hemostasis is obtained, and the nasopharynx and soft palate are carefully inspected a third time. The lateral incisions are carried to a point cephalad in the nasopharynx, such that the base of the flap will be opposite the position of maximum lateral pharyngeal wall motion, as noted previously by nasendoscopy. A useful landmark is the prominence of the first cervical vertebra. If lateral pharyngeal wall movement is relatively high in the nasopharynx, the base of the flap must also be high in the nasopharynx, opposite the level of maximum lateral pharyngeal wall motion. Once the final base of the pharyngeal flap is selected, the length of the flap is estimated and the lower end of the flap is then transected.

Turnover flaps of soft palate nasal mucosa are outlined and dissected. The plane of dissection is between the nasal mucosa and submucosa and the musculature of the soft palate. The borders of the turnover flaps are created beginning several millimeters from the peak of the nasal mucosa incision and carried approximately 1.5 cm directly lateral. The incision is then turned 90° inferiorly and laterally into the area of the upper portion of the posterior tonsillar pillar. The incision may be extended into the lateral pharyngeal wall as necessary, depending upon the intended size of the lateral pharyngeal port (as described below). If the patient has a submucous cleft palate and the levator muscles are abnormally inserted, they are dissected free at this time. Scissor dissection is used to create a plane between the levator muscles and the overlying oral mucosa. The abnormal insertion of the muscle fibers into the posterior hard palate is transected and the flap of fibers is moved posteriorly until they line a transverse plane.

The flap is attached to the palate. Different sizes of standard endotracheal tubes are used to gauge the size of the lateral port. Small pediatric endotracheal tubes (sizes 3 to 3.5) are used in patients who demonstrate minimal to moderate lateral wall motion. Larger endotracheal tubes (sizes 4 to 4.5) are used in patients who demonstrate moderate lateral movement, and sizes 5 to 5.5 are used in patients who demonstrate excellent lateral wall motion. The flap is attached to the nasal layer of the palate, beginning at the anterior peak and progressing laterally to the level of the lateral port. The final "lateral port control stitch" attaches the appropriate point of the lateral edge of the flap to the appropriate point of the incision in the superior aspect of the posterior tonsillar pillar or lateral pharyngeal wall, so that the tissues are snug around the endotracheal tube.

To assist adaptation of the flap to the palate, a single absorbable suture is placed, grasping the flap muscle layer in the midline, and brought out through the peak of the palatal incision and tied over the oral mucosa. The turnover flaps are then sutured into place so that they line the raw inferior surface of the pharyngeal flap. The flaps may be turned in an arc - anywhere from 90° to 180°, and trimmed appropriately for the final fit. The nasal mucosa on the posterior surface of the uvula is then approximated. Mattress sutures are used to close the divided levator sling, or to join the muscle fibers if an intervelar veloplasty has been performed. Several of the sutures may incorporate the underlying flap muscle layer to aid in adaptation of the flap to the palate.
The anterior oral mucosa is closed. The posterior pharyngeal wall defect is then closed. The defect is sutured loosely toward the base of the pharyngeal flap, to avoid tubing of the flap base and unnecessary strain on the lateral port.

Good postoperative care is paramount for the success of pharyngeal flap surgery and includes room mist, adequate hydration, intravenous antibiotics, and close monitoring of the airways. The endotracheal tubes used to control the lateral port size are removed the morning after the surgical procedure.

**Pharyngoplasty**

If videonasendoscopy shows relatively good velar movement and poor to absent lateral wall motion, pharyngoplasty may be the best surgical option for rehabilitation of velopharyngeal inadequacy. The technique described and illustrated is similar to that described by Jackson. A horizontal incision is accomplished in the posterior pharyngeal wall, opposite the level of maximum attempted closure of the velopharyngeal port, as previously viewed via videonasendoscopy. Bilateral superiorly based mucosal-palatopharyngeus muscle flaps are elevated, with the medial incision connected to the horizontal posterior pharyngeal wall incision. The mucosal-palatopharyngeus muscle flaps are turned 90° and sutured end to end in the midline posterior pharyngeal wall. The technique narrows the nasopharyngeal dimension laterally and posteriorly so that the velum can make contact. It is important that the mucosal muscle flaps are placed opposite the level of the maximum velopharyngeal port closure.

**Conclusion**

In summary, the goal of this chapter was to explain the need for *differential diagnosis* in the evaluation and management of velopharyngeal dysfunction. Through comprehensive, multimethod evaluation, appropriate diagnosis can lead to effective individualized management. It is the role of the speech pathologist to collect and interpret as much data as necessary to establish a provisional differential diagnosis and to interact with other medical specialists, prosthodontists, and surgeons, to develop an appropriate intervention strategy. In brief, the message of this chapter is *differential diagnosis is the first step toward differential management.*
Residual cleft lip deformities may result from avoidable errors in evaluation, planning, and execution of the primary repair, or from the severity of the initial deformity, which may preclude a perfect result. Postoperative long-term management, eg, orthodontics and prosthodontics, influences the result. Residual deformities rarely improve with age. The most important cleft lip repair is the first one. Growth magnifies asymmetry. Imperfection present in microform following primary repair produces increasing deformity and asymmetry with age.

Residual defects appear as changes from the norm in shape, configuration, proportion, and symmetry. The inevitable cutaneous scar may be hypertrophic or contracted. Without being able to articulate the problem, the lay person will be drawn to an disturbed by even slight asymmetries of the lip.

The goal of secondary lip repair is the same as that of primary repair: to create a lip as closely resembling normal as possible. Specifically, there should be the following:

1. a normal Cupid's bow with peaks of matched height and width,
2. normal and symmetrical philtral width,
3. normal and symmetrical philtral height (vertical lip length),
4. normal and symmetrical vermilion fullness in the lateral lip element,
5. a normal vermilion tubercle with appropriate fullness and protrusion,
6. a smooth vermilion-cutaneous border (white roll), and
7. a normal nostril sill and alar base insertion width.

Achieving these depends on accurate skin, muscle, and mucosal approximation with reorientation of the orbicularis oris, and production of minimal scar.

Surgical timing for soft tissue defects alone, such as lip scar, abnormal muscle bulge, and white roll mismatch, may be done as early as 12 months post-primary repair. Treatment of defects involving multiple components, such as short lip, tight lip, and long lip, must be integrated with dental, orthognathic, and maxillary treatment, and with treatment of functional defects, eg, velopharyngeal incompetence.
If possible, maximal aesthetic correction should be achieved by age 5 to 6 years when peer influence begins to influence psychological development of self-esteem and self-concept.

**Cutaneous Scars**

The vermilion produces a consistently minimal and soft scar that rarely requires revision for its own sake.

Lip skin also generally produces a good scar. Marked hypertrophic scarring is rare. Mild hypertrophic scarring that produces a red, firm, and raised scar and shortens the philtral ridge and lip height is fairly common.

Prevention of scar hypertrophy is aided by minimal skin manipulation and avoidance of deep dermal sutures. Resection of 2 to 3 mm of muscle from each segment prior to muscle approximation removes tension from the skin closure and allows skin approximation without strain.

Primary repair done in infancy produces the best scar. Reopening the lip for secondary procedures should be avoided if possible.

Hypertrophic scarring should be initially treated with time, patience, and massage. Scar relaxation and return to normal vertical lip height may take a year to 18 months. If the lip is permanently shortened or disfigured by a wide, hypertrophic scar, five possible treatments exist:

1. The scar may be simply excised with reapproximation of skin edges and careful attention to white roll alignment.

2. If the scar contracture has produced significant lip shortening, a complete revision is indicated. Scar excision must be accompanied by readvancement and rerotation of the flaps with lengthening.

3. Dermabrasion and/or laser smoothing may be used to improve the scar appearance.

4. A severely scarred philtrum may be replaced with a full-thickness skin graft.

5. Z-plasties may be used to reorient the scar. However, this distorts the philtral line and philtral groove, produces dimpling, and is not generally recommended.

**Vermilion Deformities**

Three vermilion deformities commonly occur. Superior vermilion defects exist at the white roll vermilion-cutaneous junction as irregularities of the smooth lip line. At the inferior lip border there may be a deficiency of tissue appearing as a notch, or whistle deformity, or an excessive tissue appearing as overfullness and protrusion. In the midline, deficiency appears as an inadequate tubercle or whistle deformity. Inferior border problems may involve vermilion alone or vermilion, muscle, mucous membrane, and skin.
Vermilion cutaneous white roll irregularities appear as a protrusion of skin intruding on the vermilion or of vermilion intruding on skin. The irregularity may be slight to marked.

Prevention is achieved by careful marking of the vermilion cutaneous junction before local infiltration with epinephrine and its resulting branching and by careful approximation. Tattoo marking of the white roll with two points, one above and one below the junction, makes alignment easier and more accurate than using one marking point alone.

Correction of vermilion cutaneous irregularities may be achieved in the following ways:

1. Excising the errant pigmented tissue and reapproximating the vermilion cutaneous border using a small diamond-shaped excision is especially effective for narrow fingers of skin or vermilion intruding onto the contrasting pigmented area.

2. Z-plasties of equal or unequal length can reorient the disoriented tissue. However, this can produce a large amount of scar in a small area with a lumpy result.

3. A complete revision of the lip for other component abnormalities allows correct white roll reapproximation.

4. A white roll graft may be used to correct this problem; however, excessive scar in the area can result.

5. Color-simulated tattoo may be used to camouflage the differences in pigmentation.

6. A low vermilion border, depressed in height with respect to the normal side, may be elevated with full-thickness skin excision and vermilion advancement to the proper height.

Notching of the inferior lip border vermilion is the most common vermilion irregularity. Correction of a minor notch at initial repair can be achieved by a vermilion Z-plasty. The primary cause of this problem is incision of the lateral element flap too far medially along the cleft, beyond the point of maximum vermilion fullness. Here, vermilion, skin, and muscle are underdeveloped. Lip closure then leaves a notch at the point of suture due to thin lateral lip tissues being approximated to fuller noncleft side tissues. Incision of the lateral lip element at the point of maximum vermilion fullness allows approximation of more normal tissues to the noncleft side and prevents notching.

Use of prolabial vermilion in the repair of bilateral cleft lip frequently produces a whistle deformity. Prolabial vermilion is deficient in character, being thinner, less full in texture, and more prone to peeling compared with lateral lip vermilion. Notching or whistle deformity is prevented by discarding the prolabial vermilion and utilizing lateral lip vermilion to produce a tubercle. Overcorrection is difficult to produce. Apparent overcorrection at the end of repair will reduce rapidly in volume, producing a tubercle of sufficient size.
Correction of a unilateral vermillion notch may be achieved in the following ways:

1. Equal or unequal Z-plasty or W-plasty at the muco-vermillion junction will correct small irregular notches.

2. A V-to-Y advancement flap horizontally or vertically oriented will correct slightly greater degrees of notching.

3. If notching is marked or due to incorporation of underdeveloped tissues from the lateral lip element, a complete lip takedown and revision is required with excision of deficient tissues from the lateral lip element and advancement of the flap.

The midline whistle deformity with inadequate tubercle has a multiple tissue deficiency. A midline whistle deformity may be treated in the following ways:

1. A vertically oriented V-to-Y advancement flap will produce mild augmentation of the vermillion alone.

2. A shaved Abbé flap will augment the vermillion tubercle to a greater degree. A full Abbé flap replaces deficiency in multiple layers of tissue.

3. A composite graft may also be used for tubercle augmentation.

4. If prolabial vermillion has been used, it should be discarded and lateral lip vermillion advanced from each side. This essentially requires a complete lip takedown with trimming and advancement of flaps. Inadequate bulk for the tubercle projection is augmented by advancing orbicularis oris into the tubercle area.

Excessive vermillion height or fullness, not associated with malaligned muscle, is treated with excision at the vermillion mucosal junction with appropriate reduction of bulk. This may be elliptical or a W-plasty in shape.

Muscle Deformities

Muscle deformities present as an unnatural-appearing lip with a groove beneath the lip scar and/or a muscle bulge lateral to the repair. This bulge accentuates with facial animation. Groove indentation results from muscle dehiscence or lack of initial approximation. Lateral muscle bulging results from inadequate muscle release from the maxilla with improper muscle realignment or muscle dehiscence with lateral bunching. Inadequate muscle may also present as irregular indentations along the nostril sill.

Prevention is achieved by full release of the malaligned orbicularis oris from the maxilla and full rotation into a normal horizontal position. Muscle approximation should be achieved with permanent suture material.

Correction of muscle deformity requires reoperative with adequate muscle dissection, release from the maxilla, correct alignment, sufficient readvancement, and reapproximation.
Mucosal Deformity

A normal buccal sulcus is necessary for normal upper lip mobility and pout. It is also required for dental and orthodontic access. A tight or inadequate sulcus can result from failure of primary reconstruction, necrosis, infection, or disruption of the mucous membranes. A tight sulcus must be released for a successful secondary lip revision. Failure to do so will limit revision success and may restrict maxillary and dental growth.

Absence of a normal sulcus requires detachment of the lip from the maxilla and provision of an intervening lining for opposing raw surfaces. Lining may be by mucosal flap. Flap coverage does not required donor site closure; spontaneous reepithelization will occur rapidly. Free skin or mucosal graft also provides adequate lining. Graft take is generally excellent. Maintenance of the sulcus postgrafting, however, depends on adequate splinting against graft contraction.

Vertical Height Discrepancy: Short Lip

Vertical height discrepancy is the most common late multiple component deformity of the vermilion cutaneous junction. The peak of the cleft side Cupid's bow must be level with the noncleft side's peak. A minimal asymmetry of even 1 mm is deforming and draws immediate attention to the cleft. Cleft side shortness is much more common than excessive height or length.

Prevention is achieved by producing absolute symmetry at primary operation. A lip that is short at the end of repair will not self-correct with growth. Scar contracture can shorten the lip and produce vertical height deficiency even though correct symmetry is achieved initially.

In the unilateral cleft lip, a short lip usually follows straight line or rotation advancement repair due to inadequate rotation and advancement.

In the bilateral cleft lip, two-stage repair almost always produced mismatch of Cupid's bow peak vertical height. It is extremely difficult, if not impossible, to achieve symmetry in a two-stage repair since scarring of the first site is present in differing stages of contracture at the second stage. A bilateral cleft lip should be repaired in a single stage with prolabial and lateral lip incisions of equal height allowing precise matching of philtral height and Cupid's bow peak positions on each side.

Premaxillary protrusion must be corrected prior to treatment of short lip. Lip shortness may be only apparent and not real. In this case, correct premaxillary positioning will reveal a lip of adequate length.
Corrective techniques include the following:

1. Z-plasties can produce lengthening of 2 to 3 mm. However this produces objectionable scarring across the philtral ridge.

2. Shortness of the vermillion alone can be corrected with a V-to-Y vermillion advancement flap.

3. Philtral shortness can be retreated with a V-to-Y philtral advancement flap. This produces significant scarring at the columella base with frequent dimpling of the superior philtrum. Disruption of the normally smooth columella philtral junction is additionally deforming and not recommended.

4. Moderate to marked shortness requires a complete lip revision with readvancement and rerotation of flap to provide more length. Most repairs can be converted to a rotation advancement repair with excision of old scar and advancement of lateral tissue. Rotation and advancement of lateral lip elements is also possible in bilateral lip revisions.

5. Severe shortness or severe philtral scarring calls for an Abbé flap. Although seldom required in unilateral problems, it is equally useful in this situation. The Abbé flap provides both appropriate philtral length and relaxing width.

**Vertical Length Discrepancy: Long Lip**

A long lip is most commonly seen in bilateral cleft lips and following quadrangular repair of a unilateral cleft.

In bilateral cleft lip repair, excessive length is prevented by not advancing both skin and vermillion lateral flaps below the prolabium. Vermilion only should be advanced, with rare exceptions. Marked cleft width may require maximum advancement of the lateral lip elements. This inevitably produces excessive length as the longer portions of the lip are brought medially to achieve repair.

Treatment of long lip may consist of the following:

1. Subalar resection produces lip shortening superiorly. It may be performed unilaterally, bilaterally below the alar bases and nostril sills alone, or completely transversely across nostril sills and columella. Excision generally must be full thickness including skin, muscle, and mucosa.

2. Lip shortening may be achieved by excision superior to the vermillion. Here again, skin reduction alone is inadequate. A full-thickness excision of skin, muscle, and mucosa must be achieved. If the columella is short or the lip tight, this technique produces a poor result.

3. Superior subalar and nostril sill resections plus midline excisions below the prolabium connected by philtral ridge incisions produces a bull's head pattern and allows independent tailoring of lateral and midline elements.
4. Given the extensiveness of the resections discussed above and the difficulty of accurate approximation of tissues while maintaining bridges of intact skin, a formal complete lip revision with complete redissection should be considered. Excessive length can then be excised in the appropriate area at the superior aspect of the lip flaps below the nostril sill and at the inferior aspect of the prolabium. In all techniques slight overcorrection and overshortening is recommended to counteract postoperative sagging and lengthening.

**Horizontal Discrepancy: Excessive Width**

Horizontal asymmetries are slightly less obvious than vertical discrepancies, but remain very important. The most common error is creation of excessive philtral width.

In the bilateral cleft lip, vertical symmetry is visually spoiled by an excessively wide philtrum. With proper rotation advancement of flaps, it is not necessary to maintain maximal philtral width to achieve sound primary closure. A philtral width normal for the age at which repair is done should be the goal.

In the unilateral cleft lip, the cleft side hemiphiltrum is frequently excessively wide, producing asymmetry. This results from difficulty in identifying the apex of the Cupid's bow concavity and the Cupid's bow peaks. The depth of the bow concavity is often closer to the normal peak than is appreciated. Excessive hemiphiltral width is prevent by purposefully narrowing the philtrum at primary repair by approximately 2 mm. This automatically corrects the tendency to laterally orient the cleft-sized Cupid's bow peak. Narrowing of the philtrum also reduces the amount of rotation required to achieve vertical symmetry.

Treatment of excessive philtral width demands reopening the full length of the philtral ridge scar. Generally appropriate excision of philtral skin alone suffices. However, complete lip revision with muscle excision may also be required.

**Horizontal Deficiency: Tight Lip**

A tight upper lip most frequently follows repairs of a wide bilateral cleft. Tightness occurs in unilateral repairs and more narrow bilateral repairs, if excessive tissue is excised. The lip is stretched tightly across the premaxilla with deficient mobility and softness. Premaxillary and midface anterior posterior growth may be significantly restricted.

Treatment requires addition of adequate soft tissues. A mild deformity may be corrected with readvancement of lateral lip flaps and cheek tissues utilizing the increased height and width of the more lateral tissues. The classic treatment of tight lip remains the cross lip Abbé flap. Flap placement in the bilateral cleft is in the midline with replacement of the prolabium. This may be used to lengthen the columella as required. Care must be taken to create adequate flap height so that the inset line reaches the base of the columella without tension. In the unilateral tight lip, the flap may be placed centrally with replacement of the philtrum, or eccentrically in the original defect. Despite the loss of the non-cleft normal philtral ridge, central placement is preferred, as acentric placement never produces normal landmarks and creates disturbing asymmetry.
Summary

Residual cleft lip deformities are avoidable to a large extent by precise evaluation, planning, and execution. However, even in the best and most experienced hands, perfection is difficult to achieve. About 60% to 70% of cleft lip patients will require a secondary procedure. Each component tissue must be analyzed and treated appropriately. Surgery should be kept elegantly simple with creation of normal landmarks and symmetry.
There is nothing quite so challenging as, or more exasperating than, the attempt to correct the cleft lip nose, be it unilateral or bilateral. Over the years surgeons have added various techniques, the success of which depend upon the degree of the initial deformity, the type of initial repair, and finally the surgeon's own skill.

The Unilateral Deformity

As with any surgical problem, possible solutions lie in an understanding of the underlying deformity. The unilateral cleft lip nose deformity was beautifully described by Huffman and Lierle in their classic 1949 paper.

A composite base view of the typical unilateral cleft lip nose is shown. The nasal tip is deflected away from the cleft; the dome of the cleft side is retroplaced; the angle between the medial and lateral crus is obtuse, producing a "blunting" of the nares; there is a buckling rather than an outward curve of the ala; and there is an absence of the alar-facial groove with attachment to the face at an obtuse angle. In addition, there is real or apparent deficient bony development under the alar-facial attachment. The nares shows an overly wide dorsal extremity, a circumference greater than its fellow, and the entire nares is retroplaced compared to the uninvolved side.

The columella deformities include a shorter columella on the cleft side, a medial crus on the cleft side that is retroplaced, and an obliquely slanted columella toward the involved side of the nose and lip. In addition to all of the above, Berkeley pointed out the bowstring-like contracture of the inner nostril from its apex to the piriform aperture, and finally Millard noted the asymmetry of the axis of the two nostrils, with normal being almost vertical and the cleft nostril almost transverse.

The Bilateral Deformity

There are several specific deformities with a bilateral cleft lip nose. The columella is very short, and the medial crura are displaced into the prolabium. There is poor tip projection because of separation and downward displacement of the alar domes, resulting in a bifid appearance. The angle of the dome is quite obtuse, enhancing the nasal tip flatness. The axis of the nostrils is nearly horizontal instead of vertical, with the resultant appearance of a webbing overhang of the medial alar rims and flaring of the alar bases.

Intranasally there is contracture and deficiency of the vestibular lining, flatness of the nasal floors, and retropositioning of the maxilla. In short, it is about the same appearance as you would get by depressing the nasal tip to the lip with your thumb.
Surgical Objective

The goal of cleft nose reconstruction is the same, whether unilateral or bilateral: a symmetrical, normal-appearing nose in harmony with the rest of the face and compatible with the patient's ethnic background. The projection desirable in a nasal tip in a patient of Irish background is much more than that needed in an Oriental patient. Additional goals include, of course, improvement in nasal function and patient self-image.

Timing

Nobody has the answer for the best time to perform revision surgery. Surgeons develop during their careers a feeling for timing relative to each patient. Such feelings are based on a knowledge of the psychological and physical development of children.

We usually perform nasal tip/columella reconstruction at 4 to 6 years of age, when the children are entering kindergarten and first grade. The rest of the secondary procedures, ie, further nasal tip, lip, and septorhinoplasty procedures, are delayed until the midteen years.

Since the parents are placing a great deal of trust and high expectations on each procedure, we do not perform nasal tip surgery unless there is a chance of significant improvement in appearance and function, for example, as one would expect in columellar lengthening.

More definitive procedures requiring septoplasty, onlay grafting, osteotomies, etc, are carried out at age 16 to 17 for girls and 17 to 18 for boys. A significant factor in the actual timing of the procedure depends upon the severity of the defect and the emotional strength of the patient. Discussions regarding goals, type of procedure, and postoperative course should be had with the patient as well as with the parents. We have not recommended a major nasal procedure on a 13- or 14-year-old even though Ortiz-Monasterio and Olmendo have shown it can be done. In fact, we prefer to wait as long as possible, often until the junior to senior year of high school. By 16 and 17 these young adults may decide if they really want such a procedure. Usually they have been through multiple other operations including orthognathic surgery, alveoloplasty, and orthodontia, so that rhinoplasty becomes the "icing on the cake" and hoped-for final procedure of their facial rehabilitation. There is the occasional teen who simply is not ready at 16 to 18 for revision rhinoplasty. These patients need to know that the operation can be performed at a later date. We subscribe to the approach of Marsh regarding the "need" to perform secondary surgery: "an intervention is 'needed' when the following triad is fulfilled: appropriate physical findings are present, a solution is possible, and the patient desires services." The point is that it is as important to known when to stop operating on these children as it is to know when to start.

Surgical Approaches to the Unilateral Deformity

The keys for correction of the unilateral deformity are symmetry and visualization. The problem of attaining symmetry is shown by the number of surgical procedures that have been proposed over the years by Blair, Joseph, Gillies and Kilner, and Berkeley. Surgeons recognized early on that an internal approach left major areas of the deformity uncorrected.
The scars of external approaches have become more acceptable considering the overall improvement that external approaches afford the surgeon.

During the late 1970s and early 1980s, we used an external columella incision after the technique of Bardach (personal communication, 1973). The incisions extend along the margin of the lower lateral cartilage on each side and continue along the edge of the columella. The two are then joined by a horizontal incision running across the face of the columella at its junction with the upper lip if the columella does not have to be lengthened. Usually lengthening is needed. The incision on the cleft side of the columella is extended down onto the prolabium of the lip. The length of this segment equals the difference between the noncleft and cleft sides of the columella. The longer segment on the cleft side provides for lengthening of the columella. This incision allows for wide elevation of the tip skin to the nasion and excellent visualization of the underlying deformities. The cleft side lower lateral cartilage is completely freed from the vestibular skin and mucosa. In fact, we frequently free the noncleft lower lateral cartilage as well, feeling that this gives the best opportunity to produce symmetry. The cleft side lower lateral cartilage is "recruited" to help lengthen the medial crus on that side. A fleur-de-lis configuration is produced and held with permanent white 5-0 nylon. The tip skin is redraped and suturing completed in a V-Y fashion.

Dibbell, in 1982, published a unique approach utilizing an external incision and excision of tip skin that addresses the deformities of the unilateral cleft lip nose and, of major importance, changes the long axis of the cleft nostril. It builds upon the basic concepts of the Blair alar rotation and the Cronin columella lengthening procedure for bilateral cleft nose deformity. With this procedure there is complete release of the cleft side of the nose. Essentially the entire nostril is freed including the alar cartilage and is rotated as a "sock" or "sleeve" into its new location. The incision is designed to match the noncleft side and a deformed crescent moon of skin above the cleft nostril is removed. The incision then extends inferiorly down the margin of the columella across the vestibule just inferior to the nasal sill and around the ala. The skin over the nasal tip is freed and a pocket created for rotation of the underlying cartilage-nostril complex. A permanent suture may be placed from the new dome to the opposite alar cartilage and/or the opposite upper lateral cartilage to hold the rotated nostril in place. A second mattress suture is placed over a bolster to hold the lateral ala in its new position. A wedge of cleft side lip tissue may be removed to allow the cheek to move medially to fill the space vacated by the cleft nostril.

We found the Dibbell procedure to be elegant, since it addresses a multitude of the previously described anatomic problems, and yet it is primarily a soft tissue operation applicable to most age groups. However, for those performing a limited number of such cases a year, it is not an easy operation to master. We felt that it might be possible to combine the advantages of the Dibbell procedure with that of the external approach rhinoplasty popularized by Goodman and Zorn. The advantages of the combined approach include (a) exposure of the cartilaginous structures without distortion; (b) removal of the edge of the alar rim matching the noncleft side; (c) complete freeing of the cleft nostril from the piriform aperture with rotation into a symmetric position; (d) lengthening of the columella on the cleft side; (e) exact placement of permanent sutures; (f) direct application of onlay cartilage grafts, if needed; (g) correction of associated lip deformities; and (h) excellent exposure for septoplasty and/or bony work.
For the past 7 years we have been using this combined approach incision for the majority of unilateral revisions. General anesthesia is used in all patients. The amount of local anesthetic with epinephrine used in the tip area is kept to a minimum. A crescent moon-shaped piece of alar tip skin is marked for excision. The incision on the cleft side continues inferiorly along the face of the columella about 2 mm from the edge. It extends around just inferior to the nasal sill sweeping laterally into the alar-facial groove. This incision on the cleft side joins a standard external incision on the noncleft side with the typical inverted V. The entire nostril is freed widely from the noncleft medial crus, septum, floor of nose, lip, and piriform aperture. It may be necessary to incise through mucous membrane to allow for complete freedom of rotation. The nasal skin is elevated as far superiorly toward the nasion and laterally over the alar cartilages as possible, to provide enough exposure. Once rotated into position the nostril should sit freely without any tugging on adjacent tissues.

The rotation of the nostril brings the lower lateral cartilage into its normal position, lengthens the cleft side columella, and should create a symmetric tip. The new position of the nostril is maintained with permanent sutures from the cleft ala to the noncleft upper lateral cartilage. Other deep sutures are placed as necessary. Most of the time, a cartilage strut is placed between the medial crura and sutured to them with several sutures. The strut should extend from the level of the anterior nasal spine to just inferior to the nasal domes. A longer strut may produce a visible tenting beneath the tip skin. Occasionally it may be necessary to completely free the lower lateral cartilage on the cleft side from its attachment to the vestibular skin in order for it to move to its normal position. If this is necessary then a permanent 5-0 nylon mattress suture gathering lateral-medial-lateral cartilage and back (after the technique of McCollough and English) is often used. Further onlay septal or auricular cartilage grafts may be applied as needed. Because the nostril moves medially as well as superiorly, the cheek must be advanced; thus the need for excision of a wedge of lip tissue inferiorly to the vermilion cutaneous junction in some cases. However, in a number of patients revision of the lip scar is needed anyway. If the above procedure is being performed on a child, then the skin is redraped and the columella closed with 6-0 black nylon. The intranasal incisions are closed with 5-0 chromic. The nose is taped, a cast applied, and the nostril filled with cellulose cotton.

The cast is removed along with sutures and packing under a short general anesthesia in 5 to 7 days in children. Adolescents are cared for in the office as with any other adult. If tip rhinoplasty is being combined with septorhinoplasty, we like to perform the septoplasty after all of the tip work has been completed but before permanent sutures have been placed. Because of the excellent approach afforded by the external incision, the nasal septum is easily identified and straightened as with any septoplasty. Septal cartilage is harvested and set aside for later use. Suturing of the tip cartilages is then completed as above, followed by hump removal, and medial and lateral osteotomies as needed. Skin closure, taping, casting, and packing are the same.

**Surgical Approaches to the Bilateral Deformity**

The major problem with the bilateral deformity is the short columella. Most of the time it was probably pressed into service to help close the original lip. An additional problem is poor tip projection due to separation and downward displacement of the alar domes. At least they are usually symmetrical.
There are three common procedures for addressing the bilateral deformity: the Cronin, the Millard forked flap, and V-Y advancement flaps of the prolabium, often used in conjunction with an Abbé lip switch. The reader is referred to Cronin's 1978 article for an excellent review of the multiple ways to lengthen a short columella.

The Cronin procedure is best used when there is minimal shortening of the columella, a normal-appearing Cupid's bow of the upper lip, and scars that do not need to be revised. The incisions extend along the alar-facial grooves, sweep medially just inferior to the nostril sills and then into the columella in an inverted V. A parallel incision within the nostril and along the nasal floor creates bipedicle flap on each side. Half-thickness alar wedge excisions may be used to facilitate relocation of the ala. The entire base of the nose is then freed widely and advanced medially to lengthen the columella. The flaps are sutured together in the midline and septal or auricular cartilage grafts may be placed between the medial crura to give additional support if needed.

Millard's forked flap technique is useful when the prolabium is quite wide, the philtral scars need to be redone, and there is deficient skin in the nasal floor. The forked flaps are based superiorly on the base of the columella and include the lip scars plus prolabial skin. Thus, the lip scars are revised, the symmetry of the philtrum improved, and no new scars are introduced into the lip. The lateral incisions can be extended as in the external approach to give enough exposure to reposition the alar cartilage. Wedges from the nostril floors are removed if needed to allow lip closure and decrease alar flaring. It is not as useful for lips that are tight. The procedure does tend to produce a columella that is thick and wide appearing. Secondary reduction and refinement of the columella may be needed.

The third most-common method of columella lengthening is to advance the entire prolabium into the columella closing the upper lip in a V-Y fashion. The incisions can be extended along the margins of the alar cartilages as in an external approach rhinoplasty. There is excellent visualization of the lower lateral cartilages. This is our procedure of choice for nasal tip surgery in the 4- to 5-year-old in whom we anticipate an Abbé flap from the lower lip at the time of definitive rhinoplasty at about age 16 to 18. The lower laterals are completely released from the underlying vestibular skin and "recruited" to form lengthened medial crura and new nasal domes in a fleur-de-lis configuration. If needed, an auricular cartilage strut is placed between the medial crura to give support. Two to three sutures of 4-0 or 5-0 nylon are used in a mattress fashion to suture this new tip complex together. It may be necessary to use additional onlay and plumping grafts. After the skin flap is sutured in place, bilateral mattress sutures are placed abutting the vestibular and nasal tip skin to the tip cartilages over cotton bolsters at the level of the dome on each side. The lip is closed in a V-Y manner. The nostril is packed with cellulose cotton, which is removed at 5 to 6 days. There is usually significant improvement in facial appearance from this procedure. It does have the disadvantage of placing a scar in the midline of the philtrum and may produce further tightening of the upper lip. If the prolabial V-Y flap is used in the 16- to 18-year age group, then routine rhinoplasty or septrhinoplasty can be performed through the same incision. In addition, these patients often have pouty excessive lower lips that can supply a very nice Abbé flap in the midline of the upper lip and not in a lip scar. When this flap is used in conjunction with nasal tip/rhinoplasty surgery, extreme care must be exercised in protecting and observing the airway during the first 2 to 3 postoperative days. Short straws may be placed between the lips on each side to facilitate the airway. The swelling will be significant
and the patient should be forewarned. The lips are separated under local anesthesia at 12 to 14 days.

Summary

Secondary cleft lip nose deformities offer a continuing challenge to the rhinoplastic surgeon. The goal of a symmetrical nose that is harmonious with the rest of the face and functions well is not easily attained. Emphasis has been placed on the use of external incisions, rotation of entire anatomical units, plus supporting and onlay cartilage grafts. Revisions of these major procedures will often be needed and the surgeon must keep in mind the associated problems of the maxilla, mandible, and upper lip.
Clefts of the palate or alveolus are associated with deformities in many hard tissue structures besides the local cleft area. Primary surgery usually concerns soft tissue repairs only, without addressing the underlying bony cleft defect. Furthermore, many hard tissue deformities appear or become more pronounced with growth. Maxillary transverse or sagittal hypoplasia is generally not seen in infancy and may be manifested to varying degrees any time between early childhood and the end of skeletal growth. Likewise, although dental anomalies may appear in both the primary and permanent dentitions, dental defects are generally not of concern until eruption of the first permanent anterior teeth. Thus, hard tissue cleft deformities are almost always addressed secondarily.

The timing of these secondary procedures is important and is influenced by several factors. Surgical scars have been shown to restrict growth in a direction perpendicular to the scar. Consequently, the risk of growth restriction must be weighed against the benefits of surgical intervention during growth. Transverse growth of the face continues until approximately age 8, whereas vertical and sagittal growth proceeds through the pubertal growth spurt. Orthognathic surgery is usually not undertaken before cessation of skeletal growth, to prevent return of maxillomandibular discrepancy due to disparity in growth. Psychosocial factors, however, must sometimes prevail where early surgery would aid a child with seriously low self-esteem.

Dental development and eruption are important factors in determining the best time for alveolar bone grafting. A tooth that erupts into a cleft will be lacking in periodontal support. Grafting therefore must be done before involved teeth begin to erupt. Overall dental development may also be an important factor when planning coordinated surgical-orthodontic treatment. Usually, however, skeletal maturity occurs after completion of the permanent dentition (excepting third molars) and is thus the overriding factor.

### Alveolar/Palatal Cleft

The first step in planning treatment is evaluation. The general influence of the alveolar cleft on the face should be noted. There is often a deficiency of alar base support on the cleft side, and an alveolar bone graft can be used to add bulk to this area. Soft tissue characteristics surrounding the alveolar/palatal cleft should then be examined. The presence, size, and location of fistulae should be determined. Often the fistulae are not immediately evident at the alveolar ridge level and are hidden by redundant gingiva. Here, a small, blunt probe may be helpful in the examination.

The amount of scarring should also be noted. Heavy scarring will make the soft tissue less elastic and, along with the size of bone defect, may influence flap design for an alveolar bone graft. Tissue across a scar may have compromised blood supply and may not be relied
upon for flaps.

The location and extent of the bony cleft must be determined using radiographs. The maxillary occlusal film is the most useful, although periapical dental films may also be informative. Along with the general location, the exact extent of the bony defect relative to the piriform aperture and to teeth in the area must be determined. Teeth directly adjacent to the cleft may be lacking in bone on the root surface facing the cleft, and this deficiency could lead to periodontal defects that in turn could threaten the vitality of the tooth and, rarely, that of the graft. Finally, segment mobility, especially in bilateral clefts, should be carefully documented in order to plan the placement of grafts. Stability of the host bed will enhance graft take. Mobil segments, when present, can temporarily be stabilized using a prefabricated custom occlusal or palatal splint. Alternatively, a heavy, orthodontic maxillary arch wire can be ligated to the teeth to lend support and stability at the completion of the grafting procedure.

Treatment goals of an alveolar bone graft may include the following:

1. create a continuous maxillary arch;
2. provide bony support for facial soft tissue;
3. close the fistulae;
4. allow for eruption of teeth through the graft;
5. provide bony support for teeth;
6. facilitate orthodontic movement of teeth.

A continuous arch lends stability to the bony maxilla and reduces mobility of the segments, especially in bilateral clefts where the mobility of the premaxilla is often pronounced. The reconstructed maxilla also prevents collapse of the arch segments. The graft provides bony support for the alar base and the upper lip. Therefore, grafting should extend to the piriform rim and the anterior aspect of the floor of the nose. It is also advisable to overpack the cleft site with bone. A free, onlay bone graft has a tendency to resorb unless it is stress loaded and stimulated, as in the case where a tooth erupts or is moved orthodontically through the graft.

Finally, it has been noted that fistula closure without placement of an interpositional bone graft is more likely to fail. Autogenous bone used for an alveolar graft may come from one of many sites. More important than the exact site, however, is the composition of the graft to include both marrow and cancellous bone. The more common sites where graft bone is harvested from are the cranium and anterior iliac crest. Membranous bone from the cranium reported revascularizes faster and has less of a tendency to resorb. Advocates for the cranium as a donor site also claim reduced morbidity as an advantage. However, harvesting of cranial bone graft must be performed sequentially before or after dissection of the cleft site. Anterior iliac crest dissection, on the other hand, can be performed by a second surgical team at the same time the recipient site is being prepared, which therefore reduces operatime time.
Furthermore, the anterior iliac crest provides a generous source of particulate marrow and cancellous bone for cleft grafting. This bone is highly cellular, heals rapidly, and is resistant to infection. Patients when ambulating generally favor the donor site side for a short time postoperatively, but usually are able to resume school work within a week of the operation. Rib graft, another common source of bone, is more commonly used for primary grafting. Orthodontic movement of teeth into the graft site may begin in about 8 weeks after grafting with autogenous bone.

Allogeneic and certain synthetic graft material such as porous hydroxyapatite blocks may also be used, and have been found to take well in smaller clefts with good vascular supply. This obviates the need for a donor site in the cleft patient. However, a synthetic graft will not allow tooth movement through its substance, and thus should not be used in patients in whom teeth need to be moved through the graft. Allogeneic bone does allow movement of teeth into and through the graft, provided the cementum or root surface of the teeth is not exposed. Allogeneic bone is slower to heal and is more susceptible to infection and graft loss. The potential for transmission of disease with allogeneic bone must also be considered.

As mentioned earlier, bone grafts must be placed before teeth in the cleft area begin active eruption, with the typical indicator being one-half to two-thirds of root formation, as evidenced on dental radiographs. The patient's age varies with the tooth involved. Since the permanent canine has most often been involved, the optimal age for grafting has been 8 to 10 years of age. At this age, however, the central and in particular the lateral incisor, if present, have already erupted, occasionally into the cleft. As a result, they may lack bony support and are frequently malposed. Therefore, if the cleft is in the area of the lateral or central incisor, the graft should be placed much earlier. A retained deciduous or erupted supernumerary tooth in the cleft site must also be considered when planning a graft. The tooth should first be removed in order to facilitate development of soft tissue flaps, and adequate time should be allowed for mucogingival healing. Usually, this is accomplished 6 to 8 weeks before the graft procedure. If the extraction is done too early, resorption of bone in the socket area may complicate the grafting. Furthermore, eruption of other teeth into the cleft could be stimulated by the loss of the primary tooth.

More recently, there is evidence that early secondary repair of alveolar cleft at age 5 to 6 may provide improved periodontal support of the incisors. As the teeth erupt into the alveolar graft site, they stimulate the alveolus and graft bone, reduce malposition, and bring about a healthy canine eminence and lip support. Patients who have had early secondary repair of their alveopalatal cleft with bone graft do not exhibit a higher incidence of posterior crossbite. This seems to support the theory that lateral growth in the region of the cleft defect is completed at age 5 or 6. What remains is appositional growth, which does not seem to be affected by the repair.

Another subject of ongoing discussion is whether to expand the cleft maxillary arch pre- or postoperatively. Each protocol has its advantages. Expanding the maxilla preoperatively widens the cleft, usually making the grafting easier. Postoperative expansion, however, is said to stimulate the bone graft, which enhances graft survival.

Success of alveolar cleft repair is based on careful planning and development of labial, nasal, and palatal flaps to provide tight closure without tension after placement of particulate
marrow cancellous bone chips. A local anesthetic with 1:100,000 epinephrine is injected by infiltration to the mucogingival tissue to effect vasoconstriction. Depending on the width of the cleft, the vertical incisions are placed more or less on the labial aspect of the alveolar ridge to allow adequate tissue on the palatal flap for closure. These vertical incisions adjacent to the cleft are made sharply to bone at the alveolar crest level and the two incisions connected superiorly into the labial sulcus by a partial thickness, mucosal incision. Blunt and sharp dissection is then carried out superiorly deep to the orbicularis oris muscle to expose the piriform rim. Next the palatal tissue is reflected subperiosteally extending from the alveolar crest to the depth of the fistula. The nasal mucosa that extends from the fistula is elevated from the bony cleft and separated from the palatal flap at this level. It is first closed by resorbable inverted mattress sutures to form the floor of the nose. The palatal flaps are then approximated and closed with continous or mattress sutures. Particulate bone graft is then firmly packed in the cleft from the floor of the nose and piriform rim to the crest of the alveolus. Overpacking of the graft as onlay to the labial bone adjacent to the cleft is advisable, as some degree of graft resorption is inevitable. The labial flaps are then advanced and closed over the graft bone. Releasing periosteal incisions at the posterior aspects of these flaps are usually necessary to allow tension-free advancement and closure.

In bilateral clefts, the repair is essentially the same except for the development of flaps on the premaxilla. Consideration must be given to the blood supply of the segment with its attendant scars from previous soft tissue surgical procedures. Judicious reflection of the mucosal flaps off the premaxilla is essential, and a larger portion of flap tissue for closure must come from the posterior segments. Because of increased mobility of the premaxilla, some form of stabilization is usually necessary at the completion of the procedure. This can be achieved with an orthodontic arch wire. Alternatively, a prefabricated occlusal dental splint can be used and ligated to stable, well-anchored teeth. Postoperative arch expansion, when necessary, may begin 6 to 8 weeks after the procedure before the bone is completely healed. As in conventional arch expansion, the expansion device should be maintained for at least 3 months to allow consolidation of host and graft bone.

Complication and graft failure rate is extremely low in children, especially when autogenous bone is used. Occasionally, small areas of the crestal wound may dehisce with minimal graft loss. This invariably granulates over with gentle cleansing and meticulous wound care and does not affect the final result.

Skeletal Maxillary Constriction

Skeletal maxillary constriction is a frequent finding among cleft individuals. Its etiology is not certain, but generally attributed to the scar tissue from palatoplasty. Details of evaluation and treatment of skeletal dysplasia in children are described in Chapter 23 by Albert, Kuo, and Will. Evaluation of children with cleft deformities are similar, with particular attention directed to a few areas as follows:

During the clinical examination the amount of scarring should be carefully examined. This is to anticipate potential difficulties that may arise during expansion as well as with the extent of skeletal relapse. When surgical expansion is necessary, the surgeon must assess the source of blood supply to the maxilla. In the conventional approach to Le Fort I-type maxillary surgery, surgical access is from the buccolabial aspect, leaving the maxilla to
become dependent largely on the blood supply from the soft palate. Should that be compromised because of previous palatal surgery, a different approach to the maxilla must be considered. In this case, vertical mucoperiosteal incisions in the buccolabial sulcus and tunneling approach to the bony maxilla would be an alternative. Similarly, should surgically assisted palatal expansion (via lateral maxillary osteotomies) be contemplated, the surgeon must be cognizant of the potential for total maxillary osteotomy in the future and design soft tissue and bony cuts accordingly.

Another important finding in an examination is the amount of constriction. This can be determined by measurement on dental casts or on a posteroanterior head film. It should be realized, however, that dental measurements can be misleading due to the varying buccopalatal inclination of the posterior teeth. Measurements of basal bone on a head film are more valid and are preferable. The radiographic maxillomandibular width measures the relationship between the basal bones and is more valuable than any absolute dental measurement clinically (see chapter 23 by Albert, Kuo, and Will).

In a cleft child with maxillary constriction, the deformity can be corrected by conventional orthopedic expansion using a jackscrew device. The pros and cons of pre- versus postgraft expansion have been discussed. The authors generally prefer pregraft expansion as it often allows easier access for dissection and closure of deeper structures. However, the full extent of transverse skeletal constriction in a cleft child may not be realized until maxillary growth has completed. In a skeletally mature cleft patient, maxillary constriction can be treated in two ways: by surgically assisted rapid palatal expansion via lateral maxillary osteotomies or by a multiple-piece Le Fort I osteotomy. Surgically assisted rapid palatal expansion is usually done on an outpatient basis under local anesthesia with conscious sedation. Horizontal bony cuts are made in the lateral wall of the maxilla extending from the piriform rim posteriorly under the zygomatic strut. From there, the cuts are tapered inferiorly to end in the posterior tuberosities. Occasionally, a vertical midline osteotomy is made between the two central incisors when the clefts have been grafted. Since expansion is done gradually using a jackscrew appliance, the inelastic palatal mucosa does not limit expansion as it would during a Le Fort I procedure. Accomplishing expansion at the beginning of orthodontic treatment also allows good dental and arch alignment before any subsequent orthognathic surgery. However, some patients are not good candidates for surgically assisted palatal expansion. Patients who lack good bony support in their posterior maxillary teeth should not have orthopedic forces directed laterally on these teeth. In addition, patients with a significantly rotated lesser segment may not be able to achieve correction of such a rotation with traditional palatal expansion, and may benefit more from exact placement of segments during a Le Fort I surgery.

Maxillary Retrusion

Maxillary retrusion is a common but variable feature among cleft patients. The patient with cleft deformities is subject to other concomitant maxillomandibular skeletal dysplasia such as nasomaxillary hypoplasia, mandibular retrognathia or prognathia, macrogenia, or microgenia. These may either mask or accentuate maxillary deficiency associated with the cleft. Careful evaluation is needed using several methods in order to arrive at the best plan of treatment.
The importance of soft tissue scars has been emphasized. In addition, the presence and functional status of any posterior pharyngeal flap must be ascertained. This is important both from the point of view of perioperative management of velopharyngeal competence as well as adequate mobilization of the maxilla in any reconstructive surgery.

Cephalometrics is obviously a useful method of evaluation. Steiner's Sella-Nasion A point (SNA) angle and Rickett's maxillary depth angle (FH-NA), for instance, are commonly used measurements (see chapter 23 by Albert, Kuo, and Will). These figures should be used cautiously, however, and only in the context of a complete clinical examination. Cephalometric measurements describe only the sagittal prominence of the maxilla, and three-dimensional, clinical examination including the soft tissue is critical. This examination should be both static and dynamic and should include zygomatic support, paranasal fullness, nasolabial angle, upper lip support, and incisor display. Any of these may show evidence of underlying retrusion.

The dental occlusion may also show evidence of maxillary retrusion. With a normal mandible, maxillary retrusion will usually be accompanied by an anterior crossbite and an angle class III malocclusion, in which the maxillary teeth are retruded relative to the mandibular arch. Some degree of dental compensation may occur that may partially mask the severity of the skeletal discrepancy.

Surgical maxillary advancement with the Le Fort I osteotomy is usually indicated for significant maxillary retrusion. Although the same basic technique is used for cleft and noncleft patients, there are some special considerations for cleft patients. A previously placed pharyngeal flap may need to be lengthened or temporarily taken down at the time of maxillary advancement, since its preoperative length may no longer be adequate once the maxilla is advanced. It may also limit adequate mobilization of the maxilla necessary to ensure tension-free advancement and minimize skeletal relapse. Patients without flaps may need them postoperatively, if advancement of the maxilla leads to velopharyngeal incompetence. If indicated, this should be performed 1 year after the maxillary surgery to allow healing and consolidation of bone for maximum stability. Palatal scarring also leads to concerns about blood supply. The typical Le Fort I incision is in the depth of the buccal vestibule and runs from mmmolar to molar, leaving the maxilla to receive its blood supply from collateral circulation in the palate. Alternatives to the conventional approach were described in the previous section. In cases where the anteroposterior discrepancy between the maxilla and the mandible is extensive, a combination of maxillary advancement and mandibular setback should be considered. This may facilitate adequate mobilization of maxillary segment(s) without undue risk of avascular necrosis.

Bone grafting is generally indicated where the amount of surgical advancement exceeds 4 or 5 mm. This is to counteract the relapse tendency that increases with larger advancements, presumably due to increased soft tissue stretch, and, in the case of the cleft patient, from the palatal scar and pharyngeal flap. Corticocancellous bone blocks are placed posterior to the maxilla at the lateral aspects. Porous, synthetic block grafts have also been used with success.

Whether or not a graft is placed, proper intraosseous fixation is vital to maximize stability of the advanced maxilla. Rigid internal fixation using miniplates and screws is now
commonly used and provides excellent stability especially in combination with some degree of interdental fixation.

**Dental Anomalies**

Dental anomalies are a common finding in cleft patients, due to disruption of the dental lamina in which the teeth form. Teeth in or adjacent to the cleft, usually lateral incisors or canines, are frequently missing or malformed. In addition, they may be impacted or displaced.

Congenitally absent teeth may either be prosthetically replaced or the space they would normally occupy may be closed orthodontically. The decision of which plan to follow depends upon the existing posterior occlusion and the amount of crowding. If the molar relationship is class I and if there is little or no crowding, advancing all the posterior teeth to close the space is usually not feasible, and prosthetic replacement will be necessary. If, however, the molar relationship is angle class II or significant dental crowding exists, the space can be closed orthodontically without prosthetic replacement. No matter how feasible occlusally this plan may be, it must be done with a careful eye to aesthetic and function. In the case of a missing lateral incisor, the canine may substitute for the lateral incisor and the first bicuspid for the canine. The canine crown must be altered to look as much as possible like the lateral incisor on the opposite side. In extreme cases both the lateral incisor on the opposite side. In extreme cases both the lateral incisor and the substituting canine need to be crowned dentally in order to achieve satisfactory aesthetics. Likewise, a first premolar has to be altered somewhat in order to substitute for a canine.

When considering prosthetic replacement, several options are available. In the younger patient, either a removable prosthetic appliance or a bonded Maryland bridge would be the treatment of choice. The large pulp chamber of the younger patient's teeth mandate against fixed bridgework in which teeth must be prepared. It is also not generally accepted treatment to use osseointegrated dental implants in an adolescent, due to the lack of knowledge of their behavior with bone growth. One simple but temporary measure is to incorporate an artificial tooth onto the retainer at the end of prosthodontic treatment. This is probably preferable for the first year after debanding, while the occlusion is settling and stabilizing. A bonded Maryland bridge can be placed subsequently. In skeletally mature individuals, there is no contraindication to fixed bridgework or dental implants. Dental implants come closest to a biologic tooth but require adequate thickness and depth of alveolar ridge. For that reason, many cleft patients with a grafted alveolar cleft would not be suitable candidates for implants, unless the implant is placed within a few months of grafting. Fixed bridgework can be aesthetically very pleasing, especially if the contralateral teeth are included.

In contrast to missing teeth, supernumerary teeth are sometimes found in the area of the cleft. Supernumerary teeth can impede the eruption of other permanent teeth and thus should be extracted.

Teeth adjacent to the cleft, notably canines, sometimes become impacted and will not erupt on their own. When this occurs, the tooth must be orthodontically brought into the arch, taking care to always keep bone around the root. Root surface that has lost its covering of bone will no longer adhere to bone, potentially leading to periodontal problems and eventual
tooth loss.

Lateral incisors and other teeth adjacent to a cleft are often malformed. Defects and generalized hypoplasia of the dental enamel are seen, as well as defects in shape and size of the crown. Enamel defects should be carefully monitored, and should be restored if necessary. Deformed laterals can be built up with composite dental resin to their normal size, but this should be delayed until the end of active orthodontic treatment so that the exact size for the crown can be determined after optimal occlusion and alignment have been established.

Treatment Sequencing

It is well recognized that optimal cleft care can be provided by a multispecialty cleft team. An equally important factor is proper timing and sequencing of surgical repair, dental rehabilitation, speech therapy, and hearing evaluation as well as nutritional and psychosocial counseling during growth and development.

Infancy and Early Childhood (Birth to 2 Years of Age)

General goals in the treatment of infants with clefts include facilitating feeding, closure of clefts, and forstering optimal emotional, speech, and language development. Many considerations enter into the timing decision: nutrition and feeding, speech development, and facial growth. The timing of lip and palate closure varies somewhat, but lip closure is generally performed between 6 weeks and 6 months, and palate closure between 8 and 20 months. One adjunctive procedure used in some centers is the placement of a palatal plate in the newborn period. This plate, fabricated from resilient and hard acrylic, is made from an impression that can be taken on the awake neonate. The plate's resilient flanges are trimmed to provide retention under the palatal shelves, and the hard acrylic palate extends to the height of the alveolar ridges. The plate performs three functions: First, it allows easier feeding by temporarily separating the communication between the oral and nasal cavities; it also forms a shelf against which the tongue can push during feeding. Second, it maintains the width of the posterior segments and prevents the collapse that often occurs. Third, after primary lip repair, the lip can mold the anterior alveolus while the posterior segments are stabilized. The plate is worn full time until the time of palatal repair or, in some instances, until primary alveolar bone grafting.

Childhood (Age 2 to Age 10)

The importance of speech development is addressed in Chapter 13 by D'Antonio and Crockett. During this period, as permanent teeth begin to erupt, much of the treatment attention turns to dental concerns. Treatment goals during childhood center on optimizing dental health as well as alignment, shape, position, and integrity of the maxillary arch. During this time, growth discrepancies between the maxilla and mandible may become apparent, and treatment may be undertaken to correct them. Palatal expansion using teeth as anchors may be done in the primary dentition, in the early mixed dentition before resorption of primary molar roots, or after eruption of the permanent dentition. Maxillary protraction may also be used to orthopedically advance the maxilla if necessary, but alveolar bone grafting is also done at this time, before two-thirds of the root development of involved teeth and even earlier in some cases, as previously described.
Minor tooth alignment may be carried out during later childhood, both for the purpose of aesthetics and for the purpose of allowing optimal flap development during alveolar grafting. If possible, this should be carried out as a distinct first phase of orthodontic treatment and completed at a predetermined end point. When minor orthodontic tooth movement is allowed to creep into full treatment, which usually continues until the cessation of growth, patients often become "burned out" and develop negative attitudes toward the deformities as well as toward treatment.

Adolescence (Age 11 to Cessation of Growth)

During adolescence, rapid pubertal growth gives way to the stability of skeletal maturity. Treatment goals at this stage focus on achieving final, stable maxillary position and dental occlusion. Definitive orthodontic treatment should be done at this stage, accompanied by any necessary secondary grafting and palatal expansion not already accomplished. Palatal expansion should be done as early as feasible, since fusion of circummaxillary sutures after puberty makes orthopedic expansion difficult or impossible, and surgical assistance must be obtained to reduce lateral resistance to expansion. Orthognathic surgery, when indicated, is usually delayed until the end of skeletal growth in order to maximize stability. Soft tissue revisions on the lip and nose can then be performed on a stable skeletal base.

Adulthood

Although this phase of treatment is out of the scope of this text, it should be mentioned in passing for completeness. In the context of a lifelong sequence of treatment, the goals of treatment in adulthood are to achieve the final, proper skeletal and occlusal relationships. Orthognathic surgery is done after skeletal maturity is attained and orthodontic treatment is completed; then, definitive prosthetics can be done, along with further soft tissue surgery as needed.

For the patient who begins treatment as an adult, alveolar bone grafting and palatal expansion can also be done at this stage. Surgical assistance will be required to accomplish palatal expansion, however, and alveolar grafting should only be done if benefits can be realized for the particular patient. If the maxillary segments are stabilized, the teeth have good periodontal health, and no tooth movement in the cleft area is anticipated, then alveolar bone grafting may not be indicated except to aid closure of any residual oronasal fistulae. If, however, teeth have to be moved in the cleft area or if dental implants are contemplated, alveolar bone grafting will be an important factor in achieving the desired treatment outcome.
Rhabdomyosarcoma

Rhabdomyosarcoma is the most common soft tissue sarcoma in infants and children. The peak incidence of rhabdomyosarcoma occurs in children between 2 and 5 years of age, and a second peak is seen in teenagers between 15 and 19 years of age. In the final report of 686 patients from the First Intergroup Rhabdomyosarcoma Study (IRS), the median age at diagnosis for a child presenting with rhabdomyosarcoma of any primary site was 7 years; 4% of patients were less than 1 year of age at presentation; 38% were younger than 5 years of age, and 63% were 10 years of age or younger.

Rhabdomyosarcoma occurs somewhat more frequently in boys than in girls, at a ratio of approximately 1.4:10. The annual incidence of rhabdomyosarcoma in the USA has been estimated to be 4.4 per million in white children and 1.3 per million in black children.

The most common site of origin for rhabdomyosarcoma is in the head and neck - the orbit, eyelid, and cranial parameningeal region (35%), followed by the genitourinary tract (21%), extremities (19%), trunk (9%), retroperitoneum (5%), and others.

The conventional histopathologic classification system proposed by Horn and Enterline in 1958 is still utilized widely today, though there have been many recent challenges to this system as a result of advances in the sensitivity of cytologic and histologic techniques.

Embryonal rhabdomyosarcoma is the most common form of childhood rhabdomyosarcoma, accounting for 56% of all cases, and the majority of pediatric head and neck rhabdomyosarcoma, including orbital primaries. Embryonal rhabdomyosarcoma is a tumor of early childhood, most commonly presenting between 3 and 12 years of age. The histomorphology of embryonal rhabdomyosarcoma usually consists of a mixture of small, spindle-shaped cells, with tapering bipolar cytoplasmic extensions, and small round to oval lymphocyte-sized cells with little cytoplasm.

Botryoid rhabdomyosarcoma is a subtype of embryonal rhabdomyosarcoma. The botryoid refers to the gross appearance of these tumors, which are found beneath the mucosal lining of hollow organs or of body cavities, or the nasopharynx, and which manifest clinically as grape-like exophytic masses protruding into the adjacent lumen. Histologically, these lesions have a hypocellular to acellular myxoid central region with a densely cellular peripheral region just below the overlying mucosa known as the "cambium layer". Although interesting, these tumors are quite rare in the head and neck, and account for only 5% to 6% of pediatric rhabdomyosarcoma at all sites.
*Alveolar* rhabdomyosarcoma, sometimes called the rhabdomyosarcoma of adolescence, typically occurs in individuals 10 to 25 years of age and most commonly is found in the trunk or extremities. Most series show approximately 20% or less of head and neck rhabdomyosarcomas to be of the alveolar subtype.

The histomorphology of alveolar rhabdomyosarcoma classically shows a framework of interlacing fibrous trabeculae that separates small nests of quite undifferentiated round or oval cells in which the central cells are loosely cohesive, forming a pattern that microscopically mimics pulmonary alveolar spaces.

*Pleomorphic* rhabdomyosarcoma, often referred to as the adult type of rhabdomyosarcoma, is very rare in the pediatric population, being found in less than 1% of all pediatric patients included in the IRS-I and IRS-II studies. Its histomorphology is variable, classically showing large anaplastic cells, multinucleated tumor cells, spindle and strap cells, and tumor giant cells. They often occur in extremities. The diagnosis of pleomorphic rhabdomyosarcoma is becoming rare, even in adults, as more of these tumors are now classified as pleomorphic malignant fibrous histiocytomas.

Juvenile-type embryonal and alveolar rhabdomyosarcoma, nonetheless, do occur in adults, and must be considered in the differential diagnosis of small cell tumors in the adult population. The biologic behavior of these tumors in younger adult patients has been shown to be similar to their behavior in the pediatric population.

As with any histopathologic categorization system that is applied to a spectrum of neoplasia, the conventional system as outlined by Horn and Enterline for rhabdomyosarcoma cannot be applied to all related tumors. Nearly 20% of the 1,626 specimens that were included in the IRS-I and -II studies could not be classified in these four conventional categories. Extraosseous Ewing's sarcoma (5%), small round cell sarcoma, type indeterminate (STI) (8%), and another group described as unclassifiable sarcomas (NOS) (5%), are other recognized categories of small cell mesenchymal sarcomas that are included in the treatment protocols and analyses of the intergroup rhabdomyosarcoma studies I and II.

The staging system for rhabdomyosarcoma utilized in the multicenter, cooperative group trials of the IRS is the most popular staging system in use today in the USA (Table 1). It is a classification system that depends largely on the results of the initial surgical procedure, and is therefore of limited initial value prior to surgery. Additionally, there is no way to incorporate variables such as tumor histologic subtype, tumor size, and tumor location. The IRS is attempting to compare this surgico-pathologic staging system to a new pretreatment, modified TNM staging system (the IRS TGNM system), which accounts for the other important variables previously mentioned in regard to tumor behavior. Other staging systems for pediatric rhabdomyosarcoma, some based on pretreatment criteria alone, also exist.

The histologic subtype of rhabdomyosarcoma has been shown to correlate with patient survival. Patients with alveolar rhabdomyosarcoma and the lesser known small round cell sarcoma, type indeterminate (STI), show decreased survival compared to the other histologic subtypes of rhabdomyosarcoma. Patients with embryonal rhabdomyosarcoma and extraosseous Ewing's sarcoma have intermediate prognosis, and patients with the botryoid variant of embryonal rhabdomyosarcoma have the best prognosis for survival.
### Table 1. Surgical-pathological grouping system used in intergroup rhabdomyosarcoma studies I and II

<table>
<thead>
<tr>
<th>Group</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Localized disease, completely resected (regional nodes not involved)</td>
</tr>
<tr>
<td></td>
<td>A. Confined to muscle or organ of origin</td>
</tr>
<tr>
<td></td>
<td>B. Contiguous involvement with infiltration outside the muscle or organ of origin (as through fascial planes)</td>
</tr>
<tr>
<td>II</td>
<td>A. Grossly resected tumor with microscopic residual disease; no evidence of regional node involvement</td>
</tr>
<tr>
<td></td>
<td>B. Regional disease, completely resected (regional nodes involved and/or extension of tumor into an adjacent organ); all tumor completely resected with no microscopic residual tumor</td>
</tr>
<tr>
<td></td>
<td>C. Regional disease with involved nodes, grossly resected, but with evidence of microscopic residual tumor</td>
</tr>
<tr>
<td>III</td>
<td>Incomplete resection or biopsy with gross residual disease</td>
</tr>
<tr>
<td>IV</td>
<td>Distant metastatic disease present at diagnosis (lung, liver, bones, bone marrow, brain, and distant muscle and nodes).</td>
</tr>
</tbody>
</table>

Additionally, alveolar rhabdomyosarcoma has shown the highest proportion of distant metastasis and the lowest occurrence of local progression. Alveolar rhabdomyosarcoma also may manifest a higher rate of regional lymph node metastasis, but this trait is debatable.

The behavior and prognosis of rhabdomyosarcoma depends in large part on the site of the primary tumor. In the head and neck region, rhabdomyosarcoma most commonly presents in the orbit, followed in descending order of frequency by the pharynx (nasopharynx, oropharynx, and hypopharynx), the soft tissues of the head and neck, the temporal bone, and the paranasal sinuses. Other areas of presentation include the oral cavity, parotid gland, infratemporal fossa, nasal cavity, and larynx.

More than one-fourth of head and neck sarcomas arise in the orbit. When the primary tumor is in the orbit or eyelid, the presentation is usually a progressive unilateral proptosis in a child less than 13 years of age. The requirement for surgery is limited to biopsy, and the survival rate with multiagent chemotherapy and radiotherapy is excellent (93% 3-year survival). Regional lymph node metastases are unusual because these tumors usually present early, and because the orbit is scantily supplied by lymphatics.

Parameningeal sites include the nasopharynx, middle ear and mastoid region, nasal cavity, paranasal sinuses, infratemporal fossa, and pterygoid fossa. Parameningeal rhabdomyosarcoma is of special interest because of the potential for direct spread to the meninges and into the central nervous system. Clearly, meningeal involvement at diagnosis is an unfavorable prognostic sign. Of 57 patients with nonorbital parameningeal sarcoma from the IRS-I study, 20 patients (35%) developed meningeal sarcoma and 90% (18/20) died of this
complication. This has led to the development of "intensive" IRS treatment protocols involving aggressive meningeal radiotherapy with whole cranial or craniospinal radiation, multiagent systemic chemotherapy, and intrathecal chemotherapy administered via lumbar puncture. Even with such aggressive treatment, the 3-year survival among IRS patients with nonorbital parameningeal tumors with meningeal extension has been 57% compared with 90% among those patients with meningeal involvement. Extensive erosion of the craniofacial bones or skull base, or both, independent of meningeal involvement, appears to be an important predictor for local treatment failure in controlling nonorbital rhabdomyosarcoma of the head and neck. Identified risk factors for meningeal involvement are intracranial extension of tumor, bony erosion of the cranial base, and cranial nerve palsy.

Nonorbital nonparameningeal pediatric rhabdomyosarcoma occurs in the neck, parotid region, cheek, masseter muscle, oral cavity, oropharynx (usually the tonsil or soft palate), larynx, hypopharynx, scalp, face, and pinna. As a group, these tumors have been shown to have an intermediate prognosis. However, patients with a primary tumor in the neck were found to have a significantly worse prognosis, with only a 54% relapse-free survival rate, being more likely to relapse locally or distantly. Nonorbital rhabdomyosarcoma of the head and neck has a low incidence of cervical lymphatic metastases (approximately 3%); as a result, prophylactic radical neck dissection is not recommended.

Management of a suspected head and neck rhabdomyosarcoma in a pediatric patient should begin with a diagnostic evaluation, including a complete history and a thorough physical examination. Laboratory studies should include a complete blood count, urinalysis, chest x-ray, and blood chemistry profile including creatinine, bilirubin, uric acid, alkaline phosphatase, serum glutamic oxalacetic transaminase (SGOT) or AST, and lactic dehydrogenase (LDH).

Metastatic spread of rhabdomyosarcoma occurs both by lymphatic and hematogenous routes, the most common sites of hematogenous metastatic disease being the lung, bones, and bone marrow. Metastases to the lungs are frequent, and are evaluated best by computed tomography (CT) of the chest. Evaluation for osseous metastases is accomplished with conventional bone scans and bone marrow aspirate.

Regional tumor extent can be evaluated radiographically with CT, looking for evidence of skull base erosion or other extensive bone erosion. Magnetic resonance imaging (MRI) also is helpful in delineating the precise extent of these tumors. Rhabdomyosarcoma enhances after administration of MRI contrast media and shows a high-intensity signal on T2-weighted images. Unfortunately, other relatively common tumors of the head and neck such as lymphoma and nasopharyngeal carcinoma often also have the same signal intensity characteristics as rhabdomyosarcoma. Surgical management should include an attempt at complete tumor extirpation if possible without inflicting major functional disability. At times it is ill-advised to attempt complete removal of the tumor at initial surgery. In these situations, the initial surgical procedure simply will be a biopsy of the mass. Biopsy alone is advocated for orbital rhabdomyosarcoma because of the excellent long-term results with combined radiation and chemotherapy without surgical extirpation. In certain circumstances, particularly with parameningeal head and neck rhabdomyosarcomas, definitive resection of the tumor may need to be withheld until at least a partial course of chemotherapy and radiation therapy has been given.
The importance of obtaining clear surgical margins without microscopic residual disease remains at issue. Survival curves from IRS-I data indicate that patients with microscopic residual disease postoperatively (group II cases) fare no worse than patients without any residual disease (group I). The presence of macroscopic disease postresection, however, has been a poor prognostic sign in all IRS studies.

Prior to IRS studies, radiation dosages in the range of 6.00 cGy were given routinely to patients with rhabdomyosarcoma. Though local control rates with such radiotherapy were promising, long-term complications were excessive. In IRS-I, -II, and -III, radiation dosages in the range of 4,000 to 5,500 cGy were found to result in effective local tumor control. Dosages less than 4,000 cGy led to increased local recurrence rates. Radiation therapy is currently recommended to all patients with residual disease following surgical resection. Data from IRS-I indicates that radiation treatment is not necessary in patients with group I disease. IRS studies currently are piloting the use of hyperfractionated radiation therapy (5,940 cGy) for patients with advanced stage disease. With such therapy, long-term consequences of high-dose radiotherapy may be minimized.

Chemotherapeutic agents with usefulness in rhabdomyosarcoma include vincristine, actinomycin D, cyclophosphamide, cis-platinum, doxorubicin, etoposide, ifosfamide, and melphalan. Each has shown efficacy in single agent trials. The combination of vincristine, actinomycin D, and cyclophosphamide (VAC) has become the foundation from which IRS chemotherapy studies are evaluated.

In IRS-I, patients with group I disease exhibited an 82% to 84% relapse-free survival when treated with VAC (irrespective of radiotherapy). Patients with group II disease did as well when treatment with triple agent VAC was compared to treatment only with vincristine plus actinomycin D (without cyclophosphamide). Furthermore, patients with group III and group IV disease found no additional benefits when anthracyclines (Adriamycin) were added to their VAC regimen. Three-year relapse-free survivals of 60% to 70% were seen in these advanced-stage patients.

In IRS-II, the VAC regimen was intensified by repeating the three drugs over 4-week intervals during 2 years of therapy. The benefits of using anthracyclines again were examined in IRS-II. To date, overall survival rates for all patients treated with IRS-II appear to be significantly better than for those patients treated with IRS-I. In IRS-III, the addition of cis-platinum and etoposide to VAC regimens is being examined. In pilot studies for upcoming IRS-IV protocols, trials of melphalan plus VAC, ifosfamide plus etoposide plus VAC, and ifosfamide plus doxorubicin plus VAC are being studied in advanced stage disease. Finally, most regimens recommend the use of central nervous system prophylaxis in patients with meningeal involvement with rhabdomyosarcoma. Prophylaxis generally consists of some combination of radiotherapy and triple agent intrathecal chemotherapy (hydrocortisone, cytosine arabinoside, methotrexate). Per IRS guidelines, patients whose tumors extend intracranially from the primary site should receive both cranial radiotherapy plus intrathecal therapy. In patients with cerebrospinal fluid involvement, the radiation should include the entire craniospinal axis. Patients whose tumors do not extend intracranially, but do cause cranial nerve palsies or erosion of the skull base, should receive extended field radiotherapy to the site of disease plus intrathecal therapy. Patients with parameningeal tumors that do not extend intracranially, do not erode the skull base, do not cause cranial nerve palsies, and do
not involve the cerebrospinal fluid, do not require intrathecal chemotherapy or cranial irradiation. They should receive extended field radiotherapy to the primary site, however.

Additional Comments Regarding Management

In cases of suspected rhabdomyosarcoma, preoperative consultation with the hematology-oncology service and the pathology department should be obtained. Anderson et al recommend that the initial surgical procedure in a case of suspected rhabdomyosarcoma be performed at a tertiary care center so that adequate facilities for thorough histopathologic evaluation are available. At times, fresh tissue is needed for advanced tumor marker studies. Frequently, electron microscopy, which requires special fixation of the tissue in solutions such as glutaraldehyde, is necessary to achieve a diagnosis in these cases. In addition, if frozen section suggests the tumor to be rhabdomyosarcoma, a bone marrow aspiration and biopsy can be performed at the time of surgery to evaluate the status of the bone marrow and, if a parameningeal site is involved, the child can undergo a diagnostic lumbar puncture during the same anesthetic administration.

The IRS-III program is collecting data that will determine whether there is a difference in the prognosis of group III patients whose tumors have been debulked at the time of initial biopsy in cases where total tumor extirpation is not a feasible option. The incomplete surgical removal (debulking surgery) will be categorized as less than 50% tumor excision versus greater than 50% tumor excision.

Additionally, it is hoped that the IRS-III study will provide information regarding the efficacy of secondary operations, wherein a delayed resection of the primary tumor is performed after the first 20 weeks of treatment.

Nonrhabdomyosarcoma Soft Tissue Sarcomas

Soft tissue sarcomas of childhood may be divided into two general groups: rhabdomyosarcoma and nonrhabdomyosarcoma (non-RMS) sarcomas. Each group accounts for about 50% of soft tissue sarcomas in children. Non-RMS soft tissue sarcomas of the head and neck region include fibrosarcomas, alveolar soft part sarcomas, synovial sarcomas, hemangiopericytomas, and neurofibrosarcomas. These lesions typically present as firm, slowly enlarging masses. Associated symptomatology is determined by the location and degree of mass effect exerted by the tumor itself. Both local extension and hematogenous metastases commonly occur.

Two age peaks, one in infancy/early childhood and the other in adolescence, are common with these tumors. Prognosis, in general, appears to be related to the age at presentation. Non-RMS soft tissue sarcomas of infants and young children often exhibit a benign behavior, requiring surgical excision alone. Non-RMS soft tissue sarcomas in adolescent patients are typically aggressive in nature, and require multimodal treatment.

Wide local excision remains the mainstay of treatment for all non-RMS soft tissue sarcomas. The ability to achieve a gross tumor resection is probably the most important determinant of disease-free survival. Obviously, total resections are not always possible in managing these tumors. For cases in which surgical resection is incomplete (with either
microscopic or macroscopic residual disease), adjuvant therapy is required.

For congenital fibrosarcomas in young children, there is no evidence that either chemotherapy or radiotherapy is needed in the primary treatment of grossly resected tumors. Radiation therapy in dosages of 4,000-6,000 cGy may be used for local control of unresectable disease. Chemotherapy regimens such as VAC (vincristine, dactinomycin, and cyclophosphamide), or ifosfamide plus etoposide may be used in treating unresectable or metastatic congenital fibrosarcoma. Fibrosarcomas of young adults commonly employ VAC-type regimens for primary disease control.

Grossly resected neurofibrosarcomas, like congenital fibrosarcomas, are treated best with surgery alone. No benefits have been found in using adjuvant chemotherapy after gross resections have been performed. Chemotherapy for unresectable primary or metastatic disease remains ill-defined. Trials combining doxorubicin, vincristine, dactinomycin, and cyclophosphamide have shown only marginal results in treating neurofibrosarcomas. Ifosfamide and etoposide are currently being studied in neurofibrosarcomas. Radiation therapy remains useful in achieving local control for both microscopic and macroscopic residual disease.

In contrast to other soft tissue non-RMS sarcomas, hemangiopericytomas are best treated by wide local excision in combination with chemotherapy. The high incidence of metastatic disease and the excellent response to chemotherapy have led many investigators to utilize chemotherapy in controlling primary disease. Cyclophosphamide, vincristine, methotrexate, doxorubicin, and mitoxantrone all have been tried with some success. Radiotherapy is reserved for local control of microscopic and macroscopic residual disease.

Alveolar soft part sarcomas remain extremely difficult to treat. Though 80% of patients will be alive 2 years following surgical resection, the majority of patients will eventually relapse and die of progressive disease. The most frequent sites of occurrence of alveolar sarcomas are the orbit and tongue. Females are affected more commonly than males. As many as one-third of all patients may present with metastatic disease prior to even identifying the primary lesion. Bone, brain, and lungs are the most common sites of metastases. Current initial management relies upon gross surgical resection. Chemotherapy is rarely used in the treatment of grossly resected primary disease. Responses to radiation therapy have been reported to be poor.

Synovial sarcomas occur most commonly in young adults, and rarely in children. Due to its rarity, treatment guidelines for pediatric patients with synovial sarcomas remain unclear. Combinations of wide local excision with high-dose radiotherapy (> 6,000 cGy) are being compared to excision alone. The role for adjuvant chemotherapy is not yet documented for young adults with resected disease. VAC chemotherapy regimens have shown some promise in unresected or recurrent disease.

A full discussion of osteogenic sarcomas and Ewing's sarcomas is beyond the scope of this text. Osteogenic sarcomas and Ewing's sarcoma comprise over 80% of all malignant bone tumors in children. Though extremity and trunk lesions are much more common, skull, mandibular, and maxillary lesions may occur in both tumors. As extensive debulking procedures are not often possible upon presentation, a limited resection for diagnostic
purposes is usually the initial surgical procedure performed. Adjuvant chemotherapy is then given for several courses before a more definitive resection is attempted. Multiple courses of chemotherapy subsequently follow. High-dose methotrexate, cis-platinum, and doxorubicin have shown high response rates in osteogenic sarcomas. VAC regimens with doxorubicin, and ifosfamide/etoposide regimens, have commonly been used on Ewing’s sarcomas. Whereas osteogenic sarcomas are highly radioresistant, Ewing’s sarcomas are radiosensitive.

**Neuroblastoma**

Neuroblastoma is the most common malignancy in infants under 1 year of age. About 500 new patients present with neuroblastoma annually in the USA.

Neuroblastoma arises from fetal neural crest cells of the sympathetic nervous system. The biologic behavior of neuroblastoma is one of the most fascinating of all pediatric malignancies. Spontaneous regression, as well as spontaneous and induced maturation to benign ganglioneuroma, is seen. Neuroblastoma most frequently arises in the adrenal medulla and presents as a hard abdominal mass. Other presentations include respiratory symptoms secondary to posterior mediastinal disease, urinary obstruction secondary to spinal cord compression from "dumbbell"-shaped paraspinal neuroblastoma extending through the vertebral foramen.

Only 2% to 4% of all neuroblastomas are cervical primaries. Primary cervical neuroblastoma most often occurs during the first 6 months of life and may be present at birth. Neuroblastoma may present as a firm indolent lateral neck mass. There may be associated feeding and respiratory disorders (hoarseness, stridor, dysphagia) secondary to mass effect on the pharynx, esophagus, larynx, or trachea. Ipsilateral Horner's syndrome from involvement of the cervical sympathetic ganglia and heterochromia of the iris representing a related neural crest cell abnormality may be presenting signs. Metastatic disease to the neck from more inferior primary sites is a more common etiology of cervical neuroblastoma and this possibility always must be considered.

Approximately two-thirds of all children with neuroblastoma have metastatic disease at presentation. Neuroblastoma spreads via local lymphatics as well as hematogenously to bone, bone marrow, and liver. Metastatic neuroblastoma to the head and neck may present as proptosis and periorbital ecchymosis if orbital infiltration has occurred. Bone metastases may present as masses over the skull and mandible. Subcutaneous metastatic nodules are bluish, nontender, mobile, and may be widespread.

Recommended metastatic evaluation includes CT, radionuclide bone scan, and metaiodobentylguanidine (MIBG) scanning. MIBG is a norepinephrine analogue that is taken up by neuroblastoma tissue and can be labeled with a radionuclide to detect primary or metastatic disease. The urine should be checked for elevated catecholamines and bone marrow aspiration may be appropriate. The classic staging system for neuroblastoma proposed by Evans in 1971 is based on the extent of disease (Table 2).
Table 2. Evans staging system for neuroblastoma

I  Tumor confined to the organ or structure of origin.
II Tumor extending in continuity beyond the organ or structured of origin but not crossing the midline; regional lymph nodes on the homolateral side may be involved.
III Tumors extending in continuity beyond the midline; regional lymph nodes bilaterally may be involved.
IV Remote disease involving bone, parenchymatous organs, soft tissues or distant lymph node groups, or bone marrow.
IV-S Patients who would otherwise be stage I or II but who have remote disease confined to one or more of the following sites: liver, skin, or bone marrow (without evidence of bone metastases).

Treatment

Surgery, radiation therapy, and chemotherapy all play a role in the treatment of neuroblastoma. Stage of the disease, as well as the age of the patient, influence the therapeutic regimen and prognosis. Children under 1 year of age have the best prognosis. Localized neuroblastoma that can be resected without gross residual tumor and with which there is no regional lymph node involvement usually is treated surgical resection alone. It is important to sample regional lymph nodes, if identifiable, at the time of surgery for staging purposes. Microscopic residual disease does not appear to worsen the prognosis significantly.

Stage II neuroblastoma also is treated initially by careful surgical excision, with a 90% disease-free survival rate unrelated to size of the primary tumor, intraspinal involvement, extent of resection, and subsequent treatment with radiotherapy or chemotherapy, or both. In general, chemotherapy is not recommended as primary therapy in these patients, but may be used to salvage patients who relapse. Neuroblastoma is relatively radiosensitive and radiation has been included as part of the treatment for many patients with group II and III disease. The optimum radiation dose is not known as there is not a well-defined dose-response curve for the tumor. Tumors have been treated with doses ranging from 900 to more than 4,000 cGy, together with systemic chemotherapy. Patients with metastatic disease or poor prognostic features have been treated with total body irradiation, high-dose chemotherapy, and autologous bone marrow rescue, giving long-term survival rates of 40% to 50% in selected patients.

Patients with stage III or IV neuroblastoma require multiagent chemotherapy with or without radiation therapy. Delayed surgery often is employed. Chemotherapeutic agents used for neuroblastoma include cyclophosphamide, doxorubicin, cis-platinum, etoposide, and vincristine. Purged autologous, or allogenic, bone marrow transplantation, following high-dose chemotherapeutic preparative regimens, is reserved for patients who fail standard chemotherapy.

The management of stage IV-S neuroblastoma remains controversial. Treatment approaches range from supportive care only to surgical removal of the primary tumor. Chemotherapy is debatable, but probably is required in patients with massive liver or bone marrow metastases.
Esthesioneuroblastoma (olfactory neuroblastoma) is a related tumor of the olfactory area, thought to be of either neuroectodermal or neural crest origin. Twenty percent of cases occur in childhood or adolescence. Management is primarily with surgery and radiotherapy, usually with radiotherapy following surgery. Preoperative chemotherapy and radiation therapy followed by surgical resection have been promoted in advanced cases. Recently, good results have been reported with craniofacial resection alone, without radiotherapy, in patients diagnosed with esthesioneuroblastoma limited to the nasal cavity without evidence of cribriform plate erosion.

**Thyroid Cancer**

Thyroid cancer is an uncommon disease in childhood. The annual incidence in the USA is 5 cases/million/year for children less than 20 years of age. Girls are affected more than boys and the majority of children affected are over 10 years of age.

The association of well-differentiated thyroid carcinomas with prior radiation therapy became apparent in the 1950s. In the 1940s, radiotherapy was frequently used to treat tonsillar and adenoidal hypertrophy, cystic hygromas, and hemangiomas. This led to an epidemic of thyroid cancer in children. Thyroid cancer also has been shown to follow radiation for other malignancies, in particular Hodgkin's disease.

Other factors implicated in the development of thyroid cancer include prolonged thyroid-stimulating hormone (TSH) stimulation and thyroiditis. Medullary carcinoma of the thyroid (MCT) most often occurs in patients with multiple endocrine neoplasia (MEN) type II. These are autosomal dominant disorders. Patients also develop pheochromocytoma, hyperparathyroidism and, in type IIb, mucosal neuromas.

Four histologic types of thyroid cancer are described: papillary, follicular, medullary, and anaplastic or undifferentiated. Papillary carcinoma of the thyroid is the most common type and represents 80% of cases in children. Follicular adenocarcinoma is the second most common type representing 5% to 10% of thyroid cancers in children. Many tumors have mixed papillary and follicular elements and are classified under papillary carcinoma. Treatment and prognosis for papillary, follicular, and mixed type are similar, so they are often grouped together as well-differentiated carcinoma of the thyroid. Medullary carcinoma of the thyroid develops from the parafollicular or C cells of the thyroid, and represents about 5% of childhood cases. Anaplastic or undifferentiated carcinomas are rare in children but are highly aggressive and metastasize widely.

Children with thyroid cancer usually present with a solitary thyroid nodule or cervical adenopathy, or both. Most patients (70%) will have cervical metastases at diagnosis. Pulmonary metastases are present in about 15% of patients at diagnosis. Bone metastasis can also occur. Patients with MEN IIb also may demonstrate dysmorphic facial features and mucosal neuromas.

The majority of patients are euthyroid at presentation with normal T3, T4, and TSH levels. Antithyroid and antimicrosomal antibodies are absent unless the patient has an associated autoimmune thyroiditis. Thyroglobulin levels may be elevated and can serve as markers for response to therapy or relapse. Screening tests for baseline calcitonin as well as
calcitonin levels following calcium and pentagastrin stimulation are recommended for all children with a family history of MEN IIa or IIb. Routine screening can lead to early diagnosis and treatment of MCT in these patients.

Soft tissue x-rays of the neck may show calcifications in patients with well-differentiated carcinomas. Chest x-ray should be obtained due to the high incidence of pulmonary metastases. Ultrasound studies can differentiate cystic from solid masses and can separate discrete nodules from diffuse enlargement of the gland. Radioisotope scans using I-131 and technetium pertechnetate are more helpful diagnostically. Cold nodules are highly suspicious for malignancy and must be excised. Management of hot nodules is more controversial because they are less likely to be malignant. However, because of the small risk of malignancy, some surgeons recommend biopsy of all solitary nodules in children. Fine needle aspiration is frequently used for diagnosis in adults, but there is much less experience with this technique in children. Evaluation for metastatic disease should include CT scan of chest and bone scan. Currently, there is no universally accepted staging system for thyroid cancer.

Surgery is the mainstay of treatment for thyroid carcinoma. The extent of surgery needed in well-differentiated thyroid carcinoma remains controversial. Current recommendations range from simple lobectomy with isthmusectomy to total thyroidectomy with radical node dissection. Major complications from total or near-total thyroidectomy include lifelong replacement therapy with thyroxine, damage to the recurrent laryngeal nerve, and hypoparathyroidism. However, partial thyroidectomy limits the usefulness of radioactive iodine in detecting occult metastatic disease; thus, some surgeons prefer to perform total thyroidectomy in all patients.

In patients with evidence of nodal involvement, extensive lymph node dissection, as well as total thyroidectomy, is indicated. Total thyroidectomy is also indicated in patients with gross bilateral disease and metastatic disease so that radioactive iodine therapy can be used.

Patients with a family history of MEN II should be screened routinely for medullary carcinoma of the thyroid. The screening tests consist of stimulation with calcium and pentagastrin followed by measurement of immunoreactive calcitonin. When calcitonin is elevated, total thyroidectomy with lymph node dissection is indicated. When lymph nodes are positive, a modified radical neck dissection with preservation of the sternocleidomastoid muscle should be performed.

For anaplastic carcinoma, total thyroidectomy with lymph node dissection should be performed due to the local invasiveness of this neoplasm.

The ability of many differentiated thyroid carcinomas to take up iodine introduces the possibility of selectively targeting these tumors using radioactive isotopes of iodine. Iodine 131, given systemically, may be used following surgery for papillary and follicular thyroid cancers. Radiation from this type of treatment is well confined to the tumor with relatively small amounts of radiation delivered to other areas of the body. In instances where there is gross residual tumor, particularly when it does not take up iodine, external radiotherapy with doses in the range of 4,500 to 6,000 cGy may be necessary to help control local disease. Anaplastic thyroid carcinomas are usually aggressive tumors requiring multimodality treatment
with surgery, chemotherapy, and high-dose radiotherapy in the range of 5.000 to 6.000 cGy.

Prolonged TSH stimulation of thyroid tissue may contribute to the growth of metastatic tissue. Thus, it is generally accepted that patients with differentiated thyroid carcinoma be placed on thyroid suppressive therapy with levothyroxine for life.

Results of chemotherapy in treating carcinoma of the thyroid are disappointing. Adriamycin alone demonstrates some effectiveness and, in combination with cisplatin, may have additional benefits.

Overall prognosis for patients with well-differentiated carcinoma of the thyroid is excellent. Even though children are more likely to have metastatic disease at diagnosis than adults, their disease-free survival rates approach 90%. Importantly, recurrences can occur as long as 20 years after initial presentation so lifelong surveillance is crucial.

**Lymphomas of the Head and Neck**

Lymphoma is the second most common solid malignancy in children under 15 years of age, second only to solid tumors of the central nervous system. Therefore, the otolaryngologist can expect to encounter lymphomas as the most common extracranial solid tumor of the head and neck in children. Together, Hodgkin's lymphoma and non-Hodgkin's lymphoma account for approximately 12% of all childhood cancer, but Hodgkin's and non-Hodgkin's lymphoma differ so greatly that they are best discussed separately.

**Hodgkin's Disease**

The age at onset of Hodgkin's disease demonstrates bimodal incidence peaks. In industrialized countries, the earliest peak occurs in the mid-to-late 20s, and the second peak occurs in late adulthood. It appears that less than 10% of all cases of Hodgkin's disease occurs in children 15 years of age or younger. Hodgkin's disease is quite rare before age 5 years. From 5 to 11 years of age, boys with Hodgkin's disease outnumber girls 3:1, but by age 17 the male to female ratio becomes 1.5:1, due to an increased incidence in girls at the time of puberty.

**Presentation**

The usual clinical presentation is persistent painless cervical adenopathy, "rubbery" in consistency, and located in the supraclavicular fossa or, commonly, in the lower half of the neck. Upper neck node involvement is less frequent. The nodes may be tender if they are rapidly enlarging. Nearly 80% of children have unilateral or bilateral neck involvement at diagnosis. Approximately two-thirds of patients will have some mediastinal involvement at presentation and a chest radiograph is an important part of the initial evaluation if lymphoma is suspected in order to evaluate for mediastinal tumor and to assess the patency of the thoracic airway. Hodgkin's lymphoma, less commonly, also can present as axillary adenopathy and, in 4% of cases, the primary disease will present in a subdiaphragmatic state such as peripheral inguinal, femoral, or superficial iliac lymphadenopathy.
Splenomegaly or hepatomegaly, or both, indicate advanced disease. The most common extranodal sites of involvement are intrathoracic structures such as pulmonary parenchyma, pleura, and pericardium. Extranodal involvement of these structures usually occurs when the tumor breaks through the nodal capsule to invade these adjacent structures. Extranodal involvement of Waldeyer's ring is very uncommon, but has been reported in Hodgkin's disease.

Approximately one-third of children present with systemic symptoms such as unexplained fevers, night sweats, and unexplained weight loss of 10% in the previous 6 months. These symptoms are felt to be secondary to the elaboration of cytokines by the tumor cells.

**Histologic Subtypes**

According to the currently utilized Rye classification system, Hodgkin's disease is divided into four histologic subtypes (Table 3).

**Table 3. The Rye classification of histologic subtypes of Hodgkin's disease**

- Lymphocyte predominance (LP)
- Mixed cellularity (MC)
- Lymphocyte depletion (LD)
- Nodular sclerosis (NS).

Each histologic subtype has its own characteristic histologic architecture and ratio of apparently normal or reactive cells, such as lymphocytes, plasma cells, and eosinophils, to the cytologically abnormal Reed-Sternberg cell.

In lymphocyte predominant Hodgkin's disease, the cellular proliferation involves benign-appearing lymphocytes. The lymph node architecture may be partly or completely destroyed. The histologic picture may be misinterpreted as benign hyperplasia. Often, several sections must be examined before the diagnostic Reed-Sternberg cells are found.

In mixed cellularity Hodgkin's disease, Reed-Sternberg cells are plentiful; the lymph node architecture is usually diffusely effaced. Focal necrosis may be present. Mixed cellularity subtype is seen most commonly in children 10 years of age or less.

In lymphocyte depletion Hodgkin's disease, there is a scarcity of lymphocytes and a predominance of abnormal cells. Fibrosis and necrosis are common. Lymphocyte depletion Hodgkin's disease is quite rare in children.

The nodular sclerosis variant of Hodgkin's disease is distinguished histologically by a thickened capsule with bands of birefringent collagen, which divide the tumor into discrete nodules and by the lacunar variant of the Reed-Sternberg cell. The nodular sclerosis variant occurs more frequently in adolescents and young adults. It has a propensity to involve the lower cervical, supraclavicular, and mediastinal lymph nodes.
Nodular sclerosis appears to be the most common form of childhood lymphoma, accounting for 39% to 63% of cases in larger series, followed, in descending order of frequency, by the mixed cellularity subtype (19% to 34%), the lymphocyte predominant subtype (13% to 23%), and, finally, the relatively rare lymphocyte-depleted subtype of Hodgkin's disease (1% to 5%).

**Staging**

The Ann Arbor staging classification system, adopted in 1971, remains the most popular system today (Table 4). This system is based on the assumption that Hodgkin's disease usually spreads from one adjacent nodal area to another, at least early in the disease.

If extralymphatic spread of disease remains local, so that it is amenable to treatment by radiotherapy, the substage designation "E" is applied.

**Table 4. Ann Arbor staging system for Hodgkin's disease**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Involvement of a single lymph node (I) or of a single extralymphatic organ or site (IE).</td>
</tr>
<tr>
<td>II</td>
<td>Involvement of two or more lymph node regions on the same side of the diaphragm (II) or localized involvement of an extralymphatic organ or site and one or more lymph node regions on the same side of the diaphragm (IIE).</td>
</tr>
<tr>
<td>III</td>
<td>Involvement of lymph node regions on both sides of the diaphragm (III), which may be accompanied by localized involvement of an extralymphatic organ or site (IIIE) or by involvement of the spleen (IIIS), or both (IIISE).</td>
</tr>
<tr>
<td>IV</td>
<td>Diffuse or disseminated involvement of one or more extralymphatic organs or tissues with or without associated lymph node involvement.</td>
</tr>
</tbody>
</table>

The absence or presence of unexplained fever > 38°C, night sweats, or unexplained loss of 10% or more of body weight in the 6 months preceding admission are denoted in all cases by the suffix letters A or B, respectively.

Initial clinical staging is based upon physical examination, laboratory studies, chest radiographs, thoracic CT scans, bone marrow biopsy, and lymphography. Final pathologic staging can be based only on the results of surgical staging, which usually involves a laparotomy, splenectomy, and multiple liver biopsies. Any liver involvement represents stage IV disease. Laparotomy results in a change of stage in 20% to 30% of patients. Staging laparotomy is a major surgical procedure with significant potential postoperative complications, including intestinal obstruction secondary to adhesions and postsplenectomy bacterial sepsis by encapsulated organisms such as *Haemophilus influenzae* and *Streptococcus pneumoniae*. Nonetheless, surgical staging has been shown to result in improved relapse-free survival compared to clinically staged children. In almost all cases, patients who will be treated with radiotherapy alone should undergo pathologic staging including a staging laparotomy prior to beginning treatment.
Treatment

Treatment strategies for Hodgkin's disease include radiotherapy, multiagent chemotherapy, or a combination of the two modalities. The effective control of disease by these nonsurgical means has limited the initial role of surgery to a diagnostic biopsy. Accurate pathological staging that involves laparotomy is important in treatment planning and is another important role for surgery in the management of this disease.

Hodgkin's disease is a very radiosensitive tumor, and radiotherapy has been used in the treatment of this disease since the 1920s. Current radiotherapy techniques include involved field irradiation (IFI), when radiation is directed to areas of known disease, extended field irradiation (EFI), which involves radiation to additional sites adjacent to the involved area, and total lymphoid irradiation (TLI).

Involved field irradiation alone is felt by some investigators to be appropriate treatment for selected children with favorable presentations of clinically stage I disease and favorable histologic prognosis confined to the upper third of the neck or the inguinal-femoral area. In these patients, laparotomy may not be needed, but due to the scarcity of such patients, this approach has not been analyzed completely. In general, involved field radiotherapy will lead to a high relapse rate in all but the most favorable situations and, although salvage with chemotherapy and further radiation is useful, cure rates after relapse do not approach those achieved when aggressive treatment is administered initially. Most patients with clinical stage I or stage II disease should undergo a staging laparotomy.

Total nodal irradiation has been used alone in the past for treatment of stage III disease, with a 10-year survival rate of approximately 70%, and a relapse-free survival rate of 40% to 57%. Similarly, patients with advanced-stage disease have achieved only 40% to 60% long-term survival rates when treated with chemotherapy alone. Failures in treating advanced-stage disease with either chemotherapy or radiotherapy alone have led to the development of combined modality therapies. These combined programs, such as the use of low-dose radiotherapy and combination chemotherapy, yield the highest cure rates and are applicable particularly to young children with advanced-stage bulky disease (mediastinal masses greater than one-third the size of the thorax) and to those young children who are yet to undergo their major growth and development.

The MOPP regimen (nitrogen mustard, vincristine, procarbazine, prednisone), and the subsequently introduced ABVD regimen (Adriamycin, bleomycin, vinblastine, imidazole carboxamide) are the most commonly used compounds in the multiagent chemotherapy regimens utilized in pediatric Hodgkin's disease. Although the MOPP regimen, introduced in the 1960s, is the most studied and accepted regimen, new trials with ABVD, and MOPP with ABVD plus low-dose involved field irradiation, have shown excellent survival and freedom from relapse in pediatric patients with Hodgkin's disease.

Even though dramatic strides have been made in the treatment of Hodgkin's disease, complications of therapy continue to be a major problem. Arrest of soft tissue and bony growth often occur in irradiation fields in children who have undergone prior irradiation therapy for Hodgkin's disease. Avascular necrosis and slipped femoral capital epiphyses have been reported in children receiving radiotherapy and chemotherapy that has included
Endocrine dysfunction has been associated with both irradiation and chemotherapy. Hypothyroidism is common following neck irradiation. Gonadal toxicity remains a significant problem for patients treated with MOPP therapy or with pelvic irradiation. Ovarian transposition before pelvic irradiation is recommended for affected females. ABVD appears to carry a much lower risk of sterility than MOPP in both males and females. Another major complication affecting treated patients is the increased risk of second malignancies seen in Hodgkin's disease. Patients treated with irradiation alone are at increased risk of developing secondary solid tumors, whereas patients treated with chemotherapy (or combined treatment) are at increased risk for developing acute nonlymphocytic leukemia or non-Hodgkin's lymphomas. Most secondary leukemias and non-Hodgkin's lymphomas occur within the first 5 years from the initial Hodgkin's disease diagnosis, whereas solid tumors are usually seen 10 years or more following initial diagnosis.

In summary, therapy for Hodgkin's disease in the pediatric patient should be curative in intent, and tailored to the age of the child and to the stage and extent of the disease, with concern for possible long-term complications of the therapy.

Non-Hodgkin's Lymphoma

Non-Hodgkin's lymphoma is not a single disease but a heterogeneous group of neoplasms that arise from the constituent cells of the immune system, together comprising approximately 10% of all pediatric cancers. Childhood non-Hodgkin's lymphomas differ significantly from the majority of non-Hodgkin's lymphoma seen in adults, and instead appear to be immunologically closely related to the childhood lymphoid leukemias.

In contrast to Hodgkin's disease, which has a bimodal incidence curve, the incidence of non-Hodgkin's lymphoma increases throughout life with increasing age. Approximately 97% of cases of non-Hodgkin's lymphoma occur in patients older than 20 years of age. The annual incidence of non-Hodgkin's lymphoma in children younger than 15 years of age is estimated at 9.1 per million in white children and 4.6 per million in black children. Non-Hodgkin's lymphoma occurs 1.5 times more frequently than Hodgkin's disease in childhood.

Children with acquired or inherited immune deficiency syndromes have a greatly increased risk of developing a malignant lymphoma. This group includes those patients receiving immunosuppressive agents associated with organ transplantation, patients with severe combined immunodeficiency, and patients with Wiskott-Aldrich syndrome, and ataxia-telangiectasia.

The etiology of non-Hodgkin's lymphoma, as with most neoplasms, remains unclear. An oncogenic role has been proposed for the Epstein-Barr virus (EBV), since EBV DNA and EBV nuclear antigen have been found in the tumor cells of patients with endemic Burkitt's lymphoma. This, however, has not been a consistent finding in patients with nonendemic Burkitt's lymphoma.

There are many classification schemes for non-Hodgkin's lymphoma. For childhood disease, the working formulation for non-Hodgkin's lymphoma is now widely used. Utilizing
this classification scheme, over 90% of non-Hodgkin's lymphoma can be categorized into three major subtypes, all of which histologically show high-grade malignancy. Small, noncleaved lymphoma (both Burkitt's and non-Burkitt's type), which represents 38.8% of all childhood non-Hodgkin's lymphoma, is usually a B cell-derived tumor. Lymphoblastic lymphoma represents 28.1% of childhood non-Hodgkin's lymphoma and is usually T cell-derived. Large cell lymphoma represents 26.3% of all childhood non-Hodgkin's lymphoma. Follicular pattern non-Hodgkin's lymphoma is almost universally absent in children.

The histopathology of non-Hodgkin's lymphoma is characterized by progressive effacement of normal lymph node architecture by malignant cells and, in non-lymphoid tissue, by infiltration of neoplastic cells between the normal organ-specific cells, collagen, or muscle fibers of involved tissues. The constellation of clinical findings at presentation often correlates well with the pathologic subtype.

The majority of children with lymphoma present with intrathoracic tumors, most often in the mediastinum, which can cause symptoms of dyspnea and dysphagia. Superior vena cava syndrome may develop, with swelling of the head, face, and upper extremities. Low-dose radiotherapy often is required in the initial management of such cases to control the tumor mass.

Patients with small, noncleaved lymphoma usually present with abdominal pain or swelling, a change in bowel habits, and iliac or inguinal adenopathy. Signs may mimic appendicitis. In cases of endemic Burkitt's lymphoma in Africa, jaw involvement is found in approximately 70% of children less than 5 years of age and in approximately 25% of children greater than 14 years of age.

In sporadic cases of Burkitt's lymphoma (nonendemic), jaw involvement occurs in 15% to 20% of patients, and is not age related.

Large cell non-Hodgkin's lymphoma often presents in atypical locations, such as facial, lung, and intracerebral locations as well as in the nasopharynx and bone. Large cell non-Hodgkin's lymphoma are frequently seen in children with inherited or acquired immunodeficiency syndromes and in patients receiving immunosuppressive therapy.

**Staging**

There are several staging systems for childhood non-Hodgkin's lymphoma based predominantly on tumor volume, but no staging system is uniformly acceptable. Tumor burden at presentation is the major determinant of treatment outcome in childhood non-Hodgkin's lymphoma.

Progression of a childhood non-Hodgkin's lymphoma does not follow an orderly pattern of local spread as is often seen in Hodgkin's lymphoma. Instead, rapid dissemination from apparently localized site of disease is often seen. Approximately 30% of patients with childhood non-Hodgkin's lymphoma will have head and neck lymphadenopathy at diagnosis.

In most staging systems, children with non-Hodgkin's lymphoma fall into two broad categories. Those with localized lymphoma or bulky abdominal lymphoma that has been
resected are classified as stage I or stage II. The patients with central nervous system metastases, extensive intrathoracic disease, unresected abdominal tumor, or bone marrow involvement are classified as stage III or stage IV. By convention, children with greater than 25% lymphoblasts in the bone marrow are classified as having acute lymphoblastic leukemia (ALL).

Bone marrow aspiration and lumbar puncture with cerebrospinal fluid cytologic examination are included in the staging workup, but staging laparotomy is not recommended in non-Hodgkin's lymphoma because chemotherapy is utilized universally.

**Presentation**

Approximately 24% of all non-Hodgkin's lymphomas originate in extranodal sites, with 34% of these involving head and neck structures. This compares with 15% of childhood non-Hodgkin's lymphomas, which arise in the head and neck area. Primary tumors arising from Waldeyer's ring may present with middle ear effusion or nasal obstruction. Among adults, the palatine tonsil is the most common extranodal primary site for non-Hodgkin's lymphoma, but less than 5% of pediatric non-Hodgkin's lymphoma are believed to arise in the palatine tonsils or adenoids.

Asymmetric or painless, progressive, persistent enlargement of the tonsils or adenoids without evidence of local infection, and with or without associated atypical adenopathy should alert the surgeon to the possibility of non-Hodgkin's lymphoma.

The presentation of pediatric non-Hodgkin's lymphoma can be quite varied and unusual. Burkitt's lymphoma may present with any or all of the following characteristics: a facial mass, floating teeth, odontalgia, cervical adenopathy, sore throat, or neurological changes such as diplopia, cranial nerve VI and VII palsies, and mental nerve paresthesia.

Non-Hodgkin's lymphoma of the head and neck can be difficult to diagnose even with biopsy, and MRI has proven most helpful in localizing areas of tumor involvement, often facilitating adequate biopsy. Indolent, slow-healing wounds are sometimes seen.

Head and neck non-Hodgkin's lymphoma may present as a mass involving Waldeyer's ring, including the base of tongue, lingual surface of epiglottis, paranasal sinuses, or nose. The nasopharynx is the most common site of origin, with tumor most frequently arising from the fossa of Rosenmüller or from the lymphatic tissue about the torus tubarius.

**Treatment**

The mainstay treatment for all non-Hodgkin's lymphomas, no matter the histologic type or clinical stage, is chemotherapy. Children with non-Hodgkin's lymphoma have a significantly better prognosis when treated with chemotherapy than with radiation alone. Radiation, when given in conjunction with chemotherapy, does not appear to offer any therapeutic benefits. Radiotherapy does have definite application in the treatment of some emergency situations, however, due to tumor mass effects (such as superior vena cava syndrome).
The primary role for surgery in the treatment of childhood non-Hodgkin's lymphoma is to obtain tissue for diagnosis. Tissue should be given unfixed to the pathologist for frozen section, immunologic and molecular studies, and for karyotyping. Surgical debulking of the tumor is beneficial but often not possible. Patients with large abdominal masses in which complete resection is performed have a better prognosis than patients with unresected tumors. Due to the anesthesia and surgical risks in patients with tracheal compression secondary to large mediastinal masses, surgical debulking is rarely attempted in such cases.

All children with disseminated non-Hodgkin's lymphoma should be watched closely for the development of tumor lysis syndrome. Hyperuricemia, electrolyte abnormalities, and renal failure are hallmark features of this complication. Allopurinol administration, vigorous intravenous hydration, and alkalinization of the urine (to increase urinary excretion of uric acid) should be instituted at the onset of therapy. Chemotherapy should begin promptly within 24 to 48 hr from the time of diagnosis. Children with Burkitt's lymphoma who present with large abdominal masses are especially prone to such masses. Deaths in the first 2 weeks of therapy as a consequence of tumor lysis syndrome are not uncommon.

At present, children with non-Hodgkin's lymphomas are categorized into two therapeutic groups: (a) lymphoblastic lymphomas, and (b) nonlymphoblastic lymphomas. Burkitt's lymphomas, large cell lymphomas, and undifferentiated lymphomas are grouped in the latter category. A variety of agents, including cyclophosphamide, methotrexate and vincristine, have shown efficacy in single-drug studies. Recent evidence suggests that intrathecal chemotherapy for central nervous system prophylaxis may be necessary only in patients with head and neck primary tumors.

"Localized" lymphoblastic lymphomas respond well to multiagent chemotherapy. Treatment regimens such as COMP (cyclophosphamide, vincristine, methotrexate and prednisone) or a ten-drug regimen termed LSA₂-L₂ have produced 3-year survival rates over 90%. No differences between 6 months and 18 months of chemotherapy have been noted in treating localized lymphoblastic lymphomas. Treatment of disseminated lymphoblastic lymphomas also has shown excellent promise with multiagent chemotherapy. Unlike localized lymphoblastic lymphomas, there does appear to be a significant advantage to using LSA₂-L₂ chemotherapy instead of COMP in treating children with disseminated disease. One recent trial reported a 76% disease-free survival rate for LSA₂-L₂ treated patients versus a 26% survival rate for those treated with COMP.

Treatment for childhood nonlymphoblastic lymphomas also has been studied extensively using COMP or LSA₂-L₂. Children with disseminated Burkitt's lymphoma (stage III to IV) have showed superiority with the COMP regimen over LSA₂-L₂. Anderson et al reported a 2-year disease-free survival rate of 57% for patients with Burkitt's lymphoma treated with COMP compared with a 28% survival rate for patients treated with LSA₂-L₂. Patients with localized Burkitt's lymphoma (stage I to II) appear to do equally well with COMP or LSA₂-L₂. Since COMP therapy is usually well tolerated, it generally has been favored as the treatment of choice for localized disease. Treatment of localized disease for 6 months of chemotherapy has been shown to be adequate. Much longer regimens have been used in treating disseminated disease.
Salvage therapy for recurrent disease has proven extremely difficult. Autologous or allogenic bone marrow transplantation following reinduction chemotherapy has shown only marginal results in this group of patients.

Posttransplant Lymphoproliferative Disease

Posttransplant lymphoproliferative disease (PTLD) is a relatively new disorder once felt to be related to non-Hodgkin's lymphoma. PTLD is not a single disorder but a spectrum of proliferation of B lymphocytes seen in up to 40% of the immunsuppressed pediatric posttransplant recipients. PTLD can present as a solid tumor, a parenchymal infiltrate, or as an enlargement of lymphoid tissues. Histopathologically and clinically, the process ranges from benign to malignant. There is an association with EBV infection.

In the head and neck, airway obstruction is the most common symptom either from hypertrophy of components of Waldeyer's ring, or from frank intratracheal and paratracheal masses. PTLD may mimic infectious mononucleosis.

The underlying pathophysiology of PTLD is felt to be a loss of T-cell control of B-cell proliferation in these immunocompromised patients (i.e., a failure of immune surveillance). Treatment involves removal of the hypertrophic lymphoid tissue as well as reduction of the immunosuppressant therapy. Mortality approaches 25%.

Nasopharyngeal Carcinoma

Nasopharyngeal carcinoma (NPC), a primary malignancy of the nasopharyngeal epithelium, is a rare tumor in childhood, accounting for less than 1% of all childhood malignancies. The age-incidence curve for nasopharyngeal carcinoma in North America is bimodal, with the first incidence peak occurring between ages 15 and 25. Racial differences in the incidence of nasopharyngeal carcinoma in children are not clearly established. Much work is underway investigating the relationship between Epstein-Barr virus (EBV) and nasopharyngeal carcinoma. Antibody titers against EBV may be useful in the diagnosis and in following the clinical response of nasopharyngeal carcinoma to treatment.

The World Health Organization (WHO) promotes the classification of nasopharyngeal carcinoma into three morphologic groups: keratinizing squamous cell carcinoma (WHO type 1), nonkeratinizing squamous cell carcinoma (WHO type 2), and undifferentiated carcinoma (WHO type 3), a histologically diverse group. WHO types 2 and 3 tumors are common in childhood. Keratinizing squamous carcinoma (WHO type 1), which is less radiosensitive and has a poorer prognosis than the less differentiated tumors, fortunately is rare in children.

Delay of diagnosis is common in pediatric nasopharyngeal carcinoma, often ranging from 4 to 6 months. Cervical adenopathy from regional lymph node metastasis is the most common presenting sign. The node may be tender, mimicking infection. Pain is reportedly the main symptom in children, presenting as otalgia, headache, or pain in the metastatic lymph nodes. Presentation also may include blood-stained mucoid nasal discharge, nasal obstruction, or epistaxis. Serous otitis media secondary to eustachian tube obstruction may occur.
**Treatment**

External beam radiotherapy is the mainstay of treatment for nasopharyngeal carcinoma, with or without chemotherapy. Dosages ranging from 5,000 cGy to 7,500 cGy have led to 5-year survival rates of 20% to 60%. The treatment of the cervical lymph nodes, including supraclavicular nodes, is mandatory, even in the absence of clinical adenopathy. Techniques for radiotherapy include hyperfractionation to minimize long-term side effects, and the use of intracavitary or interstitial boost radiotherapy to the site of the primary tumor to increase local tumor control. Survival rates for those patients with T3 and T4 tumors, or for those with cervical node metastases are significantly worse than those with T1 or T2 disease and no nodal involvement.

Distant metastases are the major cause of treatment failure in children with advanced primary tumors. Common sites of metastasis include the lungs, mediastinum, bones, and liver. Adjuvant chemotherapy therefore has been proposed to help control occult metastatic disease. Most studies indicate that adjuvant chemotherapy is useful in advanced cases of nasopharyngeal carcinoma of childhood, but not all studies have shown statistically significant increase in survival with chemotherapy.

The decision to use adjuvant chemotherapy may not only be based on the extent of the disease at presentation, but also on the dose of radiotherapy to be used. For example, radiotherapy dosages over 6,500 cGy may give excellent local control of nasopharyngeal carcinoma. Yet dosages in this range also may lead to significant long-term consequences in children. In such cases, adjuvant chemotherapy with lower radiotherapy dosages (approximately 6,000 cGy) should be considered. Finally, the timing of chemotherapy and radiation therapy must be considered. Whether chemotherapy should be withheld until after radiation therapy, or given both prior to and after radiation therapy, remains to be determined.

**Langerhans’ Cell Histiocytosis (Histiocytosis X)**

In 1953, Lichtenstein proposed the term histiocytosis X to describe a group of related, overlapping disorders that have in common the idiopathic proliferation or migration of histiocytes with resultant local and systemic effects. Modern pathologic techniques have identified the involved mononuclear phagocytic cell in eosinophilic granuloma (unifocal histiocytosis), Hand-Schüller-Christian syndrome (chronic systemic histiocytosis), and Letterer-Siwe disease (acute systemic histiocytosis) as a Langerhans’ cell, a dendritic cell of bone marrow origin. These three disorders are now collectively referred to as Langerhans’ cell histiocytosis (LCH). There are other disorders of histiocytes in the pediatric population, involving mononuclear phagocytes, other than Langerhans’ cells. These disorders are more rare and are not discussed here.

The etiology of Langerhans’ cell histiocytosis remains unclear, but the disease is believed to be a reactive process rather than a malignant disorder. When generalized, the condition is felt to be multifocal rather than metastatic.

The incidence of LCH is approximately 0.5 per 100,000 children. LCH occurs predominantly in childhood. About 75% of cases occur before 10 years of age and 91% before age 30. LCH can be present at birth, and appears to be more common in Caucasians.
When the diagnosis of LCH is suspected, consultation with the pathologist should precede surgery. Formalin-fixed hematoxylin and eosin-stained tissue can be highly suggestive of LCH, but definitive diagnosis requires either the demonstration of intracellular Bierbeck granules by electron microscopy or of specific membrane immunologic markers. These studies require fresh or frozen tissue in special fixatives for ultrastructural studies.

Presentation

Bony lesions of the skull and associated soft tissue lesions of the scalp are the most common clinical manifestations of LCH in the head and neck. Otorrhea, unresponsive to medical management, is the most common aural finding. Otitis externa and otitis media, with or without mastoiditis and aural polyps, may be seen. Postauricular swelling from a subgaleal mass is reported in 10% to 30% of all patients with LCH. Often, nontender cutaneous erythema is seen. Conductive hearing loss is frequent with temporal bone involvement but sensorineural hearing loss, vertigo, and nystagmus are rare, suggesting that the otic capsule is not prone to histiocytic infiltration. When the temporal bone is involved, 30% of affected patients will demonstrate bilateral disease.

Oral manifestations of LCH include loosening and premature loss of teeth. Alveolar bone loss often is associated with gingival infiltration with histiocytes. Radiographic examination reveals radiolucent lesions with well-defined margins. The mandible is more commonly involved than the maxilla.

Diabetes insipidus often is seen in patients with LCH if a condition other than unifocal eosinophilic granuloma is present. In fact, the triad of diabetes insipidus, proptosis, and membranous bone disease may be seen in Hand-Schüller-Christian syndrome. Infiltrates of abnormal histiocytes and fibrosis have been found in the hypothalamus and posterior pituitary gland of patients with diabetes insipidus, possibly explaining this endocrine dysfunction. Exophthalmos can result from lesions involving the orbital walls and the associated soft tissue reaction.

A characteristic skin rash of the scalp, intertriginous areas, and the perineum and perianal areas often is found. This has been described as a form of seborrheic dermatitis. The evaluation for metastatic disease is coordinated best by a pediatric oncologist and should include a plain film skeletal survey, which is believed to be more sensitive than nuclear scanning in detecting bony lesions. A panorex of the mandible and maxilla should be obtained in patients with oral involvement. Furthermore, an audiologic evaluation should be obtained for those patients with temporal bone involvement.

Clinical Course

The outcome of LCH is variable. The younger the child and the more extensive the disease at time of diagnosis, the poorer the prognosis. A staging system for histiocytosis that considers these factors has been proposed.

The disease tends to be benign and self-limiting when the involvement is limited to only one site. It is likely that solitary eosinophilic granuloma of bone often goes undetected.
Unifocal bony disease in the head and neck can be treated successfully with either surgery or radiation therapy. LCH is a very radiosensitive disease. When surgical removal (curettage) is not feasible, localized radiotherapy can be used. Doses of fractionated radiotherapy in the range of 800 cGy provide excellent local control with little or no side effects. Low-dose radiotherapy also has been used to treat the sella turcica in patients with diabetes insipidus. Results of treatment for this condition are not as clear, but there are reports of reversal of the disease process following irradiation.

An alternative to curettage or low-dose radiotherapy is injection of isolated bony lesions with corticosteroids. In some patients, this has proven to be highly effective, nontoxic, and convenient, with relief of pain within a week or two of initiation of treatment.

Chemotherapy is indicated in almost all patients with disseminated LCH. Solitary bone lesions are rarely an indication in adults, but chemotherapy may be considered in younger children in whom such lesions may herald disseminated disease. Initial chemotherapy is given usually with the combination of daily prednisone and vinblastine given weekly. Response to treatment is evaluated in 2 to 3 months. Failure of response to one or more agents does not preclude response to others. Etoposide, vincristine, cyclophosphamide, 6-mercaptopurine, chlorambucil, and daunorubicin also have shown efficacy in LCH.

Salivary Gland Neoplasms in Children

Primary salivary gland neoplasms occurring in children during the first decade of life, particularly during the first 2 years of life, are predominantly benign. During the second decade, the incidence of carcinoma rises. These are most often mucoepidermoid and acinic cell carcinomas; less often adenoid cystic and other malignancies are seen. The pleomorphic adenoma is the most common primary epithelial salivary tumor throughout childhood.

Neoplasms of the salivary glands are so uncommon in infants and children that the actual incidence of malignant salivary gland neoplasia has been difficult to determine. Any child that presents with a firm salivary gland mass must be suspected of harboring a malignancy. Schuller and McCabe found that 35% of all pediatric salivary gland tumors were malignant. In their review, when vasoformative lesions were excluded, the percentage increased to 57.5%. A more recent study reports a much lower rate of malignancy in solid masses of the parotid gland. Camacho et al reviewed 22 pediatric patients presenting with an unknown solid parotid mass. Only one patient had a malignancy (mucoepidermoid carcinoma). The rate of malignancy of solid parotid masses in their study was only 4%. Benign pleomorphic adenoma, cat-scratch disease, atypical mycobacteria, benign follicular hyperplasia of a lymph node, and toxoplasmosis were other causes of parotid masses in children reported in their series.

Rapid growth, local pain, and facial paralysis are signs that suggest that a salivary gland mass may be malignant. Any child with a persistent or progressively enlarging lump or nodule behind, below, or in front of the ear lobe should be evaluated for a possible parotid malignancy. Furthermore, neoplasms arising in the submandibular glands and minor salivary glands are much more likely to be malignant than are parotid neoplasms.
Mucoepidermoid carcinoma is the most common salivary gland malignancy in children, accounting for 49% of malignant salivary gland tumors. Acinar cell carcinoma accounts for 12% of malignant salivary tumors in children. Undifferentiated carcinoma, adenocarcinoma, sarcoma, malignant mixed tumor, adenoid cystic carcinoma, and squamous cell carcinoma contribute to the balance of malignant salivary gland tumors in children. The peak incidence of mucoepidermoid carcinoma is in the 10- to 16-year-old age group. The first 2 years of life are relatively spared, but mucoepidermoid carcinoma has been reported to occur at 1 year of age. The parotid, by far, is the most common site of occurrence for mucoepidermoid carcinoma; however, the submandibular gland and minor salivary glands of the palate are well-described sites also.

Salivary gland malignancies in children appear to have similar biological activity as those occurring in adults and should therefore be managed according to the same basic principles. Incisional biopsy of salivary gland neoplasms is almost always contraindicated. Fine needle aspirate for cytologic evaluation, commonly used in adults, is also useful in children in some circumstances. Fine needle aspiration cannot be used to rule out malignancy, but can be helpful in preoperative planning if malignancy is suspected. The initial surgical procedure for submandibular masses should be complete excision of the submandibular gland. The initial procedure for parotid lesions should be superficial parotidectomy, if the lesion is limited to the lateral lobe.

Lateral parotidectomy is adequate resection for benign tumors, and, according to some investigators, for low-grade malignant tumors limited to the superficial lobe. Total parotidectomy is indicated if the mass extends to the deep lobe, or if histology reveals a high-grade malignancy. Other investigators feel that total parotidectomy is indicated for all parotid malignancies.

The facial nerve is preserved whenever possible, but must be sacrificed in cases of perineural invasion. If the nerve is resected due to involvement by the malignancy, an immediate facial reanimation procedure should be performed.

The efficacy of cervical lymphadenectomy in the management of pediatric salivary gland malignancy remains uncertain. Formulation of validated guidelines has not been possible due to the relative rarity of these tumors, the variability of histologic types, and the various possible modifications of neck dissection techniques. When metastatic cervical adenopathy is present, neck dissection clearly is indicated. Elective neck dissection, even in patients with high-grade malignancy, is not felt to be indicated by some investigators. Others feel that the initial surgical procedure should involve a parotidectomy with removal of the upper posterior cervical and jugulodigastric nodes, even in clinically negative necks. If frozen section examination of the lymph nodes is positive, a modified neck dissection is performed.

Salivary malignancies of the palate are managed with wide local excision involving en bloc resection. The resultant palatal defects are noted to decrease in size by 50% to 75% over subsequent years, and can eventually be closed if no recurrence is noted after 3 or 4 years.

Postoperative radiotherapy at times is indicated for suspected residual tumor, aggressive histologic subtypes, cervical metastases, and perineural invasion. Severe long-term
radiation complications are common. Palliative radiotherapy may be offered as the sole treatment modality for undifferentiated salivary neoplasms that do not allow surgical resection at the time of presentation. Chemotherapy has been used in combined modality regimens for treating biologically aggressive high-grade salivary gland malignancies.

Late Effects of Therapy for Pediatric Head and Neck Malignancy

Over the last 20 years, strategies to combat childhood malignancies have become more successful. It has been stated that 1 in every 1,000 adults over the age of 20 today is likely to be a survivor of childhood cancer. Some of the most severe long-term effects of chemotherapy and radiation therapy occur in those who have had a malignancy of the head and neck. Long-term survivors of pediatric head and neck malignancies represent a new patient population, often afflicted with difficult and subtle treatment-related adversities that may manifest themselves years after the actual treatment.

One of the most serious delayed consequences of therapy for childhood cancer is the development of a second neoplasm, the incidence rate being approximately 8%. An increased risk of developing bone sarcoma has been found in patients whose primary malignancy was treated with alkylating agents with and without radiotherapy. Radiation to the thyroid gland in doses as small as 120 cGy in a young child increases the risk of thyroid cancer 100-fold.

Endocrine deficiencies after radiotherapy for head and neck tumors are common. Primary hypothyroidism after therapeutic neck irradiation is a well-known sequela. Growth hormone deficiency in adolescence after radiotherapy has been reported widely. Late deterioration of pituitary and hypothalamic function, after 6 and 10 years, respectively, in patients receiving incidental radiation to those structures, has been reported. Central nervous system irradiation also may lead to memory deficits and learning disabilities.

The deleterious long-term side effects of radiotherapy may include clinically apparent stunting of facial bone growth, which has been found to occur in the majority of pediatric patients who have been exposed to 3,000 cGy or more of radiotherapy. As little as 400 cGy of radiotherapy has been shown to arrest soft tissue growth.

The effect of radiotherapy on dental structures is related to both the total dose of radiation and to the stage of dental development at the time of radiotherapy. Radiation doses as low as 400 cGy have been shown to cause abnormalities in developing teeth. Radiation-induced dental deformities include hypoplastic root formation, incomplete calcification, premature closure of apices, and caries. Maxillofacial abnormalities include trismus, midface hypoplasia, other types of facial asymmetry, and malocclusion. Xerostomia and an increased disposition to dental caries may result from salivary gland irradiation. Severely reduced parotid secretory activity has been reported following radiation doses of greater than or equal to 45 Gy to greater than 50% of the parotid gland volume.

Pediatric patients requiring radiotherapy fields that include the eyes, for orbital or paranasal sinus tumors, are known to develop cataracts at doses greater than or equal to 28 Gy; at doses greater or equal to 40 Gy, xerophthalmia is common.
Radiation otitis media is a well-recognized otorhinologic consequence of local radiotherapy to the temporal bone. Although progressive sensorineural hearing loss has been reported after radiotherapy doses of greater than or equal to 60 Gy, more recent work suggests that doses less than or equal to 60 Gy are relatively safe.

Certain chemotherapeutic agents, particularly alkylating agents and procarbazine, are extremely toxic to the germinal epithelium of the tests. Ovarian dysfunction is also a well-recognized complication of therapy with alkylating agents. The surgical management of the deformities resulting from surgical extirpation and radiation therapy of pediatric head and neck malignancies is an evolving field. The problems of poor local tissue vascularity and osteoradionecrosis limit the use of many reconstructive techniques. The increased use of free tissue transfer has proven helpful in managing many of these difficult cases.

Benign Tumors of the Head and Neck

Fibro-Osseous Lesions

There are three general types of fibro-osseous lesions affecting the head and neck in pediatric patients: (a) osseous dysplasia (cementoma), (b) fibrous dysplasia, and (c) ossifying fibroma (cementifying fibroma).

Osseous Dysplasia

Osseous dysplasia is a reactive fibro-osseous lesion that most commonly occurs in the periapical alveolar bone of the anterior mandibular teeth. Radiographically, the lesions may appear as a periapical lucency or as multiple lucencies. Late lesions are radiopaque with a thin, lucent rim. The lesions, also known as cementomas, occur predominantly in black women over 20 years of age, but can be seen in younger patients. Therapy is simple excision.

Fibrous Dysplasia

Fibrous dysplasia is a congenital, metabolic, nonfamilial disorder that accounts for 7% of all nonmalignant tumors of bone. In fibrous dysplasia, normal bone is replaced by irregular trabeculae of immature, poorly mineralized fibrous tissue. The fibrous tissue may lead to bony expansion with distortion of contour and structural weakness. Fibrous dysplasia usually involves the cancellous bone, and therefore usually is found to be covered by a thinned shell of intact cortical bone. Lesion margins at the interface with cancellous bone are usually diffuse and indefinite, as opposed to the well-demarcated lesion interface in ossifying fibroma.

Grossly, the lesions of fibrous dysplasia range from white to gray to pale yellow in color, and from soft and edematous to tough, firm, rubbery, or gritty in consistency. Microscopically, the fibro-osseous tissue is composed of haphazardly arranged strands of collagen stroma and misshapen irregular bony trabeculae that are randomly oriented. Multinucleated giant cells may be found. There is an absence of osteoblastic rimming of the bony trabeculae and, under polarized light, appears woven rather than lamellar. The proportion of fibrous to osseous elements may vary considerably, even within the same lesion. Fibrous dysplasia involves a single bone (monostotic) in 70% of patients, and multiple bones (polyostotic) in 30%. The prevalence of craniofacial involvement in monostotic disease is
10% and, in polyostotic cases, nearly 50%. In McCune-Albright syndrome, polyostotic fibrous dysplasia is found in combination with areas of skin hyperpigmentation and endocrine disturbances (precocious puberty or hyperthyroidism, or both).

The average age of onset of symptoms from fibrous dysplasia is 10 years, but symptoms may occur at any age. Approximately 83% of patients with monostotic fibrous dysplasia in the head and neck region will have symptoms within the first two decades of life.

Cosmetic swelling of the facial bones is the usual initial presentation of craniofacial fibrous dysplasia. When the bones of the orbits, paranasal sinuses, or skull base are involved, symptoms may include proptosis, visual loss, diplopia, headaches, nasal obstruction, mucocele formation, epistaxis, sinusitis, epiphora, anosmia, facial paralysis, and hearing loss. Temporal bone involvement with external auditory canal obstruction or erosion of the middle ear ossicles and otic capsule have been reported. Loss of vision is the most common neurologic deficit in craniofacial fibrous dysplasia.

Radiologic appearance of fibrous dysplasia may vary from largely lucent, should the lesion be primarily fibrous, to a "ground glass" appearance, should there be considerable ossification. The surrounding bony cortex is usually intact, and there is no sclerotic bone formation surrounding the lesion. The CT scan remains very helpful in evaluating local extension of craniofacial fibrous dysplasia. In 60% to 80% of cases, dysplastic expansion of bone will become quiescent after patients reach puberty, but complete spontaneous involution of lesions is unreported. In fact, some investigators feel that growth does not stop at the end of adolescence.

**Management**

The mere presence of a lesion of fibrous dysplasia does not justify surgical intervention. Surgery is now recommended, however, as soon as the deformity becomes cosmetically substantial, or when important function is threatened. There is no evidence to suggest that surgery accelerates the growth rate of residual normal or dysplastic tissue. Other investigators promote early surgical excision of facial lesions as an interceptive technique to prevent development of subsequent problems.

There are several surgical approaches for fibrous dysplasia. Simple contouring of expanded bone back to normal dimensions in the facial and skull regions has been fairly effective and is still indicated in certain circumstances, particularly in the teeth-bearing regions of the mandible and the maxilla, and in the hair-covered regions of the cranium. Approximately one-fourth of patients treated with conservative contouring will have a recurrence of bony enlargement that will require surgical revision.

More recently, complete resection of bony lesions with immediate reconstruction is being proposed as more definitive management. The technique of removal, remodeling, and replacement of the dysplastic bone as a free graft has been reported. Large methyl methacrylate implants for cranio-orbital reconstruction have been used also. The relief of optic nerve compression, orbital hypertelorism, dystopia, exophthalmos, and grotesque orbitofacial deformities requires radical excision and reconstruction, usually with autogenous free grafts, often in combination with neurosurgical and ophthalmological colleagues. Invasion of fibrous
dysplasia into the grafted bone has not been seen.

A major concern in surgical planning for fibrous dysplasia is the extreme vascularity of these lesions and the potential for significant intraoperative and postoperative blood loss. Radiotherapy is not effective and may carry an increased risk of malignant degeneration. The incidence of malignant degeneration of fibrous dysplasia is approximately 0.4%, and can be seen in patients without previous radiotherapy.

**Ossifying Fibroma**

Ossifying fibroma (cementifying fibroma) is a histologically benign, but potentially clinically aggressive, fibro-osseous lesion. It may present as a small asymptomatic growth or as an extensive tumor with gross facial deformity. Ossifying fibromas are well defined and smoothly contoured, which contrasts with the diffuse borders seen with fibrous dysplasia.

Histologically one sees evenly spaced spicules of mature bone rimmed with osteoblasts and osteoclasts within a fibrous stroma that has loose and dense areas, with whorling.

A variation of ossifying fibroma, referred to as active juvenile ossifying fibroma, or psammomatous ossifying fibroma, is characterized by small, round, psammomatoid ossicles embedded in a cellular fibrous stroma. This represents a more aggressive variant of ossifying fibroma and occurs primarily in children and young adults. These lesions may have a rapid growth rate with local aggressive behavior, and may mimic sarcoma.

The tissue origin may be primitive mesenchymal cells, or the periodontal ligament, which is known to be capable of producing both cementum and osteoid.

Radiographically, ossifying fibroma is characterized by expansile, sharply defined margins, often with a radiolucent peripheral component. Divergence of tooth roots is seen commonly in the jaws. Ossifying fibroma is characteristically monostotic. Ossifying fibroma is usually seen between the second and fourth decades of life and women are more often affected than men.

Although it can be found anywhere, ossifying fibroma is almost exclusively found in the craniofacial area. The mandible is clearly the most common site, involved in approximately 75% of cases in most series. The ethmoid, frontal, sphenoid, orbit, occiput, and temporal bones are less common sites.

**Management**

Primary treatment of mandibular lesions includes simple curettage or expectant observation after biopsy. Ossifying fibroma of the midface, according to recent reports, may require a more aggressive, en bloc, wide, local resection initially in order to prevent recurrence. These sites, outside of the mandible, include the paranasal sinuses, maxilla, and orbit.
**Cherubism**

Cherubism is a benign, expansile fibro-osseous lesion occurring within the jaws, that manifests as a progressive, painless, and often symmetric enlargement of the mandible and maxilla, with a predilection for the mandible. Facial swelling with characteristic fullness of the cheeks is the most typical clinical sign.

Cherubism demonstrates an autosomal dominant inheritance pattern with 100% penetrance in boys and 50% to 70% penetrance in girls, with variable expressivity.

Children with cherubism typically appear normal at birth and begin to demonstrate bilateral swelling between the ages of 2 and 4 years. Cherubism has been reported as early as 14 months of age. Boys are affected twice as often as girls. Maximum jaw enlargement occurs within 2 years of onset in most cases.

The mandibular angle is the most common location for cherubism, but the maxilla may be involved in up to 67% of the cases. Premature loss of deciduous teeth and delayed eruption of the secondary dentition are hallmark signs. Jaw size usually increases rapidly during the first 2 years after onset. By age 7, the lesions become static or progress slowly until puberty. Involution of the lesion begins at puberty and continues into the late teens; the maxilla usually regresses earlier than the mandible. Not commonly, the appearance of one side will improve, while the other remains static. Cervical adenopathy is frequently associated with cherubism and usually regresses at the same time as the jaw lesions.

The radiographic findings of cherubism include irregular multilocular lucencies with sharply defined margins and thinned overlying cortex. Areas of cortex are often perforated. Teeth may be displaced, unerupted, or appear to be floating in cystlike areas.

**Treatment**

Treatment of cherubism is not standardized. The unique biologic behavior of the tumor should be considered when recommending management.

The expectation of spontaneous involution has led most investigators to recommend surgery only after puberty, unless functional or psychological problems prompt earlier treatment. Recurrence of tumor growth is common and repeat surgery is needed frequently in younger patients. Surgical contouring of these expanded fibro-osseous lesions in late stages of the disease has led to good results. Radiotherapy is contraindicated.

**Giant Cell Lesions**

Peripheral giant cell reparative granuloma presents as a sessile or pedunculated mucosally covered mass that is reddish blue in color and contains multinucleated giant cells. It usually arises from the gingiva or alveolar mucosa, most commonly in the anterior mandible. The lesions bleed easily. Frequency of occurrence is equal in girls and boys. Treatment is excision or curettage.
Giant cell reparative granuloma is an endosteal lesion seen predominantly in patients between 10 and 20 years of age. It is found usually in the mandible, anterior to the first molar. The lesion may be asymptomatic, or may present as a localized jaw deformity. Radiographically, the lesions appear as lytic expansile unilocular cavities with well-demarcated, nonsclerotic margins. However, at times, a multicystic soap-bubble appearance may be noted. The overlying bone cortex is usually intact. The lesions may cross the midline.

Simple excision of well-defined lesions and excision and curettage of more diffuse lesions is standard surgical management.

**Mycobacterial Cervical Adenitis**

Tuberculous cervical adenitis is caused by *Mycobacterium tuberculosis* and usually affects older children and adults. The history is usually positive for contact with a known carrier of tuberculosis. Multiple, bilateral, matted nodes are found frequently in the lower neck and upper clavicular area. The tuberculin skin test is usually positive. Mycobacterial tuberculosis is usually acquired by inhalation in this country, and chest radiographs often will show pulmonary parenchymal or hilar disease. Primary therapy for tuberculous cervical adenitis is antituberculosis antibiotic therapy. Surgical excision is indicated for adenopathy unresponsive to medical management.

Nontuberculous mycobacteria (NTM), also referred to as the "atypical mycobacteria", are found in the water, soil, and other environmental sources. The NTM group includes *M. avium*, *M. scrofulaceum*, *M. intracellulare*, and *M. kansasii*. Nontuberculous cervical adenopathy is classically unilateral, and localized to the submandibular high cervical, and preauricular nodes. Approximately 80% of culture-positive nontuberculous cervical adenopathy is due to *M. avium* complex and the remainder to *M. scrofulaceum*. Presently, only about 10% of culture-proven mycobacterial cervical lymphadenitis in the USA in children is due to *M. tuberculosis*; the remainder are due to *M. avium* and *M. scrofulaceum*. The individual lymph nodes are generally nontender, may enlarge rapidly, and may rupture with formation of sinus tracts. There is usually no history of exposure to tuberculosis, and the chest radiograph is normal.

Skin testing is of limited usefulness in diagnosing NTM infection. Recent work has been done to develop new skin test antigens specific for several of the nontuberculous mycobacterial species, but these remain unavailable commercially. Most children with NTM lymphadenitis given intermediate strength (5 TU) purified protein derivative (PPD) tuberculin will have a weakly reactive test (5 to 9 mm), but some children may have a negative response, and others may respond with 10 mm or more induration.

Surgical excision of all involved lymph nodes is the mainstay of therapy if NTM lymphadenitis is suspected. Incision and drainage is not performed to avoid a subsequent chronic draining fistula. It is important to submit excised tissue for mycobacterial culture and sensitivity studies. The success rate after excisional surgery without chemotherapy is approximately 95%. For children with recurrent disease, a second surgical procedure is usually performed. A multidrug antituberculous regimen should be considered only in the patient with disease recurrence after two or more surgical resections.
Cat-Scratch Disease

Cat-scratch disease is a common cause of cervical adenopathy in the pediatric population. The typical history would include exposure to a cat, often a juvenile cat, 3 to 5 days after which a small papule will develop at the site of exposure. This papule progresses to a vesicular and crusty state in 2 to 3 days, and within a week or two regional adenopathy develops in those nodes, which drain the dermal or conjunctival sites of inoculation. The adenopathy may be painless, but is often tender, and nearly always is limited to a single site. Axillary lymphadenopathy is most common. The second most common is adenopathy of the head and neck.

The disease is self-limiting and adenopathy usually subsides in 1 to 4 months. Cat-scratch disease may rarely, however, progress to a severe, systemic, or recurrent infection producing encephalitis, neuroretinitis, osteomyelitis, arthritis, hepatitis, splenitis, and other problems.

The etiologic agent of cat-scratch disease recently has been isolated and cultured. It is a small, pleomorphic gram-negative bacillus. It can be demonstrated by the Warthin-Starry-Silver impregnation stain in histologic specimens. The histologic appearance of cat-scratch disease also includes reticular cell hyperplasia, granuloma formation, and microabscesses. The diagnosis can be confirmed further by a positive Hanger-Sore skin test, which is reportedly positive in greater than 99% of patients with cat-scratch disease who have had adenopathy for longer than 1 week. The antigen for this skin test, which was originally prepared from aspirated pus from patients with suppurative cat-scratch disease, is not readily available to clinicians, and the diagnosis of cat-scratch disease must therefore be made on the basis of other criteria.

The treatment of cat-scratch disease is mostly supportive. Surgical removal of involved lymph nodes is not necessary for management. Aspiration of suppurative nodes with a 16- or 18-gauge needle after spraying the site with ethyl chloride has been recommended for relief of symptoms and for obtaining material for diagnosis. Incision and drainage may be necessary for more advanced abscesses.

Most antimicrobial agents have not proven effective in the treatment of cat-scratch disease. Some investigators feel trimethoprim - sulfamethoxazole has a therapeutic effect. Successful treatment of cat-scratch disease with Ciprofloxacin has been reported in a small series of patients; however, Ciprofloxacin is not approved for use in children.

Benign Tumors of the Peripheral Nervous System

Schwannomas and neurofibromas, two common benign tumors of the peripheral nervous system, are distinct clinical entities that may arise from multiple sites within the head and neck, including cranial nerves, motor and sensory cervical nerves, small distal nerves within the tongue and oral mucosa, and elsewhere, and autonomic nerves.

The schwannoma, also known as neurilemmoma, is a benign, solitary, often encapsulated, tumor that derives from the Schwann cell of a peripheral nerve sheath. Microscopically, the nerve of origin can be seen attached to and compressed by the
Schwannoma. Schwannomas are almost never associated with malignant change. Retrogressive changes, such as cystic alterations or hemorrhagic necrosis, are usually present.

Histologically, solitary schwannomas are composed of two tissue types: (a) cellular Antoni type A tissue with compact spindle cells in parallel orientation, often with rows of nuclei forming a palisading pattern; when these palisading areas are oriented around parallel bundles of nerve fibers, Verocay bodies are formed. (b) Antoni type B is characterized by abundant acellular collagen matrix and a random orientation of nerve fibers. Schwannomas are seen more often in the third and fourth decades of life, but also occur in children.

Management is by complete surgical excision. Since the tumor arises from the nerve sheath, it is often possible to remove a schwannoma from larger nerves and leave the majority of the nerve axons intact. The diagnosis is confirmed by histologic examination. Recurrence after complete excision is rare.

Neurofibromas may occur sporadically as well-circumscribed, solitary lesions or in association with one of the two genetic subtypes of systemic neurofibromatosis (NF-1 or NF-2).

Several types of solitary neurofibromas are found in children, including cutaneous and subcutaneous, plexiform, elephantiasis neuromatosa, and molluscum fibrosum. The cutaneous and subcutaneous lesions arise near the termination of small cutaneous nerves and appear clinically as soft, elevated, nodular pedunculated masses with increased pigmentation in the overlying skin. Neurofibromas also can be found within the upper aerodigestive tract of children, such as the larynx, and treatment is judicious total surgical excision, if possible. Often the lesion cannot be completely removed, and repeat subtotal removal is indicated.

Generalized neurofibromatosis has been classified recently into two genetically distinct disorders that are inherited in an autosomal dominant pattern with variable penetrance. The spontaneous mutation rate is high.

**Neurofibromatosis - 1**

Neurofibromatosis - 1 (NF-1) (previously referred to as von Recklinghausen's neurofibromatosis or peripheral neurofibromatosis) is considered to exist when two of the following diagnostic criteria are met:

1. six or more café au lait macules over 5 mm in greatest diameter in prepertual individuals, and over 15 mm in greatest diameter in postpubertal individuals;  
2. two or more neurofibromas of any type or one plexiform neurofibroma;  
3. freckling in the axillary or inguinal regions;  
4. optic glioma;  
5. two or more Lisch nodules (iris hamartomas);  
6. a distinctive osseous lesion, such as sphenoid dysplasia or thinning of long bone cortex, with or without pseudoarthrosis;  
7. first-degree relative (parent, offspring, or sibling) with NF-1 by the above criteria; NF-2 is diagnosed usually within the first decade of life.
Neurofibromatosis - 2

Neurofibromatosis - 2 (NF-2), previously known as central neurofibromatosis or bilateral acoustic neurofibromatosis, is diagnosed when an individual has (a) bilateral eighth nerve masses seen by MRI or CT scan or (b) a first-degree relative with NF-2 and either a unilateral eighth nerve mass or two of the following: neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity.

A full discussion of the developmental, endocrine, and neoplastic disorders associated with the neurofibromatoses is beyond the scope of this chapter and management of these patients in a multidisciplinary team facilitates comprehensive management.

The treatment for symptomatic neurofibromatosis is surgical with preservation of function when possible. The management of patients with NF-2 and bilateral vestibular nerve tumors is an interesting and dynamic topic in itself. Those patients with NF-2 may suffer cosmetic deformities from enlarging plexiform neurofibromas and also from osseous mesodermal dysplasia causing distortion of the facial skeleton, sphenoid, and temporal bones. These dysplastic changes can involve any portion of the skeleton.

Pilomatrixoma (Calcifying Epithelioma of Malherbe)

The pilomatrixoma is a benign, usually solitary cystic tumor of the skin that is believed to arise from a primitive hair matrix cell. Over half of these tumors present in the head and neck region, most frequently on the brow, lid, and cheek. There is also an affinity for the external ear. About 40% of patients present in the first decade of life, and an additional 20% present before age 20. The tumor has been found in neonates.

Clinically, pilomatrixomas present as a well-demarcated, mobile, solid or cystic nodule. The lumen typically contains cheesy amorphous debris, keratin, and possibly calcified material. The overlying skin may appear normal, or may have a reddish or bluish discoloration. The tumors are usually asymptomatic and slow growing. They may reach up to 3 cm in diameter. Complete surgical excision is the management recommended by most authors.

Teratomas and Dermoid Cysts

Teratomas are developmental neoplasms that contain elements from all three germ cell layers - endoderm, mesoderm, and ectoderm. Congenital teratomas occur with an incidence of 1:4,000 live births and affect the head and neck in only approximately 2% to 4% of cases.

Teratomas and dermoid cysts are classified into four groups according to germ layer of origin and the degree of tissue differentiation. Many authors consider any nasopharyngeal neoplasm that consists of multiple types of tissue extrinsic to their site of origin a nasopharyngeal teratoma.

Dermoid cysts (hairy polyps) are composed of epidermal and mesodermal tissue elements. They are covered with skin and contain epidermal appendages such as hair follicles, sweat glands, and sebaceous glands. Most of the mass consist of an adipose matrix with
fragments of striated muscle, cartilage, or bone. They are usually pedunculated and often occur along lines of embryonic fusion. Teratomas in the nasopharynx traditionally arise from either the lateral or the superior walls and can cause symptoms of nasal obstruction in the neonate.

*Teratoid cysts* are composed of ectoderm, mesoderm, and endoderm, but differ from teratomas in their poor histologic differentiation. The cyst lining may be stratified squamous epithelium or ciliated respiratory epithelium. Components of the tumor may contain fat, muscle, cartilage, bone, acinous glands, teeth, epithelial cysts, and nervous tissue.

True *teratomas* are composed of all three germ layers and cellular differentiation is advanced so that organs can be recognized in these masses.

*Epignathi* represent the highest degree of differentiation into a parasitic fetus that may develop organs and limbs. These tumors are usually attached to the sphenoid bone and are generally "incompatible" with life.

A classification system based on birth status, age, diagnosis of the mass, and the presence or absence of respiratory distress has been proposed (Table 5).

**Table 5. Classification system for cervical teratomas by age and clinical presentation**

<table>
<thead>
<tr>
<th>Group</th>
<th>Characteristic</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Stillborn and moribund live newborn</td>
</tr>
<tr>
<td>II</td>
<td>Newborn with respiratory distress</td>
</tr>
<tr>
<td>III</td>
<td>Newborn without respiratory distress</td>
</tr>
<tr>
<td>IV</td>
<td>Children ages 1 month to 18 years</td>
</tr>
<tr>
<td>V</td>
<td>Adult.</td>
</tr>
</tbody>
</table>

**Presentation**

Head and neck tridermal tumors are usually apparent at birth or in the immediate neonatal period and more than one-half of nasopharyngeal dermoids are seen within the first year of life.

Prenatal sonographic diagnosis of cervical teratomas is based on the presence of a fetal neck mass with solid and cystic components. Calcification within the tumor on plain radiographs is virtually diagnostic, but may be seen in only 16% of cases.

A history of maternal polyhydramnios has been noted in 18% of neonates born with cervical teratomas, presumably from direct esophageal compression by the tumor, which interferes with swallowing. Prematurity, stillbirth, and birth dystocia are reported also. The incidence of polyhydramnios is greatest in pregnancies yielding neonates of groups I and II, which are typically the larger tumors. Tracheal deviation and posterior displacement, detected by both plain films and ultrasound, is also a common finding.
Clinically, cervical teratomas appear as firm, frequently mobile, multilobular cystic lesions. Rapid enlargement of the cystic components can lead to airway compromise. The tumors can be found anteriorly or laterally in the neck. They may cross the midline and extend into the submandibular and floor of mouth region, as well as into the mediastinum.

Nasopharyngeal "teratomas" are most commonly dermoid cysts. These tumors arise from the palate or nasopharynx and may extend intracranially. Nasal obstruction may be a presenting symptom.

The incidence of malignant degeneration in sacrococcygeal and other noncervical teratomas in children is approximately 20%. In contrast, cervical teratomas found in adults, although more rare, are often malignant.

Management

Management of nasopharyngeal and cervical teratomas begins as early in the prenatal period as possible, often after finding the abnormal mass on routine obstetrical ultrasound imaging. Optimal management of the airway at the time of delivery may require close coordination between the anesthesiologist, nursing staff, neonatologists, surgeon, and obstetrician. Therefore, the management strategy and necessary instrumentation should be organized well in advance. Since premature delivery is known to be associated with teratoma and an unexpected delivery might compromise the team management of the airway, a decision regarding the desirability and timing of an elective cesarean section delivery must be made.

The immediate management of patients with nasopharyngeal and cervical teratomas requires establishment of an airway. In small tumors, positioning may be adequate temporarily. Endotracheal intubation to secure the airway is the least traumatic, preferred method in more severe cases. If intubation is expected to be difficult based on previous imaging studies, intubation with a pediatric rigid bronchoscope or endotracheal tube intubation utilizing a flexible nasopharyngoscope can be employed. Tracheotomy may be difficult if the lesion overlies or depresses the trachea. Further preparations and management options have been described, including maintaining the neonate on fetal circulation by paralyzing the baby with succinylcholine to prevent spontaneous ventilation while an airway is secured. Once an airway has been secured, prompt surgical excision should be performed to prevent airway-related morbidity and mortality.

Prior to surgical excision, the tumor must be imaged. The high fat content of teratomas cause a strong signal on T1-weighted MRI images, allowing differentiation from cystic hygroma, which may appear similar on CT studies. Particularly in the evaluation of nasopharyngeal tumors, a coronal CT scan provides valuable information regarding the bony integrity of the skull base, and will determine preoperatively if there is intracranial or intraorbital involvement while excluding congenital midline CNS lesions, such as encephaloceles. If the latter is present, a combined approach with neurosurgery or ophthalmology will be needed. Teratomas, classically, do not communicate intracranially.

Complete surgical excision results in a good prognosis. Long-term follow-up is indicated.
Dermoids

Dermoids of the head and neck are a class of benign developmental neoplasms related to teratomas. They are most commonly located in and around the nose, orbit, and submental region.

Orbital and nasal dermoids are complex entities in themselves and are discussed elsewhere in this text.

Submental dermoids are smooth, rounded, mobile nontender masses located in the midline above or below the mylohyoid muscle. If the majority of a submental dermoid is located above the mylohyoid, marked elevation of the floor of mouth and tongue may occur, compromising the airway. If a submental dermoid enlarges to compromise oral intake or the child’s airway, then it should be excised immediately.

Surgical removal of small, submental dermoids located primarily below the mylohyoid muscle can be delayed until 3 to 6 months of age without significant risk to the airway integrity.

Dermoid cysts of the tongue have been reported and are best treated by midline glossectomy and careful surgical excision of the cyst walls and tracts.

Sternocleidomastoid Tumor of Infancy

The sternocleidomastoid tumor of infancy (STOI) is a unique perinatal fibromatosis that manifests as a firm spindle-shaped mass within the sternocleidomastoid muscle of the neonate at 1 to 4 weeks of age. This lesion is also known as "pseudotumor of infancy" and has been equated to "fibromatosis colli".

The incidence of STOI is 0.4% of newborns (ie, approximately 1 in 4,000). The mass is usually not detectable at birth but becomes apparent at between 1 and 8 weeks of age with the majority occurring between 2 and 4 weeks of age. There is usually an equal sex ratio.

The tumor presents as a firm, painless, fusiform mass within the middle or lower portion of the sternocleidomastoid (SCM) muscle. Typical size varies from 1 to 3 cm. The right and left sides appear to be equally affected by STOI. Bilateral involvement is rare, but has been reported. Either the sternal or clavicular head of the SCM, or both, can be primarily involved by the lesion. The tumor typically increases in size for a few weeks, stabilizes for 2 to 3 months, and then slowly resolves by 4 to 8 months.

Sternocleidomastoid tumor of infancy is a transient disorder that can be associated with both temporary and permanent torticollis. The pathogenesis of this disorder remains unclear. About 10% to 20% of patients with STOI will go on to develop either a progressive or delayed torticollis, and 25% will have a minor residual degree of persistent asymmetry and tightness of the SCM muscle. Congenital muscular torticollis and STOI may represent different stages of an underlying SCM muscle fibrosis. Congenital muscular torticollis usually appears at 3 to 4 years of age, and may or may not be associated with a preceding history of STOI.
Torticollis secondary to STOI can lead to permanent deformities, such as plagiocephaly (a cranial deformity characterized by a rhomboid shape) and facial hemihypoplasia (characterized by an underdevelopment of half the face). Downward displacement of the ipsilateral eye and corner of the mouth are seen also. Surgical release is indicated when medical management is not adequate.

Microscopically, STOI is composed of dense fibrous tissue and an absence of normal striated muscle. Biopsy, however, is not needed routinely in the management of these tumors. Most authors agree that the diagnosis of STOI can be based safely on the clinical history and presentation. CT and ultrasound can help localize the tumor to the SCM muscle itself. Biopsy should be reserved for cases in which the diagnosis is uncertain. The differential diagnosis includes rhabdomyosarcoma, fibrosarcoma, dermoid, and neuroblastoma.

**Management**

Conservative, nonoperative treatment is the recommended initial management strategy. Neck flexion and chin rotation, head positioning, and heat and massage application are all part of the initial physiotherapy program. If craniofacial asymmetry develops or if the tumor or torticollis persist beyond 1 year to 18 months, surgery is indicated. Release of the SCM muscle distally, including both the sternal and clavicular heads, and lysis of associated fibrous tissue and platysmal banding is the recommended procedure initially. Release of the superior pole of the SCM may be necessary subsequently. Total excision of the SCM appears to offer no great advantage and it places the spinal accessory nerve at significant risk. Physiotherapy must be reinstituted postoperatively to ensure optimal results.
Historically, there have been progressive improvements in the management of injuries to children that extend from the initial emergency management and triage through improved pediatric care. Improved overall patient assessment, as well as specific wound assessment (Table 1), have led to emphasis on proper early management. Over the last several years not only have techniques been improved but the materials with which to work have shown progressive development with new suture materials and refinements in the swaged on needles available. Instrumentation is not only known to the specialist but is becoming generally available in the emergency room where not too long ago this was not always the case. The specialist has contributed greatly to the education of those in emergency medical services as the development of emergency room medicine has come about. A designated pediatrics emergency room has been a major contribution to the care of the injured child. There has been increased knowledge of wound healing, improved prognostication of outcomes, and more sophisticated management of complications.

Table 1. Wound analysis

<table>
<thead>
<tr>
<th>Type of wound</th>
<th>Contamination</th>
</tr>
</thead>
<tbody>
<tr>
<td>Location of wound</td>
<td>Foreign bodies</td>
</tr>
<tr>
<td>Landmarks</td>
<td>Blood supply</td>
</tr>
<tr>
<td>Facial nerve</td>
<td>Nonviable tissue</td>
</tr>
<tr>
<td>Parotid duct</td>
<td>Skin margins</td>
</tr>
<tr>
<td>Direction of laceration</td>
<td>Local tissue shock</td>
</tr>
<tr>
<td>Angle of laceration</td>
<td>Crushing injuries.</td>
</tr>
<tr>
<td>Condition of tissue</td>
<td></td>
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</tbody>
</table>

Soft tissue trauma in the child extends from bruises and abrasions through lacerations, avulsions with major tissue loss, and the associated maxillofacial injuries. Bites and electrical burns require special attention. The etiology of the trauma has become more and more complex as mechanization has become widespread, extending from modes of transportation to various recreational vehicles. If not actually participating, the child is often taken along and encouraged to participate before he is able to handle the power and velocity. Without safety devices, the small child in particular can become a projectile, leading to serious injury. Space heaters are not confined to lower income homes and remain a source of burns, particularly when clothing is ignited. There are innumerable electrical appliances, which often fascinate the young child and become sources of electrical burns, particularly of the oral commissure. Although increased awareness exists, there seems to be an ever-lasting number of animal bites. It seems appropriate to mention, as we emphasize safety devices in automobiles, the cooperation of the American Academy of Otolaryngology - Head and Neck Surgery and the American Society of Plastic and Reconstructive Surgery in bringing these issues to the attention of the public in 1991.
General Considerations

The reconstructive surgeon must have a fair knowledge of wound healing and the ability to anticipate various stages of healing, both for his own management and for reassurance to the child or parent. Wound healing is covered in a most scholarly manner by Koopmann in Chapter 20 of this book.

Although much research and advanced study have been devoted to wound healing, there is little in the literature comparing specifically the healing of infants and children to adults. Most observations, as with my own, are from practical clinical conclusions as one follows the child through the healing period.

In general, children heal rapidly but with the thinner skin, increased vascularity, greater elasticity, abundance of fat, and growth adding to tension, scars tend at least to spread more than in adults. In the observations of the senior author, there is no greater tendency to true hypertrophy. Various authors state frankly that hypertrophic scars are more common in children, but I am impressed that these and keloids result in relatively the same proportion as they do in the adult population, all etiologic factors being taken into consideration. There is a very real tendency for these scars to spread and appear, in later phases of healing, to have hypertrophied, but these will soften and will then present later simply as a scar that has spread.

Stitch marks are more common in children, influencing the recommendations for suture techniques.

Emergency Consultations

Starting at the beginning, the physician doing facial plastic surgery, must be willing to come as soon as possible to see the injured child, making early assessments and assuming a leadership role if this is not already assumed by someone else. Early arrival is a major consideration and plays a large part in the reassurance of the child and the parent and allows an orderly decision-making process. Perhaps before any medical treatment, establishing some control and cooperation from all emergency room personnel and with the parent should be emphasized. Personnel should be trained to present a caring, reassuring, and calm atmosphere. As pediatric emergency rooms have developed, these personnel are usually quite skilled in this important early phase of management. It is not within the scope of this chapter, but early assessment, extending from the airway and hemorrhage through multiple injuries and general conditions, must be made. Determination must then be made as to whether the child can be managed in the emergency room or office or will require general anesthesia and repair in the operating theater (Table 2). In brief, the child is frightened and the parents are anxious. Parental guilt and anger become major factors in how smooth the state of management goes.

A basic set of instruments, which the senior author recommends and which can be carried in a coat pocket folder, includes two small skin hooks (Storz N4700), Brown-Adson multiple teeth forceps (Storz N5420), Webster needle holder (Storz 5712) or the small needle holder from Anthony Products P0404 (Storz), and small Bishop-Harmon forceps (Storz PE1500). Even sterile packets of some suture materials can be brought to the emergency room by the surgeon along with a camera. Photographing the wound before and after surgical
cleanup may be vital for medical documentation, assisting the parents in insurance claims, and serving as a safeguard against any type of litigation. Even a simple automatic camera with flash captures the severity of the injury. For revisions, time for analysis and more sophisticated photographs become easier. It takes a particular conscientiousness to realize the importance of emergency photographs. The surgeon should carry his own camera, having it available for all emergency situations.

Table 2. Wound management

<table>
<thead>
<tr>
<th>Cleansing</th>
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<tbody>
<tr>
<td>Irrigation</td>
<td>Scrub brush</td>
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<tr>
<td>Removal of foreign bodies</td>
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<tr>
<td>Hemostasis</td>
<td></td>
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<tr>
<td>Debridement</td>
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<tr>
<td>Minimal</td>
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<tr>
<td>Preparation of skin edge</td>
<td></td>
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<tr>
<td>Beveled margin</td>
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<tr>
<td>Uniform thickness</td>
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<tr>
<td>Wide undermining</td>
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<tr>
<td>Beveled laceration</td>
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<tr>
<td>Trapdoor laceration</td>
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<tr>
<td>Reduction of skin tension</td>
<td></td>
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<tr>
<td>Readjustment of tissue</td>
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<tr>
<td>Management of deep scar</td>
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<tr>
<td>De-epithelization</td>
<td></td>
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<tr>
<td>Release of contracture</td>
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<tr>
<td>Exercise</td>
<td></td>
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<tr>
<td>Suture technique</td>
<td></td>
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<tr>
<td>Layered closure</td>
<td></td>
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<tr>
<td>Interrupted subcuticular closure</td>
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<tr>
<td>Skin suture</td>
<td></td>
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<tr>
<td>Interrupted, spring loop</td>
<td></td>
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<tr>
<td>Continuous subcuticular (intradermal)</td>
<td></td>
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<tr>
<td>Surgical dressing</td>
<td></td>
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<tr>
<td>Fine mesh gauze</td>
<td></td>
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<tr>
<td>Antibiotic ointment</td>
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<tr>
<td>Wound support</td>
<td></td>
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<tr>
<td>Pressure</td>
<td></td>
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<tr>
<td>± Tissue drain</td>
<td></td>
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<tr>
<td>Delayed reconstruction</td>
<td></td>
</tr>
<tr>
<td>Skin grafts</td>
<td></td>
</tr>
<tr>
<td>Skin flaps</td>
<td></td>
</tr>
<tr>
<td>Release of contracture</td>
<td></td>
</tr>
<tr>
<td>Dermabrasion</td>
<td></td>
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<tr>
<td>Immediate</td>
<td></td>
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<tr>
<td>Delayed</td>
<td></td>
</tr>
<tr>
<td>Antibiotics</td>
<td></td>
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<tr>
<td>Intraleisional steroids.</td>
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</tbody>
</table>
The degree of cooperation one can get from a child is astounding. Most of the time it is surprising how well the child will cooperate, if managed and handled appropriately. Sedation and bundling with a papoose become the next consideration. With an adjusted parent, the parent holding the child's hand and talking with him can be helpful. Many children, after the excitement of the injury, the frantic parent, the frantic drive to the hospital, and considerable crying, will simply go to sleep in exhaustion once they realize they are not being hurt. After initial irrigation at least, if not surgical preparation, it seems helpful to irrigate the wound thoroughly with 2% Xylocaine and adrenaline 1:100.000 and then to put a gauze compress saturated with this anesthetic in the wound. This technique has been used for years. More recently a topical mixture of tetracaine, adrenaline, and cocaine (TAC) has proven effective. Injection should be made through the laceration. This is often less painful and it avoids injecting through the contaminated skin. It is best to accomplish this before it is necessary to clamp any significant bleeders.

Quite often the patient has been seen "elsewhere" and the laceration may have been repaired, sometimes quite well but often omitting a layered closure. We strongly prefer that the laceration not be repaired so that appropriate layered closure can be carried out, but more particularly so that the deep wound can be thoroughly explored, both for foreign bodies and fractures. Often fractures can be approached through the laceration and it is better to have the usual, rougher management of the bone reduction before the delicate skin closure is carried out. The two greatest problems for this delicate closure are infection and hematoma. For this reason, in the fresh laceration, interrupted sutures are used so that, if necessary, opening the wound for drainage may not destroy the entire closure. Many of these wounds will require a simple tissue drain. After massive irrigation with saline, particularly if the nasal or oral cavity has been injured, we prefer irrigating with a surgical bacitracin solution. The wound must be thoroughly explored for foreign bodies with particular consideration being given to glass, paint, wood chips, and asphalt. After anesthesia, the use of the scrub brush may be helpful, both for skin margins and abrasions (Table 2).

Techniques

After the above general evaluation and preparation, specific techniques can be carried out. Wound analysis is most vital, including the condition of the tissue surrounding the laceration, the location and angle of the cut, and the involvement of vital structures, including anatomic margins (Table 1). The angle and direction of the laceration in regard to relaxed skin tension lines and major landmarks or folds may enter into the initial analysis and repair. Lacerations that extend across margins, such as the lip, nostrils, eyelids, or external ears, require particular attention to proper alignment. Vital structures may include major vessels, the facial nerve, or the parotid duct. If there is any question regarding eye injuries, it is important to get early ophthalmologic consultation, preferably in the emergency room, if this is possible.

Debridement should be minimal, or not at all, if there is a question about the structures involved or the viability of the tissues.

Wound closure is preceded by appropriate preparation of the skin margins. The skin margins are beveled or slightly undercut to assist in eversion in an attempt to counter scar separation and depression. For the beveled cut, the exposed dermis opposite the angle of the
cut is beveled in the wrong direction and must be cut away. On the side of the angle, the exaggerated beveling often requires a slightly more vertical cut matching the opposite side. It is vital that undermining be carried out on both sides of the wound with the thickness of the elevated tissue being the same on both sides. This allows appropriate skin closure not influenced by the deep tissues, which are closed separately. This preparation of the skin edges serves also for minimal debridement for foreign particles that may be ground into the wound. For the trapdoor laceration, one of the most frequently omitted portions of the repair is undermining the disturbed tissue opposite the trapdoor. This equalizes the tension and assists in closure. The margins of the wound and the deep tissue can be handled independently and generally the deep sutures are brought forward and down. Beneath the trapdoor flap, some sutures passed parallel to the flap are utilized by tacking these to the deep wound, preventing the common "humping" on the side of the trapdoor. As we have mentioned, wounds in children tend to spread and, if there is any tension at all or the sutures are left in too long, there is a tendency for stitch marks. For this reason the deep tissues must be meticulously closed and all tension possible reduced - "what counts is what is underneath". The senior author very strongly disagrees with the admonition that deep or subcuticular sutures create strangulation. If these are properly and delicately placed, and not overdone, they do not create anywhere near the degree of strangulation that skin sutures produce when closed under tension. Also, without the deep sutures, it is necessary to leave the skin sutures in longer. Again, one tries to prevent the tendency toward stitch marks in children; therefore, it is preferable to get the skin sutures in children, which are tied without tension, out within 3 to 5 days. With the active child, or if tension-reducing dressings are not practical, some of the sutures may be left in as long as a week. This is particularly true for the common cut beneath the chin, which is so often struck again and may break open. Certainly there is a tendency to spread, for this possibly most common laceration in children. In this area, this is compounded by the associated contusion when the child falls. In the deep tissues, we use both vertical and interrupted sutures and sometimes horizontal or parallel mattress sutures as the first "tension-reducing" sutures. These are passed in the deep tissue or, if necessary, lateral to the wound margin in the deep dermis lateral to where the interrupted subcuticular suture will be placed. Again, because of the tendency to spread, we have on occasion used permanent suture material, such as mersilene, but we are not comfortable with this, having seen permanent materials exude eventually or create irregularities that are permanent. For this deep suture we are now using either 4-0 vicryl (polyglactin 910) by Ethicon or 4-0 PDS (polydioxanone) also by Ethicon. These can be obtained with either a half-curved cutting needle or a round noncutting needle. In the majority of instances we prefer the cutting needle and we use 4-0 Vicryl on a PS-5 needle or PDS, which is even slower absorbing.

The interrupted subcuticular suture we have advocated is appropriately placed with the knot buried and to deliberately evert the incision or laceration lines from the surface. This means that the needle has to be placed slightly closer to the skin surface lateral to the wound than at the skin margin. This suture should bring the skin edges together but there may be some unevenness of the skin surface or level of the two skin edges, thus requiring the fine nylon skin sutures without tension. In the experience of the senior author, if only the interrupted subcuticular suture is placed, it allows more blood and serum to microscopically separate the two skin edges, and surgical tapes do not correct any differences in the skin level or assist in maintaining eversion. Therefore, we use interrupted spring loop knots, the spring loop suture, and may alternate these with simple surgical twists. We are currently using either Dermalon 5-0 or 6-0 with a CE-2 needle or Ethilon 5-0 or 6-0 on a PC-1 needle.
All wound edges are delicately handled, preferably with the skin hook. To manage small flaps and points of tissue, the Bishop-Harmon forceps may be utilized without crushing, but quite frankly do not serve as well in the practiced hand as the skin hook does for evertung the skin margin. The Brown-Adson multiple toothed forceps are used for larger tissue handling. We are depending heavily on the deep closure to prevent separation and depending on the skin sutures only for refinements.

For angled lacerations with pointed flaps, we have continued to use the "corner" stitch for angles less than 80° to 90°. This suture is passed through the skin, comes out on the margin subcuticularly, across the point of the flap subcuticularly, and then subcuticularly passed to emerge through the skin close to the original insertion. Care should be taken that the two sutures through the skin surface are not too far apart, thus avoiding a wide stitch mark. For near right angled lacerations or obtuse angles, the suture is carried straight across, bisecting the angle.

**Bites**

Bites continue to be a significant source of facial trauma with over one million bites occurring annually from dogs alone; 70% of those seen in the pediatric population are on the face and neck. In addition to salivary contamination the open wound can collect further debris en route to the emergency room. The old saw that "dog bites are cleaner than human bites" cannot influence management.

Medical management must include tetanus, prophylaxis, rabies precautions, and wound infection prophylaxis or treatment, depending on the age of the wound at presentation. Wild animals should be considered rabid and if possible the brain of the biting animal should be examined for the presence of antibodies to rabies. If possible the patient should be treated with rabbit immune globulin and human diploid cell rabies vaccine (HDCV). If no HDCV is available, then duck embryo vaccine may be substituted. If the bite is from a nonimmunized domestic animal then the animal may be observed for 10 days. If no illness or unusual behavior is observed, no treatment is required.

Dog bites, which comprise the vast majority of bites, have infection rates as high as 29% and are generally secondary to *Pasteurella multocida*. This and other pathogenic canine flora usually respond to penicillin, the first-line drug for prophylaxis and treatment. Tetracycline may be a reasonable alternative in adults but should not be given to children. Instead, IV cefoxitin is recommended. Human bites have a higher rate of infection than do animal bites, usually secondary to anaerobic streptococci or *Eikenella corroden*. Susceptibility and drug therapy are identical to that in animal bites. The wound itself must be thoroughly irrigated with massive saline solution and either a surgical prep solution or antibiotic solution. There is frequently further tissue damage from the crushing action of the injury and shearing forces create wound irregularities and fragmentation of the margins. Debridement should be minimal but devitalized, and shredded tissue should be removed, creating a better wound margin. Irregular margins can be utilized to effect a broken-line closure and reduce the need for unnecessary sacrifice of tissue. Simple lacerations may be closed directly and avulsion injuries may be repaired with local flaps or require a free skin graft. If there are great concerns, either all or part of the wound may be left open for drainage. Wounds clearly infected, or those greater than 12 hr old, should have delayed repair.
awaiting a more favorable wound without infection. Further debridement may be necessary for this second closure.

**Thermal Burns**

The management of thermal burns is a subject in itself. In brief, there are three phases of burn care: (a) cleaning and debridement, (b) skin grafting, and (c) reconstruction. After supportive care is initiated, the wound is allowed to demarcate. Debridement is then performed, usually requiring a general anesthetic for children. Following debridement the wound is dressed with silver sulfadiazine until a healthy bed of noninfected granulation tissue appears. The appearance of this granulation tissue heralds the second phase, that of skin grafting. This usually occurs 10 to 20 days after the injury. Medium thickness (0.014 to 0.018 mm) grafts are recommended to improve color match and minimize contraction. Once graft take is established, conforming splints or molds are placed to further minimize wound contracture and hypertrophic scars.

Generally 6 to 12 months after the injury the child is ready for reconstruction. The primary goals here are to restore function and cosmesis. The order of priorities when multiple procedures are involved is as follows: eyelid, perioral, neck, chin/cheek, then the remainder of the face. The ear requires some special consideration. Sulfamylon is used on the ear for its penetrating effect and in the effort to salvage all cartilage possible. Sulfamylon or silver sulfadiazine may be painful around the eyes and here Polysporin ointment may be used. Polysporin may also be used on limited, superficial, partial-thickness burns. For the eyelids, early tarsorrhaphy should be considered, both to protect the globe and to prevent contracture with ectropia. On the nose, alar rim contracture is common and prevention may require early full-thickness skin grafts. Consideration should be given to plastic stents in the nostrils and ear canals. Extra care should be given to prevent erosion of the ala and columella by intranasal tubes. Septal perforations and the loss of the columella have occurred.

Contouring pressure dressings are strongly recommended. For the ear these dressings may need to be worn 1 to 2 years. Despite the inconvenience and question of compliance, these dressings are particularly important in children. Pressure garments and clear, molded plastic masks are sometimes used.

Tissue expansion has recently been shown to be a useful adjunct in head and neck burn injury reconstruction.

**Electrical Burns**

Electrical burns in infants and children are common and usually the result of placing an electrical wire in the mouth. Perhaps the most common site of injury is the lateral commissure. Initial treatment is supportive care, tetanus prophylaxis, and the administration of fluids and antibiotics as indicated. Arguments for primary debridement and reconstruction continue to be presented. In our observation, this has either led to greater debridement than later proved to be necessary or, after primary reconstruction, additional tissue loss, either at the surface or in the depths of the wound. We advocate the conservative approach of delayed eschar removal after demarcation has occurred to prevent the unnecessary removal of viable tissue. Oral splinting should be initiated early to prevent contracture; this alone may yield
surprisingly good results. After healing by secondary intention, reconstruction can be accurately planned, measurements can be made comparing the normal and abnormal sides, and then planned skin and mucous membrane flaps, grafts, or advancements can be carried out. The key to these procedures is overcorrection to compensate for postoperative wound contracture. Measurements from the midline to the commissure of the normal and abnormal sides are important in planning.

Postoperative Care

Support to the wound is vital in the child, again hoping to capitalize on the increased proliferation of fibroblasts in children, and prevention of wound spreading. Some form of sterile, elastic tension-reducing strips are recommended over a strip of Telfa after placing Polysporin ointment on the wound. Of late we have been using Suture Strips Plus (Genetic Laboratories), which have a desirable elastic quality. On top of this, for the primary dressing, pressure is applied extending from a "clinker" built, elliptical dressing to wraparound head and neck dressings, depending on the site of the injury. This is tival to prevent postoperative hematoma and to reduce the initial edema. These dressings must be changed frequently or, after the wound is inspected, it is possible to leave on the surgi-strips only for another 48 hr. Dressings are changed and the sutures removed on the 3rd and 5th days. Support remains on the wound with antibiotic ointment and telfa for 1 week and after this, surgi-strips alone may be used for another 3 weeks. The parents must be instructed to remove the surgi-strips from each end of the tape toward the laceration to prevent separating or widening the wound.

The good academic reasoning for not using antibiotics in the sterile wound are sound. However, from the practical standpoint, most lacerations are contaminated or potentially contaminated no matter what the degree of care and delicate closure of the wound, so all can be lost if infection occurs. Several generations of antibiotics now exist and can be specific for particular bacteria. Initial treatment is usually empirical unless suppuration exists and cultures can be taken. Generally a broad-spectrum antibiotic, either Keflex or ampicillin, is used. If an intravenous line is established a bolus of antibiotics is given IV intraoperative and oral antibiotics are administered for at least an additional 72 hr. Dosage is dependent upon the age and weight of the patient. Tetanus injection depends on the inoculation history of the child.

Despite a large number of skin flora present, only Staphylococcus aureus and Streptococcus pyogenes have significant potential for infection. Appropriate antibiotics, especially after culture, may include nafcillin, methicillin, cefazolin, cloxacillin, cephalixin, or trimethoprim/sulfamethoxazole. As Pseudomonas is frequently present in the external auditory canal one should consider coverage of this organism when the canal is traumatized and infection is present or likely. Appropriate antibiotics include aminoglycosides or ceftazidime, which are given intravenously. Although ciprofloxacin would be an appropriate oral antibiotic, its safety and effectiveness in children and adolescents under 18 has not yet been established.

In regard to tetanus, the Clostridium tetani, the anaerobic gram positive, spore-forming bacilli is antibiotic resistant and extremely resistant to antiseptics. They generally live in the necrotic tissue, making adequate wound debridement imperative. Proper immunization at this point depends on the inoculation history of the patient.
Most surgeons use some form of antibiotic ointment or cream on their surgical dressings. For the dressing we prefer a fine mesh gauze impregnated with the antibiotic, or telfa placed over the wound, which is lightly covered with the antibiotic.

There is a greater argument for not using antibiotics in the clean, secondary revision; however, we would leave this decision up to the reasoning and personal security or insecurity of the surgeon in that all precise revisions can be marred with either infection or hematoma.

**Complications**

Minor complications are the tendency in the child for spreading of the scar and visible suture marks. Major complications, such as poor healing due to inanition as a result of multiple injuries, severe infection with potential bacteriemia, and allergic reactions leading to anaphylactic shock, require close attention in consultation with the appropriate specialist.

Infections and hematoma are intermediate in severity and everything possible should be done to prevent them. Thus, we have regularly used antibiotics, as discussed, above, and advocate meticulous hemostasis. A drain is most often not necessary but should one be indicated later it is applied. In the acute wound, only interrupted sutures are used so that only the necessary number may be removed to establish drainage without disrupting the entire wound, as would be necessary with a continuous suture.

In scar revision, wherever possible, we like the continuous subcuticular suture but do not advise this for the acute injury. A simple way to later combine removal of stitch marks with the broken-line repair or "W-plasty" is shown. Note the stepped closure and the need to remove a triangle of skin at each end of the excision to compensate. For revisions the soft tissue techniques described, are carried out after 6 months or preferably 1 year. Waiting until older childhood years or young adults years may be advisable, depending on the child's cooperation balanced with the psychic trauma and disfigurement may create. This requires judgment tempered by wisdom coordinating not only the maturity of the wound but the cooperation of the child and parents, along with the parents' anxiety and concern.

Hypertriophic scars stay within the margins of the original wound and tend to soften or clear. Keloids, on the other hand, proliferate beyond the original wound and increase with time. Keloids may be symptomatic with itching and burning, which may be difficult to control in the child and may add to the increased tissue reaction from scratching, leading to more severe keloid formation.

Keloid management has been addressed elsewhere by the senior author. Besides meticulous handling of the closure, one should stimulate as little as possible new tissue reaction outside the lesion into the normal tissue. Intrallesional injection and intrallesional excision are therefore in order. Kenalog injection and pressure dressings are chief additions to management. Topical steroids may help with itching and in a minor way assist in preventing buildup. After one's own second failure or recurrence, strong consideration should be given to systemic steroids, in addition to the above, and low-dose radiation therapy starting immediately after wound closure. We are more and more aggressive with initiating injections as soon as there is any hint of increased reaction in the healing wound.
Summary

This chapter has attempted to give the reader a practical approach to the management of soft tissue injuries in children. Some technical expertise is vital to the proper management of these wounds. Meticulous attention to detail, starting with the emergency consultation, can prevent unnecessary loss or disturbance of tissue and avoid the need for secondary reconstruction and a second operation for the child.

An attempt has been made to address aspects of the subject peculiar to children. In our times severe injury occurs at any age but statistically, if one includes teenagers, the frequency and severity increases as the child enters the teens. Often there is too much, and too early, personality expression with high-powered vehicles, as the teenager needs to observe his rite of passage.

Soft tissue trauma in children requires early wound management as well as the overall handling of the injured and frightened child and perhaps, no less important, the concerned, anxious, and sometimes angry parent. The objective is the prevention of immediate complications and long-term disfigurement. One should document the severity of the injury with photographs. Because of the visibility of the face and the importance to the growing child's self image, considerable attention should be given to this aspect of the injury, to the preparation of the parents for the healing period, and to the possibility of secondary revisions.

From the beginning maximum attention should be given to the primary repair with the exploration and preparation of the skin margins and meticulous layer closure. Most secondary repairs should not be carried out before 6 months and often a 1-year waiting period is preferable. With time, anticipated secondary repair may become unnecessary. There is no need to wait until adulthood to repair these secondary scars, which carry considerable psychological implications; however, there is the practical aspect of waiting until the child is able to cooperate and has been adequately prepared by the physician, his staff, and the parents.

The general considerations, surgical techniques, and management are similar for children and adults, requiring the whole spectrum of scar prevention and reconstruction. As a rule, with a child it is probably better to do the simplest repair possible at the time of the injury and reserve complicated scar camouflage techniques, Z-plasties, and regional flaps for the secondary repair.
Curious, rambunctious, energetic, and often clumsy, children routinely suffer small hurts. Fortunately, the natural elasticity of their facial bones, abundant padding, and the prominence of the skull provide anatomical protection and account for the rarity of facial fractures in childhood. Nevertheless, in our fast-paced culture, children do occasionally sustain severe injury to the maxillofacial skeleton requiring appropriate therapy. The principles in the management of facial trauma are the same for all age groups. However, the reconstructive techniques for children must be modified to accommodate their unique anatomy, rapid healing, fragile psychology, and, most importantly, their potential for facial deformity as a consequence of altered facial growth.

**Epidemiology**

Pediatric facial fractures account for approximately 5% of all maxillofacial injuries, although the incidence has been reported to be as low as 1.5% and as high as 15%. Children under the age of 5 have a substantially lower risk ranging from 0.9% to 1.3%. With increasing age, there is a rise in the incidence of facial fractures; however, it is not until late adolescence, with the maturation of the face and the adoption of a fast-paced life-style, that the frequency and distribution of facial fractures evolve into the pattern seen in the adult population.

There is an overall male preponderance among children with facial fractures varying between a ratio of 1.5:1 to 2.4:1. In children under 8 there is no difference between genders; between 8 and 15 years of age the male to female ratio rises to 4.5:1, reflecting the more aggressive and risk-taking behavior of preteen and adolescent boys.

There is a consensus that nasal fractures are the most common facial bone injuries in children. Precise statistics on their frequency are difficult to determine because many nasal fractures are treated in an office setting, whereas other types of fractures are usually managed in hospitals. In a series of 560 facial fractures, 60% were of the nasal bones. Another study reported a 45% incidence of nasal fractures among 122 children with facial injuries.
Mandible fractures are the most common facial fractures in children requiring hospitalization. In reports including nasal fractures, the incidence varies between 20.7% and 32%. The condyle is the most vulnerable part of a child's mandible and is the site of fractures 40% to 70% of the time. Body fractures (10% to 20%), angle fractures (3% to 17%), ramus fractures (3% to 10%), and symphyseal fractures (2% to 30%) occur less frequently. Dentoalveolar fractures, like nasal fractures, are often treated in an office setting; hence, it is difficult to gauge their true frequency, which is likely higher than the 14% incidence reported in one large series.

Mid- and upper facial fractures are quite rare in children. Several authors have reported a wide range of anatomic distribution, including 2.7% to 13.5% zygomatic complex fractures, 0% to 16% Le Fort-type fractures, and 0.2% to 19.4% orbital fractures. The sporadic distribution of upper facial injuries in children highlights their uniqueness and emphasizes how limited one surgeon's experience may be with these complex pediatric injuries.

Associated injuries are a common feature of childhood maxillofacial trauma. McCoy et al described 57% having injuries other than facial wounds, including 41% with skull fractures, 14% with cerebrospinal fluid (CSF) rhinorrhea, 9% with extremity fracture, 3.6% with blindness, and a 25.6% with aspiration pneumonitis. McGraw and Cole reported an incidence of 88% associated injuries with 61% multiple associated injuries in 72 children with facial fractures. Skull fractures or intracerebral trauma occurred in 61% of patients, particularly in younger children. Soft tissue injuries of the face occurred in 63% and temporal bone fractures occurred in 17%. In Kaban et al's series, 26% had associated injuries, including 6% with craniocerebral injury.

**Etiology**

The types of traumatic events that result in injury to a child's face, and the unique anatomical features of a child's facial skeleton that predispose it to the pattern of injuries observed in the pediatric population are two important factors to be considered when examining the etiology of pediatric facial fractures. In 1899, Lang described the case of "Hugh, S, age 13, who was struck on the right eyebrow, as he was running in the street, by the shaft of a cart driven at a trot". This injury resulted in a traumatic enophthalmus with diplopia and residual facial deformity. Since this early and famous case report, road traffic accidents have been a well-recognized cause of serious childhood maxillofacial injuries. McCoy et al reported that motor vehicle accidents accounted for 45% of the injuries in their series (20% pedestrian, 25% passenger). In Hall's series, 27% were due to motor vehicle accidents (5% pedestrian, 22% passenger). Reil and Kranz described a 37% incidence of motor vehicle accidents as the cause of trauma in their series (21% pedestrian, 16% passenger). McGraw and Cole found 67% incidence of motor vehicle accidents (25% pedestrian, 42% passenger). They emphasized that although the overall incidence of passenger injuries was higher and gender neutral, pedestrian injuries among boys under 5 outnumbered passenger injuries.

Although the major cause of serious pediatric facial fractures are from motor vehicle accidents, it turns out that childhood play is the most common cause of these injuries (15% to 30%), when all forms of fractures are considered, including nasal fractures and
dentoalveolar fractures. Other etiologies include bicycle accidents (2% to 20%), falls (20% to 30%), blunt strikes to the face (8% to 30%), and sports injuries (2% to 30%).

Two other causes, although uncommon, deserve special mention. These are birth injuries and child abuse. Prolonged and difficult deliveries result in a wide range of facial injuries, most of which are mild and transient. The more severe ones, such as soft tissue lacerations, facial fractures, and facial nerve injuries are usually due to forceps compression. Facial fractures, as a consequence of child abuse, have been reported. All emphasize the importance of considering such a possibility, especially in facial injuries among children under 5 years of age, since the great majority of all cases of child battering occur in this age group. Consideration of abuse requires reporting to the appropriate child protective agencies.

Both social and anatomical differences account for the disparity in the incidence and in the nature of facial fractures between children and adults. During child's play falls are frequent but generally from low heights and low speeds; hence the likelihood of serious fractures is diminished. Although children can be subject to the same hazards of modern life as adults, they usually live in protected environments that reduce the risk of major injuries.

Paralleling the changing social environment of a growing child, is the maturation of the facial skeleton, an appreciation of which is necessary to understand the difference in the patterns of fractures between children and adults. About 80% of cranial growth occurs in the first 2 years of life. Facial growth is also rapid during this period, but it is only after the second year that facial growth outpaces cranial growth. Brain and ocular growth are near completion by age 7; however, facial growth continues into the second decade of life. The cranial volume of the newborn in proportion to the facial volume is 8 to 1. In the adult this ratio is 2 to 1. The higher craniofacial ratio in children results in a greater proportion of impacting force being absorbed by the cranium, especially the prominent forehead, which overhangs the face. This accounts for the higher proportion of pediatric skull fractures compared to facial fractures and the rarity of serious midface fractures in young children, in whom the force necessary to cause major maxillary disruption often results in brain injury and death.

Another characteristic of the face of young children that reduces their likelihood of facial fractures is the more soft and elastic bone protected by thick layers of fat and muscle, and unweakened by the development of the paranasal sinuses. The greater elasticity, which is due to the thin cortical plates and a greater proportion of cancellous bone, also explains the higher incidence of "greenstick" fractures in children. The presence of unerupted and mixed dentition in the maxilla and mandible renders these structures more stable and resistant to fracture, despite fractures that can occur through the developing tooth crypts. As the child nears adolescence and adulthood the concomitant pneumatization of the paranasal sinuses, maturation of the dentition and bone, thinning of the facial soft tissues, concurrent with more risk-taking behavior, renders the face more liable to fracture.

**Emergency Management**

The initial assessment of the child with a facial fracture should follow the basic principles of trauma management. A primary survey must be made of the child's airway, breathing, and circulation. A hierarchy of airway interventions are available depending on the
severity of the injury. In most cases, where the only injury is to the maxillofacial area, careful posturing of the child is adequate. If indicated, the oral cavity should be suctioned of blood, secretions, loose teeth, and bone fragments. In case of mandibular displacement, a midline traction suture on the tongue can be most helpful.

Orotracheal intubation is generally indicated when there is coincidental cranial trauma, severe midfacial fracture associated with bleeding or oropharyngeal obstruction, and posterior retrusion of the mandible. This should ideally be accomplished after radiographic evaluation of the cervical spine. Consideration of performing the intubation in the operating room with rigid instrumentation is appropriate if there is anticipation of a difficult intubation, as with concurrent oropharyngeal or laryngeal injuries.

Tracheotomy is generally not necessary for routine facial fractures; however, it should be considered for severe panfacial injuries, and is routinely performed when a major facial fracture is associated with intracranial, thoracic, or abdominal injuries. Crash tracheotomies and cricothyrotomies on children in the emergency department should be avoided in favor of orotracheal intubation, with a controlled tracheotomy performed under general anesthesia in the operating room.

Blood loss from the highly vascular broken face of a child can rapidly result in hypovolemic shock. This blood loss may be a double threat if it is lost into the airway. Immediate volume expansion with crystalloid solution via large bore intravenous lines is necessary. With severe hemorrhage, transfusion with type specific red blood cells and other blood products, such as plasma and platelets, may be appropriate.

The secondary survey of the head and neck proceeds in an orderly fashion beginning with the neurologic assessment, evaluation of the neck and cervical spine, inspection of the eyes, otoscopy, rhinoscopy, and finally examination of the face and oral cavity. Important to the neurologic assessment of facial fractures are the sensory function of the fifth nerve and the motor function of the seventh nerve. Ophthalmologic evaluation is important to rule out intraocular trauma, and should include ophthalmoscopy as well as tests for range of motion, diplopia, and pupillary reflexes. Otoscopy is necessary since anterior canal wall injuries usually indicate a condylar fracture and a hematotympanum may suggest a temporal bone fracture. Anterior rhinoscopy is essential for the evaluation of septal injury, septal hematoma, or CSF rhinorrhea.

Examination of the facial skeleton should begin with inspection followed by manual palpation. Suggestive signs of facial fracture may include facial asymmetry with edema, ecchymoses, periorbital swelling, trismus, and malocclusion. Bimanual examination of the facial skeleton begins over the zygomatic arches and systemically proceeds down the face. The examiner feels for asymmetry, tenderness, and crepitation - all indications of underlying facial fractures. The malar eminences, the orbital rims, and the nasal bones are palpated. The stability of the maxilla is assessed by holding the head steady with one hand and rocking the premaxilla with the other, while observing for movement of the middle third of the face. Further information can be obtained by intraoral digital examination of the maxillary buttresses.
The manual examination of the mandible begins with the palpation of the temporomandibular joints and of the external auditory canals with opening and closing of the jaw. Fingers are swept across the skin covering the ramus, angle, and body of the mandible. Intra- and extraoral bimanual palpation of the body and symphysis completes the examination.

**Radiologic Examination**

Historically, pediatric facial fractures have been difficult to document radiographically. Much has changed with the advent of computerized tomography (CT), which has revolutionized the imaging of facial fractures. A CT scan in an axial plane is indicated for orbital and maxillary fractures. Direct coronal projections add substantive information especially in complex nasoethmoid and orbital fractures, but can be difficult to obtain in an injured, uncooperative child. Nevertheless, every effort should be made to obtain both projections for major injuries since the combined scans accurately define the aberrant anatomy in anticipation of correction. Three-dimensional CT reconstructions have also proved to be a valuable adjunct to two-dimensional CT for preoperative assessment and the surgical planning of facial fractures.

For mandible fractures, the most useful diagnostic radiograph is the panoramic view, which displays the total anatomy of the mandible including the condyles and the upper and lower teeth. Its proper performance requires that the child be seated and stably positioned. This is not always possible with severely injured or very young children, and alternate views may be required for documentation of their fractures. The posteroanterior projection demonstrates the entire mandible including displaced fractures. The anteroposterior projection, also known as the modified Towne's view, is specific for the condyles. The lateral oblique projection displays the condyle, coronoid, ramus, angle, and body. Intraoral projections can be helpful for the symphyseal and parasymphyseal area and with dentoalveolar fractures as well.

Axial CT scans are also useful for documenting mandible fractures, especially in a multiply injured child who cannot be readily positioned for routine mandibular radiographs. For those children who may require cranial and upper facial CT scanning, it is advantageous to get additional images that include the mandible.

The imaging of nasal fractures remains controversial because of the inaccuracy of standard nasal radiographs in isolated nasal trauma. This is especially true in children whose nasal bones are not fully fused, making radiographic interpretation difficult. Nevertheless, children being referred from the emergency department for clinical examination have routinely been x-rayed. Perhaps there is a medicolegal justification or psychological benefit; however, they are generally not useful. On the other hand, a significant nasal fracture in a child that results in flattening of the nasal dorsum warrants proper imaging with axial and coronal CT scan.

**Facial Growth**

Alterations of facial growth as a consequence of childhood facial fractures are well recognized. However, malformations attributable to early trauma are not inevitable. Normal development of the face results from the absolute growth of the face and the relative
proportional modifications that distinguish the shape of child's face from that of an adult. It is the disturbance of the differential growth rates attributable to the injury that will result in anomalies at maturity. A detailed accounting of the morphology of facial growth is beyond the scope of this chapter. However, an understanding of facial development, particularly from a pathologic and a physiologic perspective, may aid in the differentiation of injuries most likely to interfere with facial growth and facilitate the identification of reconstructive techniques that will best promote normal facial maturation.

Pathologic changes caused by accidental and experimental trauma to different parts of the facial skeleton are documented in the literature. Grymer et al studied 47 adult patients who had sustained nasal fracture during childhood and compared them to a control group of adults who did not have a history of nasal trauma. They also analyzed the effects of these fractures based on the period of facial development during which the injury had occurred. This was premised on observations that there are three periods of nasal growth: from the age of 1 to the age of 6 years, when there is rapid growth; from 6 to 11 years, when there is a period of slow growth; and from 12 to 16 years, when there is a second period of rapid growth. They found a significant increase of bony and cartilaginous deformities in the fracture group. However, they found that nasal deformities as a consequence of trauma during specific developmental periods were equally distributed among all their patients.

Rock and Brain did a cephalometric analysis of 29 adults with a history of nasal fractures and compared these to a control group without a history of trauma. They found significant reduction in the size of the midface, reduced projection of the nose, and an increase in vertical height.

Sarnat and Wexler did a series of experimental resections of septal cartilage and bone in the septovomerine angle of growing rabbits. This caused a deceleration of the growth of the snout, which resulted in a reduction in the size of the nasal and premaxillary bones, the nasal cavity, and the piriform aperture. The extent of the deformity varied with the amount of tissue resected. He concluded that the relationship of the septum to the growth of the face is comparable to the relationship of the eye to the growth of the orbit, and the brain to the growth of the skull. In a similar vein, Siegel resected the nasal septum in growing baboons and noted diminished upper facial growth.

Osterhout and Veargervic described three patients who sustained midfacial trauma in early childhood and were found during adolescence to have midfacial hypoplasia requiring Le Fort III maxillary advancements.

Precious et al described three patients, victims of child abuse, with a history of nasomaxillary fractures, who were studied with cephalometrics during adolescence. The children were found to have an elevation of the anterior palatal plane, a reduction in the length of the premaxilla and nasal spine, decreased nasal projection, and modifications in the occlusal plane of the mandible.

Bachmayer et al demonstrated that Le Fort III advancements in children with Crouzon's, Apert's, and Pfeiffer's syndromes result in a cessation of any further forward growth of the maxilla. Although they noted that these patients already had diminished maxillary growth due to premature fusion of some facial sutures, they concluded that the
combination of traumatic osteotomies and periosteal stripping stopped further growth.

Shapiro et al did Le Fort I advancements in monkeys and followed the animals for 2 years with serial cephalometrics. They found that the animals had diminished anterior maxillary growth, which they attributed to the formation of scar tissue.

Munro studied the effects of total maxillary advancement in pigs, varied with other factors including periosteal elevation, simple osteotomies, fixated osteotomies, and bone grafts. He found that growth in the areas adjacent to osteotomies was reduced, but by no more than the reduction caused by periosteal elevation. He concluded that periosteal elevation decreased local blood supply, increased periosteal adherence, and changed local bone growth.

Hellquist did an extensive study on the effect of periosteal resection on the facial growth of rodents. He found that the periosteum quickly regenerated from the denuded bone surface. Although this new periosteum appeared normal, the underlying bone showed altered morphology extending beyond the area of resection. When periosteal stripping was confined to the premaxilla, growth was not altered; however, when it involved the maxilla, growth was significantly diminished.

Lindahl and Hollender radiographically studied the process of condylar remodeling in children and adults after fracture. They found that among children between 3 and 11 years of age, extensive remodeling of the condylar processes resulted in normal anatomy. In 12- to 19-year-olds, remodeling occurred but to a lesser extent. In adults, only minimal remodeling took place.

MacLennan reviewed 180 cases of condylar fractures of which five were in children under the age of 10. He found that the great majority of these injuries healed without functional impairment. However, he did note that crush injuries of the condylar head before the age of 5 predisposed to ankylosis and growth disturbances.

Walker, in a study of the treatment of condylar fracture dislocations in monkeys, found that condylar reformation was comparable whether management was conservative immobilization, early mobilization, or direct surgical wiring.

Anderson and Alling studied subcondylar fractures in young dogs and found that there was no difference in the mature mandible if the fractures were untreated or repaired by wire fixation. They also performed a condylectomy in one animal and found complete regeneration of the condyle with normal symmetry of the mature skull.

Studies of the normal processes of facial growth date back to John Hunter, who in 1835 described the normal development of the mandible. He fed madder, the root of a plant that has the property of staining growing bone a red color, to two young pigs for a month. He sacrificed one pig after a month, whereas the other pig was fed a regular diet for an additional month before sacrifice. He then studied the pattern of madder staining on the mandibles and found that the bone that had been the condyle during the staining diet had been incorporated into the ramus, whereas unstained bone, which had grown after the madder diet had been stopped, was now the condyle. He also noted that what had been the madder-stained anterior border of the mandible in the first animal had nearly disappeared in the second
animal.

Clinical experience with the pathologic influence of trauma and experimental observations such as Hunter's provide an insight into the basic concepts of the processes of facial growth. These have been extensively reviewed by Enlow and are briefly summarized here.

Bones grow by the deposition of bony tissue on one surface and absorption on the opposite surface. This combines to produce growth movement or "drift" in a specific direction. Bones of the face are covered on the outside by periosteum and on the inside by endosteum. These osteoactive membranes cover the bone in a jigsaw-like pattern of growth fields, which are responsible for both bone deposition and resorption. This growth is not thought to be programmed within the bone itself, but in its covering osteogenic membranes and functioning soft tissues such as tendons, muscle, mucosa, and brain. The rate of activity in differential growth fields vary according to the specific function and growth of the surrounding soft tissues. Certain growth fields, such as the mandibular condyle, have special significance in the growth process. Nevertheless, it is a misconception to view them as isolated "growth centers", since all of the surrounding bone participates in the remodeling and enlargement that produces the mature facial form.

The process of remodeling and enlargement results in the relocation of the component parts to allow for overall growth and to provide structure for the changing physiologic conditions of the encapsulating soft tissues. It also results in each component bone being carried away from its adjacent neighbors, a process called primary displacement. Primary displacement occurs as a result of a bone's own growth, and its direction is generally in the opposite direction of bone deposition. For example, as the mandible is carried forward by its growing soft tissues, it is displaced anteriorly and inferiorly from the temporal mandibular joint. In response, the condyle and the ramus grow by deposition in a posterosuperior direction, in effect filling the space caused by the displacement. Similarly, as the nasomaxillary complex is displaced anteriorly and inferiorly, bone deposition occurs at the multiple sutural interfaces between the maxilla and the adjacent bones of the face. In other words, bone growth at the sutures is not responsible for the projection of the maxilla, but is the result of it. Furthermore, the bones of the face are also secondarily displaced by the enlargement of the adjacent bones, such as occurs with the downward and forward displacement of the maxilla as a result of cranial enlargement.

It is clear from the foregoing discussion that facial growth is a complex process of multiple anatomical and functional interrelationships, no part of which is independent of others. What is not obvious is the mechanism of control and coordination responsible for the overall pattern of growth. Although many possible explanations have been proposed, none completely explains how the morphogenetic process works and what occurs during facial development. A short overview of the major concepts of facial growth follows; for a detailed discussion the reader is referred to the works of Scott, Moss, and Enlow.

One need only see a parent and child who look alike to understand the genetic influence on facial growth. Nevertheless, there remains deep uncertainty in attributing all facial skeletal development to an intrinsic blueprint within the bone-producing cells of the face. Rather, contemporary thought holds that growth is controlled by the genetic program
within the enveloping soft tissues and regional growth sites. The nasal septum has received much attention for its role in nasomaxillary development. It has been proposed as the structure responsible for the dramatic anterior and inferior displacement of the growing maxilla. Although it is doubtful that the septum is the only pacemaker of midfacial development, both experimental and clinical data support its role as an important regional growth site.

Moss has proposed a broader conceptual framework to explain facial growth and osteogenic regulation. Known as the "functional matrix" theory, it holds that the predeterminants of bony morphology reside in the encapsulating soft tissues. The brain, the nasal and pharyngeal mucosa, the facial musculature, the tongue, and the teeth form a functional matrix that surrounds the bone and directs its growth and form. Direct and indirect stimuli produced by the growth and action of these functioning tissues switches on the osetoactive cells to deposit and absorb bony tissue, thus displacing, remodeling, and enlarging the facial skeleton in order to accommodate the physiologic activities of the face.

Based on analysis of the known effects of trauma in the growing face as well as on an understanding of the operational mechanics of facial growth, certain practical concepts for the repair of facial fractures in children emerge. The mandible appears to be generally resistant to abnormalities of growth as a result of trauma, unless there is alteration of its function due to injury in or near the temporomandibular joint. Early restoration of mandibular mobility is desirable in order to facilitate the restorative bony changes that result from normal function.

The nose, nasoethmoid complex, and maxilla are more prone to growth abnormalities as a result of trauma. This probably occurs because of the minor restorative functional movement that these bones are subject to, the physiologic derangements that result from the fractures, the importance of the septum as a regional growth site, and the vulnerability of the multiple suture sites to scar formation. All these factors suggest specific interventive strategies that include:

1. careful restoration of injured soft tissue, particularly the periosteum;

2. close attention to septal injuries with an emphasis on realignment rather than resection;

3. reduction of fractures into their stable anatomic locations;

4. correct realignment of suture lines;

5. minimal periosteal elevation;

6. three-dimensional, stable fixation of complex fractures;

7. use of rigidly fixed bone grafts as a substrate for growth in areas of bone loss.

The issue of rigid plate fixation in children warrants additional consideration because of the conceptual implications of its use with regard to facial growth. There is only one study that has investigated the effect of plate fixation in growing animals. Lin et al did frontoorbital
craniotomies on an expanding portion of the craniofacial skeleton in two groups of kittens. After removal, the cranial segment was replaced into its anatomically correct position. In the first group, fixation was accomplished with wire and in the second group fixation was obtained with a mini-compression plate. As controls, a third group of kittens had straight plate fixation across the coronal suture without osteotomy, and a fourth group had incision and periosteal elevation alone. Growth analysis was accomplished with volumetric determination by three-dimensional CT and cephalometrics after sacrifice. The results showed significant growth restriction in both the osteotomized groups but not in the group with the plate across a coronal suture. Moreover, the unoperated side showed a significant compensatory growth in the osteotomized kittens. It is difficult to draw any firm conclusions from this study, but it does suggest some measure of safety with the use of plate fixation.

Nevertheless, plate fixation with our current state of knowledge should be considered only for children with complex three-dimensional injuries that cannot be repaired by simpler means. There are several reasons for such caution. Access for plating often requires surgical trauma to facial soft tissues and extensive periosteal elevation, both important factors in alteration of facial growth. Bone is deposited at facial sutures as a result of tension fields across them. Plating across a suture line could potentially convert the sites to compressive fields, resulting in bony resorption. Plate and screw fixation also risks injury to the maxillary tooth buds. The circulation of the injured facial bones, already compromised by fracture and the extra trauma of exposure, is further jeopardized by the plates and screws, in that immediately after placement no circulation is seen around the drill holes and under the implant for several weeks. There is also the question of whether plates can be indefinitely left in place or should be removed after the facial bones have healed. Since the effect of plates on facial growth remains unknown, consideration of removal is valid and must be weighed against the additional injury to facial soft tissues required for their removal.

Despite these reservations, recognizing that each reconstructive technique has its strengths and its weaknesses, the use of rigid fixation is highly desirable in complex fractures where the original features are difficult to restore. The alternative of no correction is unacceptable, and the use of interfragmentary wires is tedious and may lead to reconstruction of uncertain stability. As Rahn has written, "open reduction of a fracture may sometimes appear to be an aggressive approach. It can be justified when late reconstruction and secondary surgery is avoided. Thus an approach that appears initially to be more aggressive, may be more conservative."

Nasal Fractures

The proportionate nasal anatomy of the child differs substantially from that of an adult, and this results in a different pattern of injuries in response to trauma. The primary projecting component of a child's nose is cartilaginous and readily deforms during a blow to the midface. The impacting force is dispersed across the maxillary soft tissues, resulting in a broad area of edema with a loss of anatomical specificity of the nose on examination.

The anatomy, orientation, and compliance of the lower and upper lateral cartilages allow for a ready rebound from a traumatic deformity, and these cartilages rarely sustain permanent injury, except for dislocations of the upper lateral cartilages from the bony framework. However, the more rigid septum, within its tight perichondrial covering and
surrounding bony encasement, is much more prone to long-term damage.

Three types of injury can affect the septum. The first results from the detachment of its perichondrial covering in response to the deformation of the cartilage during impact. This creates a potential space between septum and its perichondrial leaflet and allows the formation of a septal hematoma. The second type of injury tears the anchorage of the septum inferiorly from the maxillary crest and posteriorly from the perpendicular plate of the ethmoid and vomer. The resulting septal dislocation can be an immediate cause of nasal obstruction. Moreover, since the vomerine region of the septum is its most actively growing part, growth disturbances, generally of the hypertrophic variety, are a common late sequela. The third type of injury is a fracture of the septum, which can occur either vertically, horizontally, or in stellate fashion. This can result in immediate or subsequent obstruction, delayed twisting deformities, and growth disturbances. A mixed picture of the septal injuries is typically encountered.

The nasal bones are not very prominent in young children and thus uncommonly injured. They are surrounded by the nasomaxillary and the nasofrontal sutures and divided by a midline suture. Injuries will often result in "greenstick"-type sutural dislocations, commonly just on one side. When the blow is directly to the midline an "open book"-type fracture can occur resulting in central depression, lateral flaring, and dislocation of the nasal bones over the frontal process. With fractures of the nasal skeleton, one must have a high index of suspicion about more extensive but occult nasoethmoid and orbital injuries.

The initial examination of a child with a nasal fracture may be of limited value with regard to the external deformity. However, immediate intranasal examination is essential to evaluate for septal hematoma. Suspicion should be greatest in a child who has difficulty breathing through the nose following injury. The clinical appearance of septal hematoma is that of a purple bulge in one side of the nasal vestibule with contralateral deflection of the septum. The area will be compressible to palpation with a cotton tip applicator and not responsive to topical vasoconstriction.

Septal hematoma is a serious complication when left untreated and can result in a thick fibrotic and obstructive septum. It may also become infected, resulting in cartilage necrosis and subsequent saddle deformity of the nose. Stucker et al suggest that abscess formation is not always necessary for there to be cartilage dissolution and collapse from an untreated septal hematoma.

The treatment of septal hematoma should begin with needle aspiration of the suspicious area after topical anesthesia. Since this is not always possible in a child, a general anesthetic may be necessary in order to evaluate and evacuate the hematoma via a hemicranstifxion incision. This can also provide an opportunity to explore the entirety of the septal injury with suture reduction of the displaced fragments. Following exploration, the mucoperichondrial leaflet is sewn back to the cartilage with continuous through and through chromic sutures with a mini-Keith needle. If necessary, the septum can be further supported with Silastic splints. The nose is always packed with antibiotic-impregnated gauze for 2 to 3 days. Postoperatively the child is kept on broad-spectrum antibiotics.
During the anesthetic the bony nasal pyramid is examined, and if there are displaced fragments, these are manipulated into place with closed-reduction techniques. Unfortunately, severe facial edema may preclude precise examination and reduction, necessitating later reevaluation and possible rereduction.

If septal hematoma has been ruled out and there remains concern about the possibility of nasal fracture, the child is asked to return in 3 to 4 days. This waiting period will have allowed the subsidence of a majority of the facial edema and will allow for a more accurate examination. Crockett et al suggest that this is also a good time for the parents to find a pretrauma photograph for use as an additional guide to the child's normal anatomy.

Definitive management is initiated if there are demonstrable bony and septal fractures resulting in cosmetic deformity and/or airway obstruction. In most circumstances, closed reduction of the bony component is accomplished with intranasal instrumentation and external manual manipulation. However, "greenstick" fractures may not always readily reduce into the desired position. For these, open reduction with completion of the fracture with a small osteotome will allow for proper alignment of the fragments. Once the bone have been relocated, they will require support with intranasal packing and an external cast for several postoperative days.

Septal injuries are more difficult to control with closed techniques. Attempts should be made to reduce the dislocations and realign fracture segments with instrumentation, Silastic splinting, and packing. However, if it becomes obvious intraoperatively that conservative measures are inadequate, then perichondrial elevation and cartilage exposure is necessary. Care is taken to preserve the integrity of the mucoperichondrial leaflets and the emphasis of the open technique should be toward suture realignment of the septal fragments with minimal resection of cartilage.

Newborns

An occasional problem is asymmetrical tip deformities in newborns. These infants typically present with a flattening of the nasal tip to one side with the septum tilted in the same direction. The bony dorsum is invariably straight. It is difficult to know whether this type of deformity is an acute traumatic birth injury or due to prolonged intrauterine positional pressure.

There are some nasal surgeons who advocate immediate surgical reduction of these deformities by straightening and relocating the septum. However, experience has shown that these deformities self-straighten over time without late sequelae. Treatment for these children is to reassure the parents that the nose in time will straighten out. Although it is hypothetically possible for such a deformity to cause airway obstruction in the nasally obligate neonate, this has not been a problem even with severe deformities.

Mandible Fractures

The goal of mandibular fracture management in children and adults is correct alignment with proper fixation in centric occlusion. However, the techniques required to achieve this goal vary with age and the quality of dentition. Before the age of 2 the eruption
of the deciduous teeth is incomplete; hence, it is difficult to achieve adequate anchorage for immobilization. On the other hand, inaccurate alignment is generally compensated for by later growth. Between 2 and 5 years of age the deciduous incisors have firm roots, and if the deciduous molars have formed, both of these can be utilized for cap splints or arch bars. Between 5 and 9 years of age, when the deciduous incisors fall out and the deciduous molar roots are resorbed, is the period that presents the toughest problems for the establishment of occlusion. Reliance on circummandibular and circummaxillary stabilization with Gunning-type splints is often necessary during this time. After 10 years of age, the development of permanent teeth provides safe anchors for fixation.

Condylar Fractures

Condylar fractures can be classified into three anatomically distinct types. The first two are intracapsular and include crush-type fractures of the condylar head and high condylar fractures through the neck above the sigmoid notch. The third type, which is also the most common, is a low or subcondylar fracture often of the "greenstick" variety. It is extracapsular and is from the sigmoid notch back toward the posterior ramus.

There are those who advocate open surgical treatment of condylar fractures in children; however, clinical and experimental observations overwhelmingly support a conservative, closed approach to the management of most of these injuries. For the majority of condylar fractures, the primary decision is whether to immobilize or not. In most cases, unilateral condylar fractures present with normal occlusion and normal mandibular movement. A soft diet and movement exercises are all that is necessary. This may also apply to bilateral condylar fractures where there is normal function. Condylar fractures presenting with an anterior open-bite deformity, retrusion of the mandible, or movement limitation indicate a brief period of immobilization lasting 2 to 3 weeks.

In children under 2 years of age and in children between 5 and 9 years of age, whose dentition does not allow for the application of arch bars, immobilization requires unconventional fixation techniques. An overlay acrylic mandibular splint is constructed and is held in place by circummandibular wires. Its occlusal surface is placed in normocentric relation to the maxilla and immobilization is accomplished by suspending a wire from the piriform aperture and tightening it around the midline wire, which is holding the splint to the mandible.

In children with stable deciduous or permanent teeth, conventional arch bars are the preferred technique for immobilization. These can be fixed in place with 26- or 28-gauge stainless steel wire. Rubber bands will suffice to provide traction for intermaxillary fixation. Arch bars can be further reinforced with circummandibular wires and suspension wires from the nasal spine or piriform aperture.

The indications for an open surgical approach of pediatric condylar fractures are quite limited and are reserved for situations where there is a mechanical obstruction to normal movement or in the extremely rare cases of dislocation of the condyle into the middle cranial fossa. The preauricular exposure gives excellent access to the condylar head and temporomandibular joint. This approach unfortunately risks injury to the facial nerve. The submandibular technique of Risdon is safer to the upper branches of the facial nerve, but
provides exposure primarily for subcondylar fractures.

**Symphyseal and Parasymphyseal Fractures**

Condylar fractures are most commonly associated with fractures of the anterior arch of the mandible, either at the symphyseal or parasymphyseal regions. The submental musculature exerts a downward and retrusive force to these areas; hence, fragments may often be displaced accordingly and may complicate reduction. Symphyseal or parasymphyseal fractures with minimal to moderate displacement can often be realigned with careful manual manipulation under anesthesia and immobilized with a cap splint, arch bar, or interdental wiring. In younger children under 3 years of age, an acrylic splint held in place by circummandibular wires is most useful.

The problem with these simple techniques is that reducing the fracture with an arch bar or interdental wires on the side of the dentition (tension surface) will result in distraction of the lower border (compression surface). In order to better reduce serious misalignment, open reduction with internal fixation of the fragments is required. These midline areas are readily exposed via an intraoral degloving approach. The lower border can then be reduced by wire fixation or by monocortical reconstruction with miniplates. With both techniques, great care must be exercised in drill hole placement in order to prevent injury to the developing tooth buds. With both miniplate reconstruction and with interosseous wiring, a brief period of intermaxillary fixation of 2 to 3 weeks is required. In children with full, permanent dentition the principles of bicortical, compression-plate reduction are applicable.

**Body and Angle Fractures**

As with condylar fractures, body and angle fractures are often of the "greenstick" variety presenting as monocortical cracks. These children will typically present with normal occlusion and movement and thus are best treated with a soft diet and symptomatic therapy. When the fractures are displaced, treatment will depend on the availability of the dentition, direction of muscle pull, and the degree of distraction. For distracted body fractures, intermaxillary fixation with elastic traction is usually adequate. However, if misalignment of the lower border cannot be controlled in a conservative manner, then open reduction with internal fixation with interosseous wiring or monocortical minicompression plating is necessary. Although both of these can usually be accomplished intraorally, the open reduction of posteriorly placed body fractures may require an external approach in some children.

The difficulty with angle fractures is that they lie beyond the dentition and thus may not be amenable to reduction with splints or intermaxillary fixation when the fracture is displaced and under unfavorable muscle tension. Under such circumstances extraoral open reduction with wire or plate fixation is required.

**Dentoalveolar Fractures**

Dentoalveolar fractures constitute a dental emergency because salvage of the traumatized teeth requires prompt reimplantation, generally within an hour of the injury. Although loss of the deciduous teeth is not a problem, identification of tooth type, whether primary or permanent, especially during the period of mixed dentition is difficult for the
physician unfamiliar with pediatric dentition. The typical injury is to the mandibular or maxillary incisors and canines, due to their prominent anterior position. The fractures can involve the crown and the deep pulp, or there can be partial or complete avulsion with loss of the surrounding cortex. Treatment is directed at getting the child seen immediately by a dentist for definitive therapy. In the interim, the tooth is gently cleansed in saline, handled by the crown, and if the child is cooperative, replaced in the socket. If the child cannot cooperate, then the tooth is kept in saline-sauked gauze or a bowl of milk until such time as a dentist can reimplant and stabilize the tooth with either a splint or an arch bar.

Maxillary Fractures

Fractures of the maxilla in children are classified as in adults. Le Fort I fractures separate the palate from the maxilla, extending through the floor of the nose, maxillary sinus, and the pterygoid plates. Le Fort II fractures separate the midface from the cranium, extending through the pterygoid plates, along the lateral and anterior maxillary walls, the medial orbital wall, and the nasofrontal suture. Le Fort III fractures separate the entire face from the cranium extending through the zygomatic arch, frontozygomatic suture, lateral orbital wall, medial orbital wall, nasofrontal suture, septum, and the pterygoid plates.

These rare fractures present with severe facial edema, prominent orbital ecchymoses, anterior open bite, and other occlusal deformities. Associated injuries such as basilar skull fractures and dural tears with cerebrospinal fluid fistulae are common, since a force great enough to fracture the face will often be transmitted to the cranial vault.

The goals of therapy are to reestablish facial symmetry, appropriate occlusion, and normal vertical dimension. Significant fracture displacement must be reduced in 4 to 8 days or as soon as the condition of the child permits, since rapid interfragmental healing makes adequate correction extremely difficult after 10 days. Acute reduction should be considered when fractures are accessible through open wounds.

Injuries with minimal or no displacement do not require correction. Active intervention is necessary when displacement has altered form or function. Reestablishing occlusion by intermaxillary fixation with splints, arch bars, or interdental wiring is generally the first maneuver in the repair of these injuries. Intermaxillary fixation techniques utilized to overcome the unique problems of pediatric dentition are those previously described for mandible fractures. Additional simple intervention such as the suspension of Le Fort I fractures with piriform aperture wires and suspension of Le Fort II fractures from circumzygomatic wires remain time-tested alternatives to open reduction and internal fixation.

Historically, complicated Le Fort II and Le Fort III fractures, as well as panfacial fractures, have required craniomaxillary and craniomandibular fixation. This is based on the concept that the solid foundation of the cranium and correct occlusion are required for accurate reconstruction of the face. Traditionally this was accomplished with a plaster head cap with outrigger suspension wires, which supported the fragments by external traction. Later, halo-type external fixateurs with direct pinning or fragmentary suspension were developed for the management of complicated fractures. Although these techniques were often successful, they presented problems of patient incompatibility, particularly in young children.
Over the last decade, the availability of miniplate and microplate screw fixation systems have made internal, three-dimensional, rigid fixation a preferable alternative to external suspension. The injuries are approached via hidden facial degloving incisions, inferiorly via the maxillary gingivobuccal sulcus and superiorly through a bicoronal incision.

The inferior approach provides access to the entire maxilla, laterality to the masal bone and zygoma, anteriorly to the infraorbital nerve, and medially up to the lacrimal fossa. The exposure provides for the reestablishment with individually contoured plates of both the lateral zygomaticomaxillary buttress and the medial nasomaxillary buttress. It also allows for rigid interfragmentaly fixation of the entire midface below the level of the orbital rim. Additional exposure of the inferior orbital rim and floor can be obtained by subciliary or transconjunctival techniques.

The superior approach, which is subperiosteal over the cranium and subfascial over the temporalis muscle, provides access to the superior and medial orbital rims. Orbital roof and nasoethmoid exposure can be obtained by drilling the bone from around the supraorbital nerves. Detachment of the temporalis fascia from the lateral orbital rim and zygomatic arch reveals the bones of the entire upper face from the zygomatic root on one side completely around to the other. Access to the lateral orbital can be obtained by reflecting the temporalis muscle posteroinferiorly. The exposure provides a means to realign and rigidly fix the frontozygomatic suture, the entire zygomatic arch, and the nasal bones. It also allows for harvesting of cranial bone grafts for orbital reconstruction and cantilever nasal reconstruction in complex nasoethmoid fractures. Intracranial access for neurosurgical intervention or basicranial fracture reduction is also possible via this technique.

**Orbital and Nasoethmoid Fractures**

The orbit and its contents occupy a prominent position in the face of a child, and injuries to this area can have serious functional and cosmetic consequences. The scope of these injuries are related to the magnitude of the impacting force and can vary from relatively minor fractures, such as "blow-out" of the orbital floor, to complex fractures involving the rim, multiple walls, and the apex, with alteration of orbital volume, ocular mobility, and visual acuity. Management depends on the extent of the injury, ranging from observation to surgical intervention requiring craniofacial exposure, rigid fixation, and bone grafting.

Appropriate treatment requires accurate diagnosis best accomplished by physical examination and CT scanning. The first priority is an assessment of visual acuity. Periorbital edema, ecchymoses, and subconjunctival hemorrhage are indicative of orbital trauma. The position of the globe, often obscured by edema, is inspected for exophthalmos, enophthalmos, and vertical dystopia. Intraocular pressure is measured. The extraocular musculature is tested for voluntary range of motion and if necessarily with forced ductions under anesthesia. Intercanthal distance and the length of the palpebral fissures are measured, and the locations of the medial canthal ligaments are identified. The rims are palpated for disruption. The supraorbital and infraorbital nerves are tested for sensitivity.

Classified by its pattern of fractures, the orbit has three anatomically distinct regions. The anterior component is the hard bone of the orbital rim, which divides into three subsections. The first is the supraorbital rim, which is a portion of the frontal bone. The
second is the infralateral rim, which is part of the zygomaticomalar complex. The third is the medial rim, part of the nasoethmoid complex, and to which attach the medial canthal tendons. The middle component consists of thin lamellae of bone forming the roof, the floor, and the medial and lateral walls. The posterior component consists of the orbital apex, including the orbital fissures and the optic foramen.

**Orbital Rim Fractures**

**Zygomaticomalar Fractures**

Paskert et al have suggested a useful classification system of three types of zygomaticomalar fractures based on patterns of fracture seen on CT scan. Type 1 is incurred with low-impact trauma and results in separation of the frontozygomatic suture, infraorbital rim disruption with separation through the zygomaticomaxillary suture, fracture of the zygomatic arch, and cracks along the lateral orbital wall and floor. Type 2 fractures result from more forceful impacts and present with comminution of the inferior rim, the zygomaticomalar buttress and the orbital floor, along with separation of the frontozygomatic suture. Type 3 injuries are the most severe and often associated with Le Fort-type fractures. These result in total disruption of the zygomaticomalar complex. Typically, there is marked retrodisplacement of the malar fragment, comminution of the infraorbital rim, zygomatic arch, lateral orbital wall, orbital floor, and loss of both buttresses.

The surgical correction of type 1 and 2 injuries requires exposure of the infraorbital rim, frontozygomatic suture, and zygomaticomaxillary buttress. This can be accomplished via the combination of a brow and subciliary incision or a transconjunctival incision with lateral canthotomy in conjunction with an upper buccal sulcus incision. The bony fragments are manipulated into alignment utilizing the contralateral side as a guide to correct positioning. Type 1 injuries can be fixed with interosseous wires or microplates at the frontozygomatic suture and infraorbital rim. Type 2 injuries will often require additional plating of the zygomaticomaxillary buttress. Exploration of the orbital floor may reveal prolapsed orbital contents and the need for additional support with alloplastic implants or bone grafts. The zygomatic arch can be repositioned by elevation through the brow incision or the lateral canthotomy. Type 3 injuries may require a bicoronal craniofacial approach to reconstruct the zygomatic arch and lateral orbit.

**Supraorbital Rim Fractures**

In young children the stout bone of the supraorbital rim is not affected by the expansion of the frontal sinus and is thus resistant to injury. However, when high forces impact this area the consequences are serious, since fracture of this portion of the rim will often extend into a frontal skull fracture with intercranial injury. Moreover, without the protection of the frontal sinus, large forces will also be transmitted into the inner bone of the orbital roof, resulting in orbital "blow-out" fractures.

When these injuries occur they are often associated with overlying lacerations that allow for direct repair with either interosseous wiring or plating. If there is no ready access via an existing laceration, a bicoronal craniofacial exposure is appropriate. A brow incision may be sufficient when the need for exposure is limited.
Older children, in whom the frontal sinus has developed, and who have sustained supraorbital rim fractures must be evaluated and treated as adults with frontal sinus fractures. Isolated anterior wall injuries can be managed with appropriate interfragmentary reconstructions with wire or plates. Complex injuries require basicranial exploration for evaluation and repair of dural tears, assessment of the nasofrontal ducts, and reconstruction of the anterior wall. With supraorbital fractures, the injury is typically above the nasofrontal duct, so sinus obliteration is generally unnecessary. Should obliteration be required, the standard techniques of stripping of sinus mucosa, burring of the walls, muscle, or bone pate obstruction of the ducts, and filling the cavity with homologous fat can be performed.

**Nasoethmoid Fractures**

The three-dimensional saddle shape of the nasoethmoid region, with its strongly defined vertical and horizontal axes, subtle curves, careful proportions, and prominent symmetries, make injuries of this region the most difficult to reconstruct. Nasoethmoid fractures can vary from simple dislocations of a fragment of the orbital rim, to comminuted compound fractures with bilaterally shattered medial orbital rims, medial orbital walls, nasal bones, and complete disruption of the ethmoid labyrinth. Various classifications have been described, but all try to differentiate between simple medial rim injuries as opposed to extended complex injuries seen in conjunction with other midface fractures. Anatomically, a nasoethmoid fracture is defined by fractures of the nasofrontal suture, nasal bones, medial orbital rim, and inferior orbital rim. Such a four-sided fracture yields a core of bony pieces that Paskert et al call "the central fragment", the appropriate reconstruction of which will provide the optimal function and cosmetic results. The varying displacement and mobility of this central fragment also provides the physical clues for accurate diagnosis.

Measurement of intercanthal distances is mandatory but poses some difficulty. The bony intercanthal distance is often obscured by edema and there is no well-documented data about soft tissue intercanthal distances as a consequence of trauma in children. Nevertheless, some guidelines are available. Recognizing ethnic, gender, and normal anatomic variations, the mean bony interorbital width by 4 years of age is 19.5 mm, by 8 years of age it is 22 mm, by 12 years of age it is 23 mm, and at adulthood it is 25 mm. Experience suggests that the soft tissue intercanthal distance is about 5 mm wider than the bony interorbital width. An additional 5 mm of soft tissue intercanthal distance is indicative of, and 10 mm diagnostic of, displaced nasoethmoid fractures.

The contemporary management of nasoethmoid fractures requires adequate exposure to facilitate optimal correction. Existing lacerations can be utilized and, if necessary, extended into camouflaged areas such as the brow. Coronal exposure is the aesthetically superior choice when lacerations are absent. The major fragments and the medial canthal ligaments are identified. Great care is taken to preserve the attachment of the ligament to its bony insertion. The central fragment is mobilized in order to facilitate interfragmentary reduction and transnasal wiring.

The maneuver of greatest importance in the successful correction of nasoethmoid fractures is the correct setting of the medial canthal ligaments by transnasal wiring. Conversely, the easiest mistake in children is to set this intercanthal distance too widely, since interorbital growth is nearly complete by 8 years of age. A drill hole is made into the anterior
lacrimal crest just above the insertion of the anterior limb of the ligament. A second drill hole is made in the posterior lacrimal crest just behind the insertion of the posterior limb. Contralateral drill holes are similarly placed and 28-gauge stainless steel wire is passed transnasally between the two fragments and tightened in an effort to overcorrect the deformity. An alternative technique is to use a small screw as the anchor for the transnasal wires. Interfragmentary wiring is complete and, if unstable, further supported by plate fixation of the medial orbital rim.

Another detrimental aesthetic feature of nasoethmoid fractures is the loss of nasal dorsal support secondary to combined fractures of the septum, ascending nasal process of the maxilla, and the nasofrontal suture. Under such circumstances bone grafts should be used to correct the deformity. These must be rigidly fixed to prevent reabsorption and this is best accomplished by lag screw fixation of the graft to the remaining nasal dorsum or by plate fixation of the graft to the frontal process in a cantilever fashion.

**Orbital Floor Fractures**

Orbital floor fractures are the most common injuries of the middle component of the orbit. They occur as isolated "blow-out" fractures or in conjunction with zygomaticomalar or Le Fort-type fractures. In children the occurrence of these injuries parallels the pneumatization of the maxillary sinus and are generally not seen before 5 years of age.

The clinical findings suggestive of an isolated fracture are diplopia, infraorbital hypoesthesia, periorbital ecchymoses, and edema. Several "blow-out" configurations occur. In a "trapdoor" fracture, the "blow-out" is hinged on one surface with the orbital contents herniated past it and trapped by the fragment of bone. In a "saucer" fracture, there is a depressed fracture of the floor with an increase in orbital volume resulting in enophthalmos.

The surgical treatment of isolated "blow-out" fractures has been the subject of controversy. Putterman suggests observation alone, whereas Converse and Smith advocate early exploration and reconstruction. Although these opposing perspectives remain unresolved, a generally accepted approach in children is to observe them for 1 week. If at that point they continue to have enophthalmos, restriction or pain on movement, and ptosis of the globe on upward gaze, then exploration is undertaken. Large fractures are routinely explored as are fractures that on CT have muscle entrapment. Orbital floor fractures concurrent with other maxillary fractures are reconstructed as part of the overall facial fracture repair.

The surgical approach to isolated orbital fractures is via a transconjunctival or subciliary incision. The orbital septum is exposed and followed to the rim where the periosteum is incised, the entire orbital floor exposed, and the fracture site identified. Herniated orbital tissue is carefully teased back into the orbit and the bony fragments are elevated into position if possible and occasionally removed. If a defect or weakness persists, an implant of two layers of saline-soaked Gelfilm is usually adequate in children. In more severe multiple fractures with large defects, calvarial bone implants secured to the rim by 28-gauge wire may be necessary.
**Orbital Roof Fractures**

Orbital roof fractures in young children have traditionally been considered a rare injury. However, several recent reports suggest that these are more common than previously suspected. They all attribute this to the availability of direct, coronal CT evaluation in conjunction with an increased awareness of the clinical presentation of these injuries.

Characteristically, isolated orbital roof fractures occur in children under 7 years of age prior to the pneumatization of the frontal sinus. There is usually a history of a blow to the brow from a fall or a blunt object often associated with a late-developing periorbital hematoma. This delayed swelling can be an important clue in differentiating roof fractures from other orbital injuries. Proptosis or dystopia can occur but may not be immediately evident. Concomitant intracranial injury is present in a substantial proportion of cases. Although permanent morbidity is an ever-present concern, usually as a consequence of neurologic damage, the orbit and the globe do not frequently sustain long-term damage. Orbital encephaloceles have been reported as a late sequela, presenting with vertical dystopia, axial proptosis, and pulsation of the globe.

Pediatric orbital roof fractures also occur as a component of more extensive craniofacial fractures. These tend to be seen in older children and are associated with greater impacts such as high falls and motor vehicle accidents. These children have a much higher incidence of acute neurologic injury as well.

Messinger et al have suggested a classification system for orbital roof fractures based on the pattern of injuries seen on coronal CT. In type 1 fracture, there is comminution of the orbital roof but no displacement of fragments. In type 2, the fracture fragments are superiorly displaced toward the anterior cranial fossa. In type 3, the fracture fragments are inferiorly displaced into the orbit. Type 1 and type 2 injuries do not need surgical repair; however, large type 3 fractures will require combined intracranial and extracranial exploration with cranial bone graft reconstruction of the deficit to correct dystopia and exophthalmos and to prevent encephaloceles. For more extensive craniofacial injuries, where there is orbital roof fracture, neurosurgical intervention takes priority. However, surgical management may in fact require concurrent reconstruction.

**Medial Orbital Wall Fractures**

Medial orbital wall fractures in children are typically associated with nasoethmoid fractures. Isolated fractures of the lamina papyracea are relatively uncommon, occurring usually as a consequence of blunt trauma to the nose, rim, or eye. Orbital emphysema is commonly seen on CT. Enophthalmos and entrapment may occur with isolated injuries and, under these circumstances, surgical correction is necessary. This can be accomplished by an external ethmoidectomy-type incision, which exposes the medial orbital wall. Soft tissue reduction and bone grafting, if necessary, can then be performed.

**Orbital Apex Fractures**

Fortunately, orbital apex fractures are rare injuries. Although there are no reports in the literature of these injuries specific to children, it is doubtful whether the mechanism,
pattern, and consequence of trauma to this vital area varies with age. Fractures of the apex are usually due to posterior extensions of complex craniofacial injuries. Blindness is the greatest concern, occurring as a result of optic nerve injury and vascular injury to the ophthalmic artery. Treatment with steroids or optic nerve decompression via either a transsphenoidal or intracranial approach should be considered; however, reversal of blindness is unusual. Injury to the neurovascular structures entering the superior orbital fissure can result in ophthalmoplegia, ptosis, and fifth nerve hypoesthesia. Treatment is expectant but long-term deficits are common.

Temporal Bone Fractures

Temporal bone fractures are not traditionally considered within the realm of maxillofacial trauma. Because of frequent association between these two types of injuries in children, as well as the profound aesthetic and functional facial changes that can be the result of temporal bone fractures, a brief review of the topic is presented.

Temporal bone fractures are classified by their anatomic pattern of injuries into three types: longitudinal, transverse, and mixed. Longitudinal fractures are the most common and account for 70% to 85% of temporal bone fractures. The mechanism of injury is generally trauma to the temporal or parietal areas. These injuries are parallel to the long axis of the petrous bone, typically starting at the posterior half of the squamosa, extending medially, and slightly anteriorly. Along its course, the fracture runs through the posterosuperior wall of the external auditory canal, the epitympanum, and the carotid canal, ending in the middle cranial fossa near the foramen lacerum. Classically, the fracture is lateral and then anterior to the inner ear. Although the cochlea and vestibule are not directly injured, sensorineural hearing loss is reported in 5% to 35% of longitudinal fractures. Conductive hearing loss is much more common, occurring in about 50% of cases as a result of hemotympanum, tympanic membrane tears, and ossicular chain disruptions. Hemorrhage from the external auditory canal is also a common feature resulting from tears of the canal skin. Facial nerve dysfunction occurs in about 10% to 20% of longitudinal fractures, generally as the consequence of a physiologic block secondary to stretching, ischemia, or edema in the tympanic segment of the nerve. CSF otorrhea is rare in longitudinal fractures and is due to medial extension of the fracture to the tegmen. Bilateral longitudinal fractures are seen in 10% to 30% of cases.

Transverse fractures account for 10% to 20% of temporal bone fractures. The mechanism of injury is usually from severe blows to the occipital or frontal areas of the skull. These injuries are perpendicular to the long axis of the petrous bone, typically starting at the foramen magnum, crossing the petrous pyramid, and ending in the middle cranial fossa at or near the foramen lacerum. As the fracture passes across the petrous pyramid it involves the cochlea and vestibule, resulting in universal sensorineural hearing loss and vertigo. Facial nerve injury occurs in 50% of cases, generally due to a tear within the labyrinthine segment. If the fracture line involves the medial wall of the middle ear then hematotympanum is seen. Middle ear involvement can also result in CSF otorrhea.

Mixed fractures account for 10% to 20% of temporal bone fractures. The mechanism of injury is usually from crushing blows to the side of the head. There is no generic fracture pattern as in longitudinal and transverse types, but a variety of fracture lines involving the petrous pyramid, cochlea, vestibule, and middle ear are seen. There is thus a variable clinical
picture of conductive and sensorineural hearing loss, vertigo, facial nerve paralysis, hematotympanum, canal hemorrhage, and CSF otorrhea.

In children, these types of injuries are due to pedestrian-motor vehicle accidents (40%), falls (30%), passenger-motor vehicle accidents (25%), and blows to the head (5%). Hence, the temporal bone fracture is only considered after the more life-threatening injuries have been taken care of. Important diagnostic studies include axial and coronal CT and early audiologic evaluation. Facial nerve testing is indicated when paralysis is present.

Management of temporal bone fractures depends on the deficits incurred. The most common problem is conductive hearing loss. When this is due to hematotympanum alone it generally resolves without intervention. In most cases, tympanic membrane tears will also heal on their own. This can be facilitated with paper patching, if the child allows, or concurrently with a general anaesthetic for another problem. Persistent perforations have a potential for cholesteatoma formation and should be repaired with standard tympanoplasty techniques within 3 to 6 months of the injury.

Ossicular disruptions account for 15% to 20% of conductive hearing loss in temporal bone fractures. The two most common injuries are incudostapedial separation and incus dislocation. Fractures of the stapedial crura occur less frequently. An air bone gap of 35 dB or greater 3 months after the injury suggests ossicular disruption and is an indication for exploratory tympanotomy. Incudostapedial separation or incus dislocation warrants ossicular reconstruction with the homograft incus. This can be accomplished either as a "mushroom cap" interposition between the tympanic membrane and the stapes or as a Y strut between the malleus and stapes. Crural fracture is best treated with stapedectomy in older children. In younger children, the footplate should be left intact.

Sensorineural hearing loss can occur with all types of temporal bone trauma and is seen in 5% to 10% of children with head injury. Recovery rates are reported between 10% and 50%. With longitudinal injuries, where there is no fracture of the cochlea, sensorineural hearing loss may be a consequence of hydraulic trauma to the inner ear membranes or to disruption of central auditory pathways. With transverse fracture there is direct cochlear injury resulting in a 90% to 100% incidence of sensorineural hearing loss. When there is anakusis or flat hearing loss, little recovery can be expected; however, there may be some with fluctuating or low-frequency losses. If there is serviceable hearing in the involved ear, amplification can be provided. Suspicion of perilymphatic fistulae, as evidenced by fluctuating sensorineural hearing loss and positive fistula tests, indicates immediate exploratory tympanotomy.

Vertigo can also occur with all types of temporal bone injuries. It is more likely with transverse and mixed fractures, where there is direct vestibular injury. Both post-concussive vertigo and cupulolithiasis can be seen with longitudinal fractures. Vestibular testing is difficult in children and may not add much to the outcome. Due to the plasticity of a child's brain, dizziness and disequilibrium will extinguish as compensation occurs.

Facial nerve paralysis is more likely with transverse and mixed fractures. Longitudinal injuries are much more common; hence, they account for a higher incidence of facial paralysis as a consequence of all temporal bone trauma. With longitudinal fractures, facial paralysis
will often have a delayed onset. This is an important clue since it suggests subsequent recovery. Unfortunately, this information is not always available and prognostic insight must await facial nerve testing. Electroneurography is the diagnostic procedure of choice and can be done as soon as 3 days after the injury, the time required for denervation to occur. Fisch has published guidelines regarding the prognostic value of electroneurography along with indications for surgical intervention. If there is 90% or greater denervation within 6 days of the injury, the likelihood of recovery is small and facial nerve exploration and decompression is indicated. Denervation of a lesser degree, occurring over a longer period of time, is associated with a greater likelihood of spontaneous recovery and surgical intervention is not necessary. The surgical approaches for facial nerve exposure are via the transmastoid, translabyrinthine, and middle cranial fossa routes. Each approach has its advocates, but all agree that decompression of the geniculate ganglion is desirable for optimal results. For cases with loss of nerve tissue, cable grafting or hypoglossal facial anastomosis can be utilized.

Cerebrospinal fluid leaks presenting as clear otorrhea or rhinorrhea can be the one life-threatening consequence of temporal bone fractures because of the potential for meningitis. They occur in 5% of basilar skull fractures and will spontaneously seal more than 90% of the time. In the first 2 weeks, treatment is directed toward promoting natural closure by bed rest, head elevation, and spinal drainage. If the leak persists, an attempt should be made to diagnose its source with metrizamide-contrasted CT or radionuclide studies. Unfortunately, these are not universally successful in identifying the site of leakage. Surgical closure by either a transmastoid or craniotomy approach is necessary for persistent leaks, late-onset leaks, recurrent meningitis, or brain herniation.

Conclusion

In our highly mechanized society children sustain serious maxillofacial injuries that require appropriate repair. The primary factor that differentiates the treatment of pediatric facial fractures from those of adults is facial growth. The anticipation of growth of the mandible simplifies fracture repair, since most of these injuries can be managed with intermaxillary fixation. On the other hand, inadequate treatment of upper facial injuries will result in serious alterations of facial growth. The techniques of three-dimensional reconstruction of complex fractures have been revolutionized over the past decade with the use of rigid plating systems, craniofacial exposure, and bone grafting. There are solid theoretical grounds for the application of these techniques and short-term results of their use appear promising. Only time and careful study will prove their long-term value.
Pediatric Facial Plastic and Reconstructive Surgery
James D Smith, Robert M Bumsted

Chapter 20: Wound Healing and Scar Revisions in the Pediatric Patient
Charles W Koopmann

This chapter discusses the basis of wound healing in respect to not only the phases of response to injury but also the factors affecting the healing of wounds, including the effect of vitamins, age, foreign bodies such as sutures, anemia, and drugs. The chapter then describes abnormal wound healing including hypertrophic scars and keloids. Special problems such as electrical and thermal burns to the lip and neck, and principles of scar revision will complete the chapter. It is the author's strong feeling that meticulous attention paid to the management of the physiologic principles of wound healing is essential for optimal cosmetic and functional results.

Phases of Wound Healing

There are three (some authors say four) phases of wound healing: the exudative or inflammatory phase (sometimes subdivided into hemostatic and inflammatory phases); the proliferative phase; and wound contraction or the remodeling phase.

Inflammatory or Exudative Phase

The inflammatory or exudative phase begins immediately and can extend for several days. In the early phase, the disruption of blood vessels causes the wound to fill with blood, serum proteins, clotting factors, and platelets. Collagen is involved in the reparative process from the initiation of the wound throughout the end stages of remodeling. It is in the initial phase of the inflammatory (or hemostatic) portion of wound healing that there is interaction between collagen and platelets. The activated platelets not only initiate coagulation but will also release biologically active substances such as growth factors, fibrinogen, von Willebrand's factor, fibronectin, and other substances that promote cell migration into the wound. The fibrin clot itself not only assists in hemostasis but also acts as a scaffold for later fibroblast migration.

Within the first 5 to 6 hr postinjury, neutrophils enter the wound as a result of the release of chemotactic factors. This is considered by some to be the latter portion of the inflammatory exudative phase or, by others, the beginning of the second phase (inflammatory) of wound healing. Monocytes will begin to accumulate within 24 to 48 hr. While the neutrophils are present to fight contamination from bacteria, the monocytes (macrophages) not only phagocytose bacteria, but will digest the neutrophils and bacteria and release biologically reactive substances that will assist in the tissue debridement and release of growth factors (platelet-derived growth factor (PDGF)) and transforming growth factors, type beta (TGF-beta), which is necessary for the initiation of the granulation tissue.

The biological consequences of the phagocytic portion of this phase will result in the increased oxygen consumption, phospholipid metabolism, glucose utilization, and production
of hydrogen peroxide. Polymorphonuclear (PMN) dysfunction can occur in patients with thermal injuries. The PMN will serve the role of generating electrically excitable oxygen molecules that will assist in proline hydroxylation. This helps to explain that one way to accelerate wound healing is by producing an inflammatory response.

Macrophages produce a growth factor that stimulates fibroblasts, fibronectin, and angiogenesis or neovascularization. Fibronectin will become an insoluble matrix that will cross-link with collagen and other cells or cell surfaces (platelet aggregation to injured vessel walls). Later this same fibronectin will cause clot retraction. Thus, although PMNs are present for a relatively short time, the monocytes remain longer and play a major role in the transaction between the inflammatory and granulation tissue phases.

**Proliferative or Granulation Phase**

The second phase of wound healing occurs approximately 3 to 4 days after injury. It is characterized by a rapid increase in fibroblast numbers and in epithelial cell mitoses, as well as an increase in the synthesis of extracellular collagen and proteoglycans. Reepithelialization of the wound will begin within hours of the injury, but it increases during this rapid mitosis as cells migrate along the new fibrin bridge. The epithelial migration will continue until cells touch one another, causing contact inhibition and signaling the end of epithelial expansion and the beginning of keratinization. Most sutured wounds will have epithelial coverage within 4 days after the insult.

Following the fibroblasts is a proliferation and ingrowth of capillaries, Angiogenesis is very complex, and involves chemotactic-stimulated migration, which causes the wound to become filled with granulation tissue (fibroblasts, macrophages, and new capillaries). The angioneogenesis will begin approximately 48 hr after injury and the ingrowing fibroblasts will produce collagen, elastin, and proteoglycans within 3 to 4 days. The collagen synthesis itself is extremely active 5 to 7 days after injury. The interdigitation of collagen in the midportion of the wound with collagen along the wound edges forms a source of intrinsic wound strength.

**Wound Contraction (Matrix Formation) and Remodeling Phase**

The third phase of wound healing (wound contraction or matrix formation and remodeling) is best characterized by a reduction in the numbers of fibroblasts, macrophages, and wound vascularity. Fibronectin is eliminated from the wound matrix, while at the same time type I collagen will accumulate. Later, type III collagen will form with fibronectin. The contraction component in this phase is due to myofibroblasts, causing the wound to contract by 0.6 to 0.75 mm per day. This occurs when random collagen fibers are replaced by parallel cross-link fibers, yielding most commonly a flatter, softer wound with increased tensile strength. The capillary reduction occurs within 6 to 18 weeks and the remodeling process is completed between 6 to 18 months.

During the remodeling phase, the scar size is a function of wound tension, pressure, age of the patient, and oxygen supply to the area. Mechanical stress will promote collagen synthesis and deposition, often yielding a hypertrophic scar. Hypoxia will also stimulate collagen formation and deposition. As the scar matures, it will usually become more dense
as a result of loss of fluid and volume (corresponding to wound contraction). For a summary of cellular activity in wound healing, see Table 1.

**Table 1. Summary of cellular activity in wound healing**

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<th>Platelet</th>
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<td>Aggregation</td>
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<td>Coagulation</td>
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<td>Release of active substances</td>
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<td>Chemotactic factors</td>
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<td>Vasoactive factors</td>
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<td>Growth factors</td>
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<td>Enzymes (proteases)</td>
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<td>Scavenger functions</td>
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<td>Bacteria and other pathogens</td>
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<td>Tissue debris</td>
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<td>Neutrophils</td>
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<td>Release of active substances</td>
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<td>Vasoactive mediators leading to angioneogenesis</td>
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<td>Chemotactic factors</td>
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<td>Enzymes (proteases)</td>
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<th>Platelet</th>
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<td>Wound contraction</td>
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<td>Migration into wound</td>
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<td>Deposition of fibronectin and collagen</td>
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<td>Contraction</td>
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<td>Formation of wound matrix</td>
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<td>Fibronectin</td>
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<th>Platelet</th>
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<td>Formation of new epidermis</td>
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<td>Migration over wound surface</td>
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<td>Formation of new tissue</td>
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<td>Release of active substances</td>
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<td>Vasoactive factors necessary</td>
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<td>Enzyme (proteases)</td>
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<td>Growth factors.</td>
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Factors Affecting Wound Healing

Local Factors

There are multiple local factors that have a profound influence on the quality of wound healing. One of the first and most important factors is the manner in which the wound was inflicted. Adverse wound healing can be expected to be seen in contused wounds, severely infected wounds, and traumatic tattooing in and about the wound. Perhaps the most important factor is the orientation of the wound in relationship to normal skin tension lines (the "relaxed skin tension lines" (RSTL) of Borges). The RSTLs are generally at right angles to the direction of underlying muscle pull, and are most easily seen in the aging wrinkle lines in the geriatric patient. Wounds crossing the RSTL are difficult to close accurately (without tension) and have a higher tendency to become widened and hypertrophic. Tension along the suture line, especially when associated with motion, tends to lead to widened, hypertrophic scars. Immobilization helps facilitate initial healing. The mechanical stress associated with movement leads to a decrease in the mitotic activity and an impairment in the ingrowth of vessels.

The nature of the injured skin is of extreme importance also. The tight, wrinkleless skin of the pediatric patient will do well in hiding scars from simple surgical incisions or lacerations. However, when the wound crosses the natural tension lines, or is closed under tension, the pediatric patient will be more likely to show scar hypertrophy than the elderly patient with loose, wrinkled skin. Also, the location of the wound is important since wounds of the scalp will contract very little, whereas a wound near the commissure of the mouth will have a tendency to cause lip deformity because scar tissue contracture is a virtually unopposed force.

The tissue type and complexion of the patient is also extremely important since an individual with a darker, olive complexion will be more likely to form pigmented, hypertrophic scars. Surprisingly, the converse can also be true in black patients where deep abrasions and burns may cause scars that will remain unpigmented for very long periods of time. A fair-complexed patient may be more predisposed to widened scars that remain reddened for longer periods of time.

Blood supply and hematoma formation are also extremely important. Ischemia will cause failure of the wound to heal. Fortunately, in the pediatric patient who has scalp, facial, or neck injuries or incisions, the blood supply is usually excellent and is rarely a problem. Hematoma formation, however, is one of the most common wound-related complications, not only causing problems with basic wound healing but also predisposing the patient to a wound infection. Thus, prevention of hematoma formation should be the goal. Where possible the surgeon should attempt to assure adequate coagulation by having the patient avoid medication that tends to interfere with clotting (anti-inflammatory drugs, salicylates, etc), and to correct bleeding disorders (in hemophiliac patients or patients with decreased platelet or fibrinogen deficiencies). If there is significant oozing at the time of surgery, and ligation and/or electrocautery is inadequate, the use of suction drains (either the standard Jackson-Pratt or, in smaller wounds, the adaptation of a scalp vein IV infusion set to make a small suction drain) is advised. One can further assist coagulation by the use of compression dressings if the wound's supply will tolerate the external pressure. If these methods fail, the surgeon may
be well advised to pack the wound until hemostasis is secure and return after 48 to 72 hr to perform a delayed primary suturing.

Wound infection is one of the most common problems associated with suboptimal wound healing. The best treatment for infection is prevention. Factors influencing the incidence of infection include impaired local blood supply, hematoma formation, foreign bodies, patient status (nutrition, electrolytes, metabolism (diabetes), anemia, and drug ingestion - steroids or antimetabolyte chemotherapeutic agents), and surgical technique. It is interesting to note that in the early stages both infection and wound healing are very similar in their cellular and vascular responses to insult. It is well known that a delay in wound healing occurs when wounds are inoculated with bacteria. Also, it has been shown that there is a decreased tensile strength in wounds contaminated with *Staphylococcus*. The principal biochemical abnormalities in infected wounds appear to be a disturbance in collagen metabolism, probably via lysosomal enzymes present in PMN cells, and a depressed effect on fibroblasts leading to a disturbance in collagen synthesis. In a wound that is significantly contaminated, the most efficient method for reducing the risk of infection is to dissect and remove devitalized tissue without attempting to remove an entire wound surface. When this is impractical, one can attempt to dilute the bacteria by irrigation with a physiologic saline solution, employing the physiologic solution in combination with some type of antibiotic (bacitracin is most commonly used), or using a surface antibacterial agent (povidone-iodine (Betadine) is most commonly used in this fashion). Once a wound becomes infected, closure is not advised since that may cause the formation of an abscess. Instead, it is best to leave the wound open until signs of inflammation are gone, and then consider secondary wound closure.

Surgical Technique

Surgical technique is very important in the creation of an adequate wound. Borges and Alexander recommend that the orientation of the wound in relation to normal skin lines of tension (RTSL) is possibly the single most important factor in scar prognosis. The lines of tension should be at right angles to the direction of the underlying muscle pull. Any wound that crosses the lines of skin tension will have a tendency to widen and hypertrophy. The width of most scars is proportional to the magnitude of skin tensions. Thus, not only the planning of incisions is essential, but also the conversion of suboptimal wounds using rotational flaps or Z- or W-plasties is important. Also, the reduction of skin tension by the use of undermining and approximation with multiple layers of wound closure is essential. The concept of having wound edges in apposition prior to the final subcuticular or skin suture layer is very important. In the face, the dynamic skin tensions are perpendicular to natural skin wrinkles and parallel to the direction of contraction of the underlying mimetic muscles. Thus, a linear scar intersecting the wrinkle lines can result in severe scar tissue contracture. When the directional orientation of a wound tends to predispose to a widened, unattractive, or hypertrophic scar, it is very important that the physician warn the patient and the family of the probability of unacceptable scar formation and the likely need for scar modification later. At a later date (usually 12 months or later), the scar could be revised and placed in more optimal skin tension geometric patterns with either a W-plasty or a Z-plasty, making the orientation of a portion of the wound perpendicular to dynamic skin tensions.
Wound closure techniques depend on the type of wound. Basically, two types of wound exist: (a) one characterized by loss of tissue; and (b) one with no evidence of loss of tissue. Usually primary closure can be accomplished in the wounds without loss of tissue, whereas wounds with significant tissue loss require flaps or grafts to close the defect.

Timing of wound closure is also important. The main decision is whether to close the wound primarily, or whether to delay closure. In the head and neck, time is not the major criterion. If the wound appears to be in an area with minimal devitalized tissue, and without evidence of significant inflammation, usually closure can be accomplished even several hours after the initial injury. If, however, there is severe devitalization of tissue, or if there is evidence of wound infection, open wound management prior to delayed primary closure is reasonable. This can be accomplished by the use of fine mesh gauze covered by a sterile dressing.

The method of wound closure is important. Most commonly, sutures are inserted to close both deep and superficial layers. Dermal sutures are utilized to reduce skin tension. These dermal sutures serve as a precaution against wound dehiscence. Most commonly, minimally reactive absorbable sutures such as polyglycolic acid or polyglactin sutures are used. Not only will these sutures reduce skin tension, but also they will tend to obliterate potential dead space and reduce hematoma formation. The skin is then closed with 5-0 or 6-0 suture using the technique of approximating the midportion of the wound and then, with further sutures, bisecting the residual portions of the unclosed wound. For skin sutures, one might utilize a 5-0 or 6-0 nylon-propylene suture on a swage cutting needle. In wounds that are stellate or jagged-edged, percutaneous suture closure of the skin edges gives much better results than taping.

General Factors

Age

Wounds in children tend to heal more rapidly and actively than those in the older patients. This is frequently due to increased blood supply, as well as less systemic disease. However, there does appear to be a slight increase in the tendency for reddened or hypertrophic scars in children. Aging affects all stages of wound healing. In open wounds in the elderly patient, the time before contraction begins is lengthened, the healing rate is slower, and the final degrewe of epithelialization is late. In the elderly, the breaking strength of incisional wounds is lower and the bursting force is less. The rate of wound healing varies inversely with age. Collagen and elastin are markedly altered in the elderly with soluble collagen decreasing, whereas insoluble collagen increases with age. Elastin also increases in the elderly, but the quantity of elastin fibers decreases, causing atherosclerosis and poor wound vascularization.

Nutrition

Malnutrition plays a major role in wound healing complications. Proteins are necessary for collagen synthesis, fibroblast proliferation, revascularization, and the formation of lymph channels. Severe protein deficiency will inhibit all phases of wound healing, including humoral and cell-mediated antibody responses, and phagocytic action, and it will delay fibrous
tissue matrix formation. Hypoalbuminemia will cause edema and interfere with microvascularization. Vitamin deficiencies will also affect wound healing. Specifically, vitamin A enhances epithelial rate of collagen synthesis, collagen cross-linking, and synthesis of glycoprotein and proteoglycans, as well as collagenase production. Viral and bacterial infections are enhanced by vitamin A deficiency, and vitamin A supplements reverse the adverse effects of glucocorticoids.

Vitamin B serves as a cofactor for enzyme reactions, and is required for a white cell function and antibody formation. Clinical vitamin B deficiency states do not appear to delay wound healing. Vitamin C is required for hydroxylation of lysine and proline and collagen synthesis, capillary formation and strength, macrophage migration, neutrophil function, and inflammatory response, as well as immune reactions. Since humans do not store vitamin C, deficiency may occur rapidly in chronically ill patients. Vitamin D is essential for new bone formation and for transport and metabolism of calcium, but there is no evidence that a deficiency state reduces bone healing in human surgical subjects. Vitamin E has anti-inflammatory properties because it reduces functioning free radicals. It also may improve cell-mediated immunity and inhibit platelet aggregation. Vitamin K is essential for the synthesis of clotting factors VII, IX, X, and prothrombin. Deficiencies of vitamin K lead to increased risk in bleeding diatheses and hematoma formation. Finally, zinc deficiency can result in retardation and delay in the repair process as a result of impairment of synthesis of nucleic acid and protein (especially collagen). It is also essential in cell membrane function and for vitamin A transport. although there is no therapeutic value in the administration of zinc in the nondepleted state, zinc-deficient patients may be aided by an intake of 220 mg of zinc sulfate three times a day. Zinc excess (greater than 40 mg/dL) can impair wound healing.

**Anemia**

Anemia, especially in hypovolemic states, will cause tissue hypoxia and result in poor wound healing. Anemia is also associated with an increased infection rate, although it is usually difficult to separate anemia from problems leading to poor nutritional states.

**Diabetes Mellitus**

Patients with diabetes mellitus have wound complication rates of up to 10% incidence. This is usually related to infection, although additional factors include neuropathy and atherosclerosis. Improved management of diabetes mellitus has reduced the risk to the patient. Specifically, preoperative modification of insulin requirements is important. Diabetics also appear to have white cell defects, which contribute to their propensity for postoperative wound infections. However, the greatest improvement in diabetic wound management is related to proper control of blood sugar in the perioperative period. Thus, it is important to make certain that the diabetes is well controlled with the appropriate endocrinology consultation.

**Medications**

Glucocorticoids (corticosteroids) exhibit significant inhibitory effects on wound healing. Low doses (less than 10 mg prednisone per day) have minimal, if any, effect in adult patients, but repeated high dosages (40 mg or more prednisone per day) will adversely affect...
wound healing, including (a) reduction of fibroblastic proliferation and granulation tissue; (b) polymorphonuclear leukocytes with reduced motility, phagocytosis, and adhesiveness; (c) reduction of lymphocytes in tissues and blood; (d) macrophages with reduced mitosis and phagocytosis; (e) vasoconstriction resulting in reduction of blood and nutritional supply and nutrient supply; and (f) reduction of synthesis of collagen, proteoglycan, and glycosaminoglycan.

Anticoagulants such as sodium warfarin (Coumadin) and heparin increase the risk of hematoma formation, bacterial infection, and wound dehiscence. Nonsteroidal anti-inflammatory agents (aspirin, phenylbutazone) will decrease tensile strength of wounds, and increase the risk of bacterial infection and hematoma formation.

Although antineoplastic agents have been implicated in delayed wound healing, the results are conflicting. It has been suggested that actinomycin D or bleomycin in the perisurgical period may cause some impairment of wound closure. However, it has also been surmised that although limited clinical data do not confirm significantly diminished wound healing when these agents are used, most surgeons prefer withholding chemotherapeutic agents in the first 7 to 10 postoperative days.

Colchicine, an anti-inflammatory agent, interferes with fibroblasts and their secretion of collagen precursors, as well as stimulates collagenase and reduces the secretion of histamine granules from mass cells. It was used by Peacock in combination with penicillamine or beta-aminopropionitrile in the treatment of keloids and hypertrophic scars. Penicillamine can reduce the tensile strength of healing wounds by affecting collagen cross-linking, and can also be used to modify hypertrophic scars. Large doses of penicillin would theoretically yield enough penicillamine to cause wound healing problems, but in fact, this has not been shown to be clinically significant. Beta-aminopropionitrile is a lathyrogenic agent that yields reduced tensile strength and reduces collagen fiber formation. It has also been utilized to modify adhesions and keloid formation.

**Jaundice, Uremia**

Theoretically, jaundice can affect wound healing in the realm of a poor absorption of vitamin K, and thus decreased clotting factors yielding increased wound hematoma. Hepatic dysfunction will also cause problems with protein metabolism. However, the role of jaundice in predisposing to wound healing complications is multifactorial in that these patients are also usually malnourished and have protein abnormalities. Thus, one must consider not only the effect of the hepatic dysfunction on clotting mechanisms but also the patient's dietary intake. Uremia will also show evidence of reduction in wound strength with a higher incidence of wound dehiscence. Uremia will cause inhibition of fibroblast ingrowth. Again, the poor nutritional status of the uremic patient may be of primary importance.

Thus, as this portion of the chapter illustrates, optimal wound healing is dependent upon multiple factors. The surgeon must be moderately cognizant of the effect of the above elements so that he can optimally prepare his patient for surgery.
Hypertrophic Scars and Keloids

When one is dealing with either hypertrophic scars or keloid formation, one must realize that there may be some intrinsic abnormality in the patient's physiologic reaction to tissue insult that will cause difficulties in attaining optimal satisfactory results. In reviewing the literature in preparation for this chapter, the author finds the words "hypertrophic scar" mentioned in conjunction with keloid formation, but rarely does he find a definition of the term. Most commonly, it is utilized by various authors to mean noticeable scar tissue that may be raised, reddened, or widened, but it is not the exuberant deposition of huge, raised amounts of collagen in healed skin wounds. Peacock et al have clinically differentiated between the two entities by defining a keloid as an excessive collection of scar tissue that has no resemblance to the shape of the original wound and extends beyonds the confines of the wound, whereas a hypertrophic scar is excessively large but retains the original shape and remains within the confines of the original wound.

In both conditions, there appear to be abnormalities in both the synthesis and degradation of collagen. Specifically, Cohen and Diegelmann have shown that collagen synthesis in both conditions is greater than in normal skin, and that in the keloid collagen, synthesis is greater than that in hypertrophic scars. They hypothesized that hypertrophic scars are merely raised because the rate of collagen synthesis was greater than the usual scar tissue. It is not clear whether the increased collagen synthesis in keloids is due to an increased number of collagen-synthesizing fibroblasts, a normal number of fibroblasts stimulated by some factor to make more collagen, or fibroblasts growing at a rapid rate.

The tendency to form hypertrophic scars is related to several significant factors. One factor is the tension placed upon the scar tissue. Wounds that tend to have more tension will be at a higher risk to form excess scar tissue. This tension may be in the form of crossing the RSTL of Borges in regions of tension near joints or in circumferential scars. At times it may be necessary to splint the area or immobilize the region (if possible).

The age of the patient is also important since fibroblastic activity in patients under age 30 is much greater than in patients 60 years of age and over. Thus, the chance for a bulkier scar is greater in the younger patient, although the remodeling process may cause the scar to flatten nicely, and in the early stages it may show evidence of hypertrophy.

Crushed tissue, large areas of dead space, and infection are also stimuli for scar hypertrophy. Finally, noticeable skin suture marks may occur when the sutures are left in place for excessive periods of time. Specifically, in the presence of percutaneous sutures, epithelial cells will tend to migrate downward, following the suture. If the sutures are removed before the 8th postoperative day, this epithelial invasion will regress, leaving minimal deformity. However, after 8th day, the suture track reaction to the foreign body becomes extremely intense and a permanent deformity (permanent needle scars) may result.

Race may also play an important role in healing, especially in patients with keloid formation. Large hypertrophic scars can occur in fair-skinned individuals, but they are more commonly seen in darker-skinned persons. Keloids are most commonly seen in black-skinned individuals.
The treatment of hypertrophic scars and keloid formation is obviously one of prophylaxis. In patients with a history of increased scar formation, one should make every attempt to reduce the tension on the wound, place incisions within the appropriate skin creases or areas of reduced tension (or reduce the tension by primary W- or Z-plasties or rotation flaps), and remove skin sutures before the 8th postoperative day. However, in spite of the above treatments, one still may encounter hypertrophic scars or keloid formation. Intralesional injections of triamcinolone are useful in modifying the wound healing. One can inject triamcinolone (10 mg per cc) utilizing a 25- to 27-gauge needle. If hypertrophic scar formation (or keloid) is suspected, the injections can begin within the first 3 postoperative weeks. Further 3-week courses may be given if there is itching in the scar, or if the scar hypertrophy appears to be unresolved. In patients who do not have a history of hypertrophic scar formation, one can initiate injection at the first sign of the problem. If the result of injections is unsatisfactory, then one could proceed with scar revision and immediate injection of steroids, or if the lesion is in a readily accessible and optimal site, pressure dressings can be applied. The pressure appears to reorient collagen bundles parallel to the surface of the skin and will cause a reduction in the level of chondroitin sulfate associated with scar hypertrophy. The disadvantage of this treatment is that it must be maintained continuously for at least 9 to 12 months, since premature release of the pressure will cause recurrence of the lesion. Also pressure dressings are not effective in lesions that have been present for 6 to 12 months or longer.

Radiation therapy has been a controversial modality utilized for the treatment or prevention of keloids and hypertrophic scars. It has no effect on established lesions, but it may be of benefit in the prevention of lesions in the immediate postinjury or revision case. Doses of 1,000 rad, given singly or in divided doses can suppress proliferation of fibroblasts. One recommendation is giving three to four daily treatments of 300 to 400 rad to each surface with a 100-kV machine. This would be a very unusual course of therapy in the pediatric patient, especially over active bone growth sites or in the region of endocrine glands. For the keloid or hypertrophic scar that has been present for 6 months or longer, the treatment will usually depend upon the size and location. As we have mentioned, radiation therapy and pressure dressings will not be successful in the management of these lesions. Intralesional steroid injections may be of some benefit in the long-standing case. Again, the injection can be performed using 10 mg per cc triamcinolone injections at 3- to 4-week intervals. If this is unsuccessful, then the keloid could be excised and the area closed primarily, if possible. If primary closure is possible, it would be recommended that one perform a subtotal excision. Specifically, the bulk of the central keloid would be excised with a very minimal keloid frame left on the sides of the wound to act as a splint to prevent skin retraction. The central portion of the keloid should be removed as deeply as possible. Then the wound could be either closed primarily or, if this is impossible, then a skin graft placed to cover the bed. In the postauricular area, one should excise the keloid completely and cover the defect with a full-thickness graft from the groin. If the patient has extensive resurfacing needs, it is recommended that one test a small area to see if the patient will heal appropriately, rather than try to graft the entire area at once. All grafts should be extremely thin (10/1.000 inch) split-thickness skin grafts to avoid hypertrophy at the donor site.

The complications of the therapy for hypertrophic scars and keloid formation can include local effects of the steroid injections, such as atrophy of the skin with telangiectasia. Also, complications include recurrence of the hypertrophic scar and keloid.
Burns involving the head and neck region in children involve specific special problems. Often these burns involve the ignition of clothing. The mortality rate for victims whose clothing is ignited is at least four times higher than those whose clothing is not ignited; the severity and extent of injuries is almost 100% greater in the former than latter group; the percentage of full-thickness injury is six times greater, and the number of days of hospitalization is approximately 60% greater. The injuries are most likely due to open space heaters, kitchen ranges and matches, flammable liquids such as gasoline or pain thinners, or hot water.

Electric burns in infants and young children will most commonly occur when the child puts an electric plug or cord in the mouth. Thus, the lips, oral commissure, and tongue are the most common areas involved in head and neck electric burns. Electrical burns can be of two natures: arc and contact. The arc type of burns will involve temperatures of 2,500°C to 3,000°C, causing charring of soft tissue and bone. These are the types most commonly seen when there is contact with live electrical sockets. Physical examination will most commonly reveal a third-degree burn with a centrally depressed crater, and a slight, pale elevation of the surrounding tissue. The surrounding blood vessels are markedly injured, as are the sensory nerves. Immediately after the accident, one finds that the area of necrosis is not usually apparent, and since it may take several days to demarcate, immediate surgical debridement or reconstruction is usually contraindicated.

For electrical burns, special acrylic or Silastic appliances are usually not immediately placed in the wound. Early management includes the use of antibiotic ointments in an attempt to prevent severe infection. After the extent of tissue destruction can be determined, reconstructive procedures will involve excision of the involved tissue. If the burn is in an anatomically favorable position, immediate reconstruction after debridement is possible. However, when the burn involves a corner of the mouth, it is usually preferable to await spontaneous separation of the eschar and to allow some softening of the wound before definitive repair. Early excision can also risk the sacrifice of uninvolved tissue. In these instances, special acrylic or Silastic appliances placed at the commissure to prevent severe adhesions may be useful. Final reconstruction can be achieved after a significant period of time has elapsed and the scars are allowed to soften (most frequently 1 year is a reasonable time frame).

In the reconstruction of the patient with a thermal burn to the face, the surgeon must be cognizant of certain wound healing problems following the injury. Specifically, wound contraction will frequently be present in both second- and third-degree burns. Four areas in the face are markedly susceptible to contracture due to mobility and flexibility. These are the eyelids, the cheeks, the lips and oral tissues, and the cervical region (neck). The upper lip is pulled superiorly toward the columna via intrinsic contracture forces, whereas the lateral portions of the upper lip are pulled towards the cheeks. The lower lip and chin are pulled downward by both intrinsic scar tissue contracture and also the muscle contractions and scar contracture seen in the cervical region. The oral cavity may have circumferential contraction (purse-string manner) limiting the patient's ability to open the mouth. Similar circumferential contracture can occur in the nasal vestibule region. An interesting observation is that the
position of comfort in the burn wound is one of contracture. Also, the burn wound will contract until an opposing force is met. This opposing force can be splinting and/or exercise. This therapy may be very effective in reducing of contracture in the lip region and the neck.

The timing of repair in the facial burn patient is extremely important. One must keep in mind that allowing the wound to demarcate and show the areas of most contracture is not unreasonable, and therefore a concept of "slow is always bad and fast is not always best" is reasonable. A conservative approach including topical antibiotic therapy, debridement of scar and eschar, and eventually skin grafting is recommended in burns of the face. This conservative approach is advocated because the diagnosis of depth in most facial burns in the early postinjury wound is often misleading. An exception would be partial-thickness burns that can be converted to full-thickness loss from infection during the healing phase. In these patients, excision by shaving the involved tissue, and immediate grafting is warranted, often between the 3rd and 5th postburn days. Usually it is safe to perform the definitive correction of facial burn deformities at approximately 1 year or longer after the injury, when scars and initial grafts have had a chance to mature. In the intervening time between the injury and definitive reconstruction, splints and pressure masks, as well as local steroid injection, may yield improved results with hypertrophic scars. Earlier intervention is indicated in regions around the eyelid where damage to the cornea may be possible, or around the mouth where lower lip eversion causes drooling or severe neck scarring.

In reconstructing the face, one should divide the face in a map as shown. Any flap or graft applied to the face should, if possible, cover the entire aesthetic unit in order to avoid a patchwork appearance. The stages of treatment in these patients include the acute period, which has a pregrafting and skin grafting phase, and a chronic period, which requires a waiting period (1 year or longer, as mentioned above), as well as an early and final reconstructive phase. Since the early or acute periods have been discussed above, we will concentrate here on the oral (lip) and upper neck regions. In the waiting period, one can apply stents, either intranasally or in the oral cavity, or pressure dressings over the entire face or neck. Also during this phase, the patient is encouraged to exercise the periorbital regions and the mouth and neck. The early reconstructive stage mainly involves temporizing measures such as Z-plasties or skin grafts in a nondefinitive fashion.

The final reconstructive phase is not begun until hypertrophic scars have shown signs of becoming less active and scar tissue contracture appears to be stable. The relief of contractures with local flaps and Z-plasties is the first objective of therapy. Secondly, skin grafts are usually applied where possible in preference to skin flaps. Thick split-thickness grafts are much better than thin grafts, since they will be less likely to contract. Burn deformities of the perioral area and chin can be divided into upper lip, lower lip, and neck. In this region, one must again divide the area into aesthetic units as shown. The incisions for this area are shown.

When one encounters ectropion of the upper lip, this is best corrected by full-thickness grafts from the supraclavicular or postauricular regions, or by very thick split-thickness skin grafts if one cannot obtain full-thickness tissue. One should attempt to graft the entire aesthetic unit between the nasolabial folds. When the scar is excised from the upper lip, it is very important to leave some residual scar on the philtrum to avoid a flat upper lip. The contracture in the upper lip should be released by making an incision near the base of the
Reconstruction of the lower lip and chin again follows lines of aesthetic components. When scar tissue is excised from the lower lip and chin, one must remember that a circle of scar should be left on the apex of the chin, since this gives a slightly better profile. Microstomia is a very difficult problem, and may initially be treated by stenting and dental appliances. Deifinitive surgical repair would require one of a multitude of possible flaps, which is beyond the discussion of this chapter. Scar tissue contracture of the neck is usually corrected with multiple Z-plasties to lengthen the web-like vertical bands, and by excising the majoruity of the scar and replacing it with thick split-thickness grafts. Postoperative splints or neck braces are also helpful. When the scar extends over the chin or lower border of mandible, one attempts to excise the area so that a splint can be made with an extension to press against the grafts. In the cervical region, one can attempt to have an interval of 4 to 5 days between scar excision and skin grafting to allow a decent layer of granulation tissue to begin to form. Postoperatively, a splint is applied that will keep the neck extended, mold the chin-neck angle, and apply even pressure over the grafted area. Splint application is usually completed during the 2nd week postgrafting. The use of regional transposition flaps, distal flaps, or microvascular flaps is also occasionally helpful but again is beyond the realm of this chapter.

At this time the author would like to mention that when one finds hypertrophic scarring in burn patients, the surgeon is faced with a decision of an intramarginal or extramarginal excision of the hypertrophic scar. In an evaluation by Engrav et al, it was found that intramarginal excision of lesions seemed to be more efficacious in the younger patient. There are two theoretical reasons for this. First, one would feel that a scar base is an ideal area for wound healing. Second, the perimeter of the scar tissue acts as a physical restraining splint, thus reducing the effect of tension on the margins of the incision. Undermining can still be utilized even in intramarginal excisions unless the surrounding tissue has altered vascularity. In some cases, it is even reasonable to perform an overgrafting technique in which all that is removed from the hypertrophic scar is the epithelial portion, leaving the underneath thick, fibrous tissue in place as a bed for a thick split-thickness or full-thickness graft. This reduces the amount of healing necessary, and also reduces wound tension.

Scar Revision

Scar Analysis

A discussion of scar revision is, in and of itself, a topic that could be a complete chapter, or in some cases, a small monograph or book. Thus, this discussion will be limited to some basic principles and concepts, rather than a multitude of specific cases.

The surgeon should be cognizant of evaluating multiple factors involving the patient and the scar areas before proceeding with the therapeutic recommendation. One of the first concepts is to evaluate the individual host in regard to tendency to form keloid or hypertrophic scars in other areas of the body, or with a family history of similar problems. One should also evaluate the person's psychological makeup in order that one is not trapped into dealing with a patient who has unreasonable expectations. This is especially true if
previous scar revisions have been unsuccessful. One may find that the patient may indeed have a lesion that cannot be improved. Younger patients will have greater skin tension, and thus will have a tendency to cause scar spreading and hypertrophy. This must be pointed out to the parents of pediatric patients, especially in areas where there is constant motion. The scars in these patients will usually become redder and remain erythematous for much longer periods of time.

One must also evaluate the time lapse between the injury and the patient's request for revision. Some surgeons feel that scars can be revised 2 to 4 months after injury in adult patients and older children, whereas in children less than 7 years of age, one should wait at least 6 months before revision. The rationale behind this concept is that the scar is most noticeable between 2 weeks and 4 months, and thus if revision is done, there will be a more obvious improvement with greater satisfaction from the child and parents. However, this author feels that since scar tissue continues to remodel and improve up through at least 12 months postinjury, one is justified in waiting until between 6 and 12 months postinjury before attempting scar revision. However, if there is functional deformity such as ectropion or difficulty in neck extension or opening the mouth, early revision is definitely warranted.

The surgeon must also pay special attention to various characteristics of the scars, which include pain, step-off deformities, pigmentation, length, shape, depression, width, hypertrophy, and direction. The treatment of painful scars is difficult and most likely will not lead to great success in the alleviation of pain. Distortion of anatomic landmarks is significant and is a definite indication for early repair, especially with ectropion or difficulty in extending the neck or opening the mouth. Step-off deformities along the vermilion-cutaneous border of the mouth or along the nostrils will require opening the wound and usually a multiple-layer closure. Stitch marks may indicate a tendency toward hypertrophic scar formation unless the previous surgeon's notes show that the sutures were left in for a lengthy period of time. If stitch marks are present, this may require dermabrasion, planing with the scalpel, or, if extremely pronounced, excision or incorporation in a running W or geometric pattern revision. Pigmented scars may be revised with dermabrasion or surgical planing if the lesions are superficial, but if they are deep-seated, complete correction may require full-thickness skin excision. The patient and family must be prepared for full-thickness excision if the latter is the case.

When planning a scar revision, the surgeon must remember that the scar has three components: the line, the contour, and the color. It is the integration of these three components into one final entity, the scar, that makes a patient either satisfied or dissatisfied with the result.

The line of the scar may be extremely minute, but still be visible if it passes across or near normal structures that are misapproximated, or it may be noticeable because it is widened (even though it is flat).

The contour of the scar involves a three-dimensional depression or elevation. Both depressed and elevated scars contribute to many patients' dissatisfaction. Scar contracture may not only be of significance in itself (this leads to scar hypertrophy or keloid formation), but also may cause displacement of normal structures such as the eyebrow, lower eyelid, lip, or nasal ala or columella. The contour may be altered also in the trapdoor type of scar.
Finally, the color of the scar is significant because any deviation from the color match with the surrounding tissue will usually make the scar noticeable. Examples of this are deep pigmented scar tissues in black individuals, darkly pigmented scar tissue in Caucasians, traumatic tattoos, or the markedly reddened scars seen frequently in the pediatric population. Thus, in selecting individuals for scar revision, each of the above factors must be taken into consideration when counseling the patient. The patient and his family must be reminded that a scar line is going to be inevitable when any scar is excised. Thus, one can only attempt to improve the result, but one cannot guarantee that an improvement will come to pass.

When discussing timing with a patient, one again must take into consideration such problems as malalignment, a spreading scar, a contracted scar, or trapdoor scar. Whereas color changes may improve with time, and thus repair could be delayed in these cases, the trapdoor defect or functional deformity caused by severe contraction should have early intervention.

**Methods of Scar Revision**

There are two factors in wound healing that are beyond the control of the surgeon when dealing with scar tissue and scar revisions. These are the area where the wound has occurred, and the amount of normal tissue that is lost. The surgeon should attempt, when at all possible, to place incisions in relaxed skin tension lines (RSTL). Unfortunately, there are occasions where the skin tension is considerable in any direction, and this will widen even the most meticulously revised scar. Scars overlying joints, or where there is marked skin motion, will also tend to be difficult to revise satisfactorily. The choice of excision and revision techniques will be discussed later, but during scar excision the scalpel should be held at right angles to the skin to prevent beveling. One can either utilize interrupted skin sutures with subcutaneous closure of an inverted absorbable suture, a single layer running closure (this is extremely unusual), or a running subcuticular suture to oppose skin edges after the subcutaneous tissue is closed with an absorbable inverted interrupted suture. The running subcuticular suture should be a 4-0 nonabsorbable material (5-0 nonabsorbable would be adequate if the wound is very short). When using the subcuticular suture, the wound edges may be taped together with Steri-Strips.

**Simple Excision - Primary Closure (Fusiform Excision)**

A simple excision is the most basic type of scar revision. This can be utilized in scars that parallel the RSTL and are reasonably short. The long axis of the incision must follow the RSTL even if it is curvilinear. The excision should not cross anatomic landmarks such as vermillion of the lip. Occasionally an M-plasty is needed for shorter scars. A variation of this technique is serial excision in which the scar may be extremely wide and thus must be excised in multiple stages. One disadvantage of this is the multiple-stage technique and thus the use of skin expanders has reduced the need for serial excision.

Occasionally the excision of a lesion and/or scar may be such that a single advancement flap is optimal (often in the forehead or facial groove regions). In order to reduce tension on the flap as it is advanced, triangles at each end of the flap (called Burrow's triangles) are removed, and the skin undermined in advance. At times, a bilateral advancement flap is necessary, as is often seen near the philtrum and columella of the upper lip and nose. Here, one performs a V-to-Y advancement.
Rotation Flaps

Rotation flaps are extremely useful in not only excision of small lesions, but also large lesions, especially in the nasolabial crease, and thus near a natural skin line. Also, large areas of cervical or buccal scars may be treated with large rotation advancement flaps. The general principle of this is shown, where a scar is excised and the flap is undermined. A triangle is removed at the distal end of the flap to reduce a dog-ear deformity.

Z-Plasty

Z-plasties are one of the most widely utilized scar revision techniques in reconstructive surgery. The principle of this technique is threefold: (a) the lengthening of the linear scar contracture, (b) realigning the scar within lines of minimal tension, (c) a breaking up of a straight line scar into a multiple Z configuration. A Z-plasty results from a transposition of two triangular flaps. The central line of the Z-plasty is called the central limb or incision. An attempt is made to construct the limbs of two equilateral triangles so that the angles are 60° with limbs of equal length. After excision of an unsightly scar, the flaps may be undermined and then transposed in a more satisfactory configuration in regard to the RSTL. Thus, the rotation allows these scars lie in a more satisfactory alignment but the price paid is that of two additional scars. Usually this trade-off is beneficial. Although the above discussion talked about equilateral triangles with 60° angles, one can adjust the lengthening by altering the angles at the apices of the transposition flaps. The greater the angle, the greater the lengthening. However, the majority of Z-plasties will have angles from 30° to 60°, with a 30° Z-plasty increasing the length approximately 25%, a 45° Z-plasty increasing the length 50%, and a 60° Z-plasty increasing the length by almost 75%. This technique can be utilized in a serial fashion and thus one could have multiple Z-plasties running along a very long line in an attempt to break up a prominent scar. Usually, certain basic principles can be followed to simplify Z-plasty utilization: (a) In the majority of cases, a simple Z-plasty with an angle from 45° to 60° is optimal. (b) The minimum angle utilized should be at least 30°, since an angle less than this would risk avascular necrosis of the tip. (c) Long scars are best treated with a multiple Z-plasty technique. (d) The central limb is placed along the scar line or line of contracture, with the lateral limbs drawn in one of either two directions, but utilizing the principle that the lateral limbs, which are parallel to each other, should also parallel the RSTL. If drawn incorrectly, they may lie perpendicular to the RSTL, and thus negate the entire concept of the scar revision.

W-Plasty

The concept of the W-plasty, or zigzag-plasty, is that of a series of interposed triangular advancement flaps that break up a straight line scar. Once the eye perceives any part of a scar, it will immediately follow it and see the entire scar. Multiple tiny Z-plasties are more likely to be visible to the eye than are running W-plasties, because the Z-plasties are more predictable and will lengthen the scar line. To perform a W-plasty, the scar is excised along with some normal tissue in a zigzag fashion. The completed W-plasty causes the scar to be irregular without adding to its length. It also breaks up the forces of contracture, and thus lessens scar tension. Since the W-plasties are irregular, this makes the scar less visible without causing any increase in the longitudinal aspect. The angles between
the interposition flaps are usually between 45° and 75°. The main drawback of the running W-plasty is a sacrifice of some healthy skin plus the difficulty in making the interdigitating parts conform to one another.

A Z-plasty has an advantage in that it elongates the contractile scar while changing the scar's direction, and thus will more likely place the resultant wound more nearly parallel to the RSTL lines. The Z-plasty will also utilize all the available skin without excising normal tissue. Finally, the Z-plasty will allow one to adjust the location of adjacent or displaced tissue. The W-plasty is an excellent procedure in the forehead, temporal area, chin, and cheeks, as well as the lower lip. It has an advantage of being less likely to displace normal anatomic landmarks since it does not involve the transposition of tissue. The W-plasty is indicated where the scars are perpendicular or nearly perpendicular to the lines of minimal tension. Unfortunately, a W-plasty will increase the tension in the scar because it does sacrifice some normal tissue and thus should be utilized only where there is adequate surrounding tissue. When the tissue surrounding the scar is somewhat deficient, or when the scar should be elongated, a Z-plasty is preferable.

A variation of the running W-plasty is a geometric broken-line closure, which is a design of irregular, unpredictable geometric figures as shown. The geometric broken-line closure came into use because the W-plasty was predictable. Therefore, one can excise a scar and make irregular, unpredictable geometric patterns to attempt to correct this deficiency of the W-plasty. These geometric closures can be triangles or rectangles of varying sizes, configurations, and patterns. Again, all incisions should run as close as possible to parallel the RSTL. This type of closure will not lengthen the scar but does cause the sacrifice of some normal tissue. It is usually wise to design the flaps to have a progression of heights so that the height is greater in the middle and less at either end.

**Dermabrasion**

Dermabrasion is a method of modification of facial scars by mechanically removing the epidermis and some, but not all, the dermis. This can be used as an adjunct to scar camouflage and occasionally can be used on the acutely injured patient to feather or touch up the lateral margins of the laceration or traumatic area. Dermabrasion can also combine this with a running Z-plasty, W-plasty, or geometric broken-line revision, but one must be careful to preserve hair follicles and sebaceous glands.

Dermabrasion is performed using any one of a number of devices designed for this procedure. These devices are lightweight and should have a very rapid RPM (ideally, 20,000 or greater). Although one could use wire brush abraders, these can be dangerous in the hands of the novice, and therefore diamond fraises of varying shapes (tapered, wheels, and cylinders) are available. The surgeon should be careful to protect both the patient and the operative team with the use of safety glasses, and also by keeping surgical sponges out of the operative field. This technique is best utilized for smoothing out depressions in the skin, including depressed scars, mild acne pitting, and fine wrinkles. It should not be utilized for keloids or hypertrophic scars. Postoperatively, one can utilize multiple types of wound care, including Telfa or Adaptic dressings, Debrisan ointment, or any of multiple antibiotic ointments.
The patient must be well informed about postoperative care and potential complications with dermabrasion. Specifically, erythema will occur and may last for 3 months or longer. Areas of telangiectasia may occur temporarily or less commonly be permanent. Small inclusion cysts (milia) are also a possibility. Hyperpigmentation has occurred, especially in dark-skinned individuals, and thus this technique should be used very sparingly in these patients. Hyperpigmentation can be brought about by exposure to sunlight, which patients are advised to avoid during the summer months. If sun exposure must occur, then the patient must utilize sun-screen of a high sun protective factor. Hypopigmentation can also occur postoperatively. The surgeon has minimal control over this hypopigmentation. Individuals who tend to form keloids or hypertrophic scars are not good candidates for this procedures. Finally, the patient must be warned that it may be necessary to perform more than one dermabrasion procedure.

The "Trapdoor" Scar Deformity

The trapdoor deformity is a description of a healing problem where one has the effect of retraction of curved scars, giving rise to an outward bulging projection over the surrounding skin. These scars are frequently seen in lacerations that are deep and have beveled edges. Multiple theories have been put forth concerning the trapdoor deformity, including lymphatic and venous obstruction, the piling up of fatty tissue, a bevel-shaped flap, scar hypertrophy, and mechanical factors. Indeed, the trapdoor deformity probably comes about because of a combination of factors. Specifically, when one has a horse-shoe-shaped, bevel-edged laceration, there may be three very important factors playing significant roles: (a) retraction of the curved scars with a depression in one area and a thickening of the skin over the beveled edge, with a bulging of the curved flap; (b) an interruption of tension lines by the curved scar; and (c) local movements of muscles in varying areas of the body (especially around the nasal and oral cavities), which leads to unequal retraction of muscle segments caused by differential divisions at the time of injury. The combination of these factors frequently will lead to the trapdoor deformity.

The trapdoor treatment depends upon the size of the scar. If the scar is small, then one can excise the area in a fusiform fashion and place the final scar parallel to the RSTLs. However, one must follow certain specific rules in utilizing this technique: (a) the skin incision must be perpendicular to the surface, (b) there must be slight undermining of the border, and (c) the deep layers must be reconstructed and closely approximated with subcuticular stitches. If the wounds are large, and thus preclude a fusiform excision along the long axis of the RSTL, then one should perform serial excisions in approximation to the RSTL, utilize Z-plasties or V-to-Y plasties in scars that are in an alignment that is not situated in the direction of the RSTL, and utilize a running Z-plasty or W-plasty on long scars that are perpendicular to the RSTL. The W-plasty is extremely useful in trapdoors along the mentum and chin regions.

Complications of Scar Revision

The cosmetic surgeon is faced with similar complications to those found in any other part of the body. These include hemorrhage, hematoma formation, infection, and wound dehiscence. However, certain other difficulties tend to surprise and frustrate both the surgeon and patient. These include reactions to the suture, which are usually heralded by the use of
subcutaneous or subcuticular catgut or chromic catgut. In these cases, the wound will become erythematous and there will be spontaneous drainage as the patient rejects the foreign body. One can only suggest to the patient that they treat the areas topically with antibiotic ointment, and the surgeon should remove any sutures that extrude toward the surface. Sebaceous hypertrophy also can be seen in the revision of facial scars, especially in patients who have seborrhea. This occurs with stimulation of the sebaceous apparatus in the areas of the incision margin. Thus, rather than attempting to excise these wounds (where the scar appears to be depressed), it would be better to either shave the edges with a scalpel, or to utilize dermabrasion. If excision and reapproximation are necessary, the wound should be beveled slightly in an attempt to alleviate the situation. If skin sutures are left longer than 8 days, one may find epithelial tunnels, which will then show suture marks. Thus, early removal of the skin sutures is necessary. Also, one may find milia (firm, pearly white cysts), which result from the trapping of hair follicles in the scar tissue beneath the epithelium. These are best treated by incising the area overlying the cyst and allowing the cyst and its lining to extrude. Finally, scar hypertrophy and keloid formation remain a distinct possibility and must be treated as mentioned in the earlier portions of this chapter.

In conclusion, the author would like to encourage the reader to continue to review the literature for new and better techniques in the evaluation of, and nonsurgical and surgical management of, patients with facial lacerations and wounds. The subject of wound healing is a rapidly changing area of otolaryngology, and it will require constant vigilance on the surgeon's part to keep abreast of new findings.
Facial nerve abnormalities in children represent a broad spectrum of pathologies, including numerous congenital and acquired etiologies. However, the principles of managing facial paralysis in children are essentially the same as those for adults. What follows is a review of the various facial nerve abnormalities encountered by otolaryngologists. In addition, patient selection and the surgical procedures used in managing specific facial nerve disorders are presented.

**Facial Nerve Abnormalities**

**Congenital**

**Möbius Syndrome (Congenital Facial Diplegia)**

Möbius syndrome is a rare congenital disorder that usually includes bilateral seventh nerve paralysis and unilateral or bilateral sixth nerve paralysis. Since the disorder was described, multiple authors have studied families with the syndrome (for review see McKusick). It is considered to have an autosomal dominant inheritance pattern with variable expressivity. The inheritance pattern is thought to be no higher than 1 in 50 in families in whom myopathies or other extremity anomalies such as clubfoot, arthrogryposis, or digital anomalies are not present.

The etiology of Möbius syndrome is unclear. Neuropathologic studies have noted that the nuclei of cranial nerves VI, VII, and XII are abnormal, with lesser abnormalities being found in the nuclei of cranial nerves III and XI. Other authors have reported that the facial nerves are smaller or absent at autopsy. Alternatively, primary failure of facial muscle development has been proposed as the etiology by Pitner et al, who also noted normal facial nuclei in their postmortem study.

The clinical observation of congenital extraocular muscle paralysis and facial paralysis is the typical presentation of this disorder. No mass lesions will be found on magnetic resonance imaging (MRI). Ophthalmologic consultation and management is mandatory. Reinnervation procedures such as cross-face grafts or hypoglossal-facial nerve anastomosis yield poor results, either due to the paucity of motor end plates or the atrophic seventh nerves. Significant improvements of resting tone and voluntary animation can result from temporalis muscle transposition, which brings in a new neuromuscular system.

**Hemifacial Microsomia**

The term hemifacial microsomia refers to patients with unilateral microtia, macrostomia, and mandibular hypoplasia. Goldenhar's syndrome (oculoauriculovertebral
dysplasia) is considered to be a variant of this complex and is characterized by vertebral anomalies and epibulbar dermoids. Although approximately 25% of patients with hemifacial microsomia have facial nerve weakness, one patient with Goldenhar's syndrome has been reported to have had aplasia of the facial nerve.

**Osteopetrosis**

Osteopetrosis is a generalized dysplasia of bone that may have an autosomal dominant or recessive inheritance pattern. The recessive form is more rapidly progressive and causes hepatosplenomegaly and severe neural atrophy secondary to bony overgrowth at neural foramina. Optic atrophy, facial paralysis, sensorineural hearing loss, and mental retardation are common in the recessive form, and death usually occurs by the second decade. However, in these severe cases of osteopetrosis, which were previously fatal, bone marrow transplantation has been reported to be of value.

The dominant form causes progressive enlargements of the cranium and mandible and clubbing of the long bones. Increased bone density is seen radiographically. Progressive optic atrophy, trigeminal hypoesthesia, recurrent facial paralysis, and sensorineural hearing loss are common. Complete decompression of the intratemporal facial nerve should be performed in patients with recurrent facial paralysis and radiographic evidence of osteopetrosis.

**Acquired**

**Trauma**

Approximately 90% of all congenital peripheral facial nerve paralysis improves spontaneously and most can be attributed to difficult deliveries, cephalopelvic disproportion, high forceps delivery, or intrauterine trauma. These types of congenital facial paralysis are often unilateral and partial, especially involving the lower division of the facial nerve. Since these etiologies involve extratemporal compression, surgical exploration or bony decompression are not indicated.

Blunt trauma resulting in temporal bone fracture is an unusual cause of facial paralysis in children. The lower incidence of skull and temporal bone fracture in children is due to adults sustaining head injuries more frequently and the increased skull compliance in children. These fractures are best evaluated with high-resolution temporal bone computed tomography (CT) scans.

Temporal and parietal blows to the head may occur anywhere along a coronal arc from the vertex to the cranial base. When the vector of force is directed toward the base, it classically passes toward the external auditory canal, deflects off the otic capsule, and extends anteromedially along the anterior edge of the petrous bone to the foramen lacerum and foramen ovale. The resulting fracture is described as a longitudinal temporal bone fracture. This is the most common type of temporal bone fracture (approximately 90%) and is also the most common type of fracture associated with facial nerve injury. The geniculate ganglion region of facial nerve is most frequently injured. The indications for facial nerve decompression and exploration are the same as those discussed in detail under the Bell's palsy section of this chapter.
Frontal and particularly occipital blows to the head tend to result in transverse fractures of the temporal bone. More severe head injury is usually required to cause these fractures. Since they often extend through the internal auditory canal or across the otic capsule, hearing loss and vertigo commonly result. Although only 10% to 20% of temporal bone fractures are transverse in orientation, they cause facial nerve injury in approximately 50% of patients. The anatomic region of the facial nerve most commonly injured is the labyrinthine segment.

Penetrating injuries to the extratemporal facial nerve should be explored urgently in order to facilitate identification of the transected distal branches using a facial nerve stimulator. If primary repair is not possible, the principles of facial nerve repair using cable grafts described later in this chapter should be followed. In infected wounds, urgent exploration and tagging of identified distal branches should precede control of the infection and granulation. Subsequent repair usually requires the use of cable grafts.

The risk of otologic surgical injury of the facial nerve is particularly high in children with congenital ear malformations. The discussion of specific malformations and the relative risk to the facial nerve is beyond the scope of this chapter. An additional group at higher risk for injury to the facial nerve are newborns who are undergoing mastoid surgery since the mastoid tip has not become pneumatized and the facial nerve exits the stylomastoid foramen laterally. In these children a semihorizontal, curvilinear incision should be used and as is the case with all otologic surgery, a facial nerve monitoring system should be used.

Infection

Bacterial

Facial paralysis as a complication of otitis media has become a rare disease in children due to the ready access to medical care and antibiotics. Takahashi et al reviewed their series of over 1,600 patients with facial paralysis and found that only 11 of these patients were younger than 20 years old and had facial paralysis due to otitis media (0.69%). They described the facial paralysis in this group of patients as having a slower progression and less complete paralysis than that seen in Bell's palsy. Temporal bone CT should be performed in all patients in order to eliminate the diagnosis of coalescent mastoiditis. Intravenous antibiotics in combination with myringotomy and tympanostomy tube placement remain our initial management algorithm for bacterial otitis media complicated by facial paralysis. Bacterial cultures should always be obtained at the time of myringotomy, and antibiotic selection should be tailored to the culture results.

Facial paralysis complicating mastoiditis or cholesteatoma is also rare. In Sheehy's series of over 180 children undergoing surgery for cholesteatoma, only one patient (0.6%) had facial nerve weakness. The surgical management of these patients includes mastoidectomy, excision of the cholesteatoma, and appropriate antibiotic therapy.

Infection with the spirochete *Borrelia burgdorferi* (Lyme disease) can result in facial paralysis. This tick-borne infection is endemic to the Northeast, and is named for the town of Lyme, Connecticut. Widespread infections have been reported from the West Coast, Midwest, and East Coast as well as throughout Europe and Australia. As is the case with
other spirochete infections, the clinical manifestations of Lyme disease are protean. Facial diplegia has been reported in Lyme disease and should be considered in children presenting with facial paralysis. Serologic diagnosis should be followed by antibiotic therapy. Tetracycline is considered to be the agent of choice; however, erythromycin and penicillin have been successfully used.

Viral

Herpes zoster oticus (Ramsay Hunt syndrome) is the etiology of facial paralysis in 3% to 12% of adults and approximately 5% of children reported in various series. It is characterized by severe pain and small, clear vesicles in the external ear canal and pinna, and rapidly progressive facial paralysis. Cranial polynuropathy is frequently seen, particularly in the vestibulocochlear nerve with resulting auditory and vestibular dysfunction. Involvement of cranial nerves V, IX, X, XI, and XII is much less common. Although surgical management using the middle cranial fossa approach for facial nerve decompression in herpes zoster oticus has been recommended, we currently do not advocate this specific surgical therapy since the facial nerve is more diffusely involved in this disorder than in Bell's palsy. In adults we routinely use intravenous acyclovir in the management of acute herpes zoster oticus; however, due to the mechanism of action of this drug, ie, DNA incorporation of acyclovir triphosphate and subsequent termination of DNA synthesis, its use in children is controversial. If a child is immunocompromised and has herpes zoster oticus, particularly with cranial polynuropathies, we would, however, administer this medication in consultation with our pediatric colleagues. Other viral infections such as primary chicken pox, mononucleosis, mumps, and poliomyelitis can result in facial paralysis that may or may not resolve spontaneously. For these specific viral infections, immunization when available, is the most effective preventative measure, and supportive care is required during the active infection. Facial reanimation procedures are sometimes required after adequate follow-up suggests that spontaneous recovery will not occur.

Benign or Malignant Neoplasms

Tumor involvement of the facial nerve should be considered in children with facial paralysis if one or more of the following clinical features are present: facial paralysis that progresses slowly over 3 weeks, recurrent facial paralysis involving the same side, facial weakness associated with twitching, long-standing facial paralysis (greater than 6 months), facial paralysis associated with other cranial nerve deficits and/or deficits referable to the brain stem, or evidence of malignancy elsewhere in the body. Gadolinium-enhanced MRI is extremely useful in imaging solid tumors involving the facial nerve, and high-resolution CT scans are useful in identifying bony erosion of the fallopian canal. In May’s series of 280 patients ranging in age from newborn to 18 years, he reported 15 patients (5.4%) with tumors causing facial paralysis. This group of benign and malignant tumors included the following diagnoses and number of patients: leukemia (3), schwannoma (3), meningioma (2), capillary hemangioma (1), granular cell myoblastoma (1), brain stem glioma (1), mesenchymal mass (1), congenital cholesteatoma (1), sphenoid giant cell tumor (1), and neurilemmoma (1). Rhabdomyosarcoma is one of the most common neoplasms in children, accounting for 10% to 15% of all childhood neoplasms, and therefore should be considered in children presenting with unilateral facial paralysis. If the underlying disorder requires surgical resection, the resulting gap in the facial nerve should be reconstructed at the time of resection with a greater
auricular nerve or sural nerve graft, as described later in this chapter. If the management of the disorder involves chemotherapy rather than surgical intervention, facial reanimation procedures as described by May are indicated if the child has persistent facial nerve dysfunction.

Miscellaneous Disorders

Guillain-Barré Syndrome

The onset of simultaneous bilateral facial paralysis suggests Guillain-Barré syndrome, sarcoidosis, sickle-cell disease, or some other systemic disorder. Guillain-Barré syndrome is a relatively common neurologic disorder affecting children and is an acute inflammatory polyradiculoneuropathy that progresses to varying degrees of paralysis. The etiology remains unknown; however, autoimmune or viral mechanisms have been considered. Classical histopathologic features of the syndrome include a lymphocytic cellular infiltration of peripheral nerves and destruction of myelin. The facial paralysis seen in these children is typically bilateral in nature and often resolves spontaneously after a prolonged course of paralysis. Although there is no role for surgical decompression of the facial nerve in this disorder, reanimation is only considered late in the course of the disease.

Melkersson-Rosenthal Syndrome

Merkersson-Rosenthal syndrome is a neuromucocutaneous disease with a classic triad of recurrent facial (labial) edema and recurrent facial paralysis associated with a fissured tongue. Patients with Melkersson-Rosenthal syndrome may not present with the complete triad, and although facial paralysis is the best-known neurologic symptom, it is not mandatory for the diagnosis. Headache, granular cheilitis, trigeminal neuralgiform attacks, dysphagia, laryngospasm, and a variety of cranial nerve and cervical autonomic dysfunction may also occur. The patient with Melkersson-Rosenthal syndrome may present at any age with any variety of classic and associated features that may wax and wane. Approximately one-third of the patients have recurrent facial paralysis as part of their syndrome. The underlying etiologic factor has been thought to be a neurotropic edema causing compression and paralysis of the facial nerve as it passes through the fallopian canal. Since the anatomically most constricted area of the fallopian canal is the meatal foramen and because most prior reports observed recurrence after transmastoid decompression, Graham and Kemink elected to decompress the proximal segment in addition to the mastoid segment of the facial nerve in all such cases by performing a combined transmastoid and midle cranial fossa facial nerve decompression and neurolysis of the nerve sheath. The preliminary data presented by Graham and Kemink suggest that edematous involvement of the facial nerve in recurrent facial paralysis does occur intratemporally and that the recurrent paralysis can be prevented by transmastoid and middle cranial fossa total facial nerve decompression with neurolysis of the facial nerve sheath. Recurrent paralysis over a prolonged period of time usually results in increasing residual dysfunction. If evidence of residual paresis exists, facial nerve decompression of the labyrinthine segment and geniculate ganglion through a middle cranial fossa exposure is recommended at the time of the next episode of paralysis.
Bell's Palsy

As is the case in adults, Bell's palsy is the most common diagnosis in cases of facial paralysis in children. Spontaneous recovery of normal function of the facial nerve has been reported to be 90% in children, as compared to 71% in the adult population. However, the incidence of Bell's palsy has been reported to be greater in adults (for review see Jenkins et al). It must be noted that Bell's palsy is a diagnosis of exclusion and represents loss of facial nerve function without a proven etiology.

Gadolinium-enhanced MRI has been advocated as a diagnostic tool in assessing facial paralysis. Gadolinium enhancement of the normal facial nerve does not occur. Therefore, enhancement of this structure would be due to increased extracellular fluid from edema, inflammation, or neoplasm. Our observations with gadolinium-enhanced MRI, as well as those of others, are supportive of Fisch's hypothesis of axoplasmic damming at the meatal segment with subsequent edema and nerve conduction impairment. However, a recent study demonstrated that there was no prognostic significance of gadolinium enhancement of the facial nerve on MRI in patients with Bell's palsy. Therefore, gadolinium-enhanced MRI is not indicated in every child with facial paralysis. In patients suspected of having a tumor from clinical or electrodiagnostic data, gadolinium-enhanced MRI along with a high-resolution CT of the internal auditory canal, fallopian canal, skull base, and parotid should be performed.

In addition to the MRI evidence of axoplasmic damming at the meatal foramen, Jackson et al published histologic data that also support this entrapment hypothesis of Fisch. In their case report, they presented the pathologic observation that there was a sharp demarcation between sclerotic nerve proximal to and necrotic nerve distal to the meatal foramen within the fallopian canal. A morphometric temporal bone comparative study of the fallopian canal at the meatal foramen in children and adults was published by Eicher et al. They found that the nerve diameter to canal diameter ratio at the meatal foramen was statistically significantly smaller in children. These findings that the facial nerve is not as tightly contained at the meatal foramen in children may explain the higher rate of spontaneous recovery in children with Bell's palsy.

Electroneurography (ENoG) provides a quantitative assessment of facial nerve function and allows a relative comparison between the normal and affected sides. Electroneurography is widely used in adults, and in our experience, as well as that of others, ENoG is equally useful in the evaluation of facial paralysis in children. Our criteria for surgical decompression include ENoG degeneration greater than 90% relative to the unaffected side and the operation within 14 days of onset. The figure illustrates the outcome of a child who was followed for 10 days with serial ENoGs that showed progressive degeneration, until she reached 100% degeneration preoperatively. Her complete facial paralysis resolved by 2 months postoperatively when her facial nerve function was assessed as a House-Brackmann grade 1.

Surgical Anatomy of the Facial Nerve

Detailed knowledge and familiarity with the complex course of the facial nerve and its anatomic relationship to other vital structures are essential to the surgeon who plans to operate in this area. The facial nerve (cranial nerve (CN) VII) exits the brainstem at the pontomedullary junction approximately 1.5 mm anterior to the vestibulocochlear nerve (CN VII).
VIII). The facial nerve is smaller in diameter (approximately 1.8 mm) than the oval CN VIII (approximately 3 mm in the largest diameter). A third smaller nerve, the nervus intermedius, emerges between CN VII and CN VIII and eventually becomes incorporated within the sheath of CN VII. After leaving the brain stem, CN VII follows a rostrolateral course through the cerebellopontine cistern for 15 to 17 mm, entering the porus of the internal auditory canal (IAC) of the temporal bone. Other important structures in the cerebellopontine cistern include the anterior inferior cerebellar artery (AICA) and the veins of the middle cerebellar peduncle. The AICA passes near or between CN VII and CN VIII; the veins are more variable in position and number. On entering the IAC the facial nerve occupies the anterosuperior quadrant of this channel for 8 to 10 mm. Then it enters the fallopian canal at the fundus of the IAC. The IAC is anterior to the plane of the superior semicircular canal. Superiorly, the bone overlying the IAC is within a 60° angle, whose vertex is the superior semicircular canal ampulla. At the entrance of the fallopian canal (meatal foramen) CN VII narrows to its smallest diameter, 0.61 to 0.68 mm. Only the pia and arachnoid membranes form a sheath around the nerve at this point, since the dural investment terminates at the fundus of the IAC. Many authors believe that the small diameter of the meatal foramen is an important factor contributing to the etiology of facial paralysis in certain diseases such as Bell's palsy and Ramsay Hunt syndrome.

The intratemporal course of the facial nerve has three distinct anatomic regions: the labyrinthine, tympanic, and mastoid segments. The labyrinthine segment is shortest (approximately 4 mm), extending from the meatal foramen to the geniculate ganglion. This segment travels anterior, superior, and lateral, forming an anterior medial angle of 120° with the IAC portion. The basal turn of the cochlea is closely related to the fallopian canal and lies anteroinferior to the labyrinthine segment of the facial nerve. At the lateral end of the labyrinthine segment the geniculate ganglion is found and the nerve makes an abrupt posterior change in direction, forming an acute angle of approximately 75°. Anterior to the geniculate ganglion the greater superficial petrosal nerve exits the temporal bone through the hiatus of the facial canal. The hiatus of the facial canal is quite variable in its distance from the geniculate ganglion. The hiatus of the facial canal also contains the vascular supply to the geniculate ganglion region. The tympanic, or horizontal, segment of the nerve is approximately 11 mm long, running between the lateral semicircular canal (LSC) superiorly and the stapes inferiorly, forming the superior margin of the fossa ovalis. Between the tympanic and mastoid segments, the nerve gently curves inferiorly for about 2 to 3 mm. The mastoid, or vertical, segment is the longest intratemporal portion of the nerve, measuring approximately 13 mm. As the nerve exits the stylomastoid foramen at the anterior margin of the digastric groove, an adherent fibrous sheath of dense vascularized connective tissue surrounds it. The stylomastoid artery and veins are within this dense sheath.

**General Principles in Facial Nerve Surgery**

Whenever the facial nerve is to be surgically exposed, several technical points must be observed. First, a system for monitoring facial nerve function during the operation should be employed. One of the simplest monitoring methods is visual observation during critical stages of the operation. Needle electromyography can also be used if the equipment is available. No matter which monitoring technique is used, it is necessary that the side of the face in which the nerve is to be exposed be draped in a manner that allows visual observation.
The forehead, eye, mouth, and chin should be visible. The endotracheal tube should be secured to the opposite side without placing tape on the side of the mouth to be observed. Towels drape the posterior, superior, and inferior margins; a fourth towel is placed along the anterior profile. An abdominal clear, noniodinated plastic adhesive drape is placed over the face and operative area. An observer is thus able to see the entire face during the procedure and determine if any of the muscles move in response to surgical manipulation or electrical stimulation of the nerve. The scrub nurse or circulating nurse has been found to be the best observer if the surgeon asks for observation during critical periods of the procedure. Electromyographic needle electrodes can be placed in the orbicularis oculi and oris muscles and attached to electromyographic equipment for auditory feedback of neural activity throughout the surgical procedure.

Instrumentation is crucial to a successful exposure of the facial nerve. The largest diamond burr that the operative site can safely accommodate should be used when the surgeon is near the fallopian canal. Cutting burrs have a tendency to catch and jump unexpectedly and can cause severe injury to the nerve. Continuous suction-irrigation keeps the burrs clean and also dissipates heat, which can induce neural damage.

The final layer of bone over the nerve should be removed by blunt elevators specially designed for this purpose, such as the Fisch raspatory. These instruments are thin but strong enough to remove a thin layer of bone. Stapes curettes are usually too large and can cause compression injury to the nerve. If a neurolysis is to be performed, disposable microblades are available (Beaver no 59-10). Sharp dissection is less traumatic than blunt elevation when the nerve must be lifted out of the fallopian canal. The medial surface of the nerve usually adheres to the bone and contains a rich vascular supply. Cauterization near the nerve should be performed only with bipolar electrocautery and insulated microforceps.

Middle Cranial Fossa (Transtemporal) Approach: IAC Porus to Tympanic Segment

The middle cranial fossa exposure is used to expose the IAC and labyrinthine segments of the facial nerve when preserving existing auditory function is desirable. The geniculate ganglion and tympanic portions of the nerve can also be decompressed from this approach.

Technique

The patient is placed supine on the operating table with the head turned so that the involved temporal bone is upward. The hair is shaved 6 to 8 cm above and anterior to the ear and 2 cm posterior to it. The surgeon is seated at the head of the table with the instrument nurse at the anterior side of the patient's head. A 6x8 cm posteriorly based trapdoor incision is marked in the hairline above the ear. If exposure of the mastoid is necessary, the inferior limb of the incision can be carried postauricularly. The skin flap is elevated to expose the temporalis muscle and fascia. A 4x4 cm temporalis fascia graft is harvested for use during closure of the IAC dural defect. An anteriorly based trapdoor incision is used to elevate the temporalis muscle and periosteum. Staggering the levels of the muscle and skin incisions provides for a double-layer, watertight closure at the completion of the procedure.
The temporal root of the zygoma is exposed during elevation of the temporalis muscle. This landmark represents the level of the floor of the middle fossa. Dural fishhooks are placed in the skin and temporalis muscle flaps for retraction. A 3x5 cm bone flap for facial nerve decompression, or 4x5 cm bone flap for tumor excisions, centered above the temporal root of the zygoma, is fashioned with a medium cutting burr. It is important to keep the anterior and posterior margins of the craniotomy parallel to facilitate placement of the self-retaining retractor.

Branches of the middle meningeal artery are occasionally embedded within the inner table of the skull; therefore, elevation of the bone flap must be performed in a controlled manner. Bipolar coagulation and bone wax may be necessary to control bleeding. Elevation of the dura from the floor of the middle fossa can be one of the most difficult steps. Blunt dissection and magnification greatly facilitate dural elevation. The dura is elevated from the posterior to anterior direction to prevent accidental injury to an exposed geniculate ganglion and greater superficial petrosal nerve. Bipolar coagulation is used to cauterize dural reflections within the petrosquamous suture before transection with a scissors.

The elevation proceeds until the petrous ridge is identified medially and the arcuate eminence, meatal plane, and greater superficial petrosal nerve are exposed anteriorly. No attempt is made to identify the middle meningeal artery and accompanying troublesome bleeding veins. The tip of a self-retaining (House-Urban) retractor is placed at the petrous ridge anterior to the arcuate eminence and medial to the meatal plane. A medium diamond burr and a suction-irrigation apparatus are used to identify the blue line of the superior semicircular canal (SSC). A preoperative Stenvers' projection radiograph helps to determine the level of the SSC in relation to the floor of the middle fossa and the degree of pneumatization above the SSC.

Drilling begins posterior to the arcuate eminence over the mastoid air cells until the dense yellow bone of the otic capsule is identified. Otic capsule bone is slowly removed until the blue outline of the SSC is seen. The IAC is located by removing bone with a 60° angle anterior to the blue line of the SSC, and with the vertex based at the SSC ampulla. This dissection is continued until approximately 180° of the IAC are exposed for facial nerve decompressions, or 270° of the IAC are exposed for schwannomas. Because of the close proximity of the SSC and the basal turn of the cochlea, only approximately 120° of the circumference of the IAC can be safely removed in its lateral 5 mm or so. The facial nerve occupies the anterosuperior portion of the IAC. Laterally the vertical crest (Bill's bar) marks the division between the superior vestibular nerve and the meatal foramen containing the facial nerve.

The entrance to the fallopian canal is the narrowest, most delicate portion of the facial nerve and consequently the most challenging portion of the dissection. At the meatal foramen the facial nerve turns anterior and slightly superior. The basal turn of the cochlea can be within 1 mm inferiorly, and the ampulla of the SSC can be directly posterior to the nerve. The labyrinthine segment is followed to the geniculate ganglion. If the facial nerve needs to be exposed distal to the geniculate ganglion (eg, as with facial neuromas or with some traumatic injuries to the facial nerve) the tegmen tympani is removed with care to avoid injury to the head of the malleus and incus. The tympanic segment is easily seen to turn
abruptly posterior; it is followed to where it coursed inferior to the lateral semicircular canal (LSC). It is advisable to leave a thin shell of bone covering the nerve until its entire course is identified. The final layer of bone is removed by small blunt elevators. The nerve is tightly confined within the labyrinthine segment of the fallopian canal; larger curettes should be avoided to prevent compression injury. If the nerve is to be decompressed, a neurolysis is the final step. A disposable microscalpel (Beaver no 59-10) is used to slit the periosteum and epineural sheath.

Alternative methods to locate the facial nerve may be necessary, especially in traumatic cases. The greater superficial petrosal nerve can be traced posteriorly to the geniculate ganglion, or the tegmen tympani may be fractured and the tympanic segment visible through the fracture. The tympanic segment is then used to locate the geniculate ganglion and labyrinthine segments.

At the end of the procedure a free temporalis muscle graft is placed within the IAC and a corner piece of the bone flap is fashioned to cover the defects in the tegmen tympani and IAC. This prevents herniation of the temporal lobe into the middle ear or IAC. The temporalis fascia previously harvested is placed over the free bone graft to help seal the dural defect at the IAC. The squamosal craniotomy bone flap is replaced, and the temporalis muscle is closed with interrupted absorbable sutures. The skin is closed in layers with particular care in closing the galea. No drain is placed. A mastoid-type pressure dressing is applied.

Advantages and Uses

The middle cranial fossa route is the only method that can be used to expose the entire IAC and labyrinthine segment with preservation of hearing. This, in combination with the retrolabyrinthine and transmastoid approaches, enables visualization of the entire course of the facial nerve and still preserves function of the inner ear. The middle cranial fossa technique is the most commonly used for decompression of the facial nerve in Bell's palsy, herpes zoster oticus, and longitudinal temporal bone fractures. However, as described earlier in this chapter, this approach may be useful in the management of patients with schwannoma of CN VII or CN VIII, as well as with patients with Melkersson-Rosenthal syndrome.

Postoperative Care - Complications and Their Treatment

The anatomy of the floor of the middle cranial fossa is quite variable and presents some difficulty in identification of landmarks. The Stenvers' projection radiograph provides important anatomic information regarding the degree of pneumatization above the superior semicircular canal, and should be performed in all cases to minimize the risk of surgical injury to the SSC. In addition, the surgeon must have a precise knowledge of three-dimensional anatomy of the temporal bone. Many hours in a temporal bone dissection laboratory are required to attain the delicate microsurgical skills that are necessary for this type of surgery.

Conductive and sensorineural hearing losses can both result from middle cranial fossa facial nerve decompression. Conductive hearing loss can be secondary to temporal lobe herniation or ossicular disruption during dissection in the attic. A free bone graft, as already described, prevents temporal lobe herniation. Sensorineural hearing loss can result from direct
injury to the inner ear by the drill exposing the cochlea or semicircular canals or from translational injury by the drill striking an ossicle. Should the SSC be entered during the surgical dissection, the fenestration should be immediately occluded with bone wax. Injury to the internal auditory vessels within the IAC can also result in loss of inner ear function. Loss of vestibular function can occur by the same mechanisms.

Postoperative intracranial complications including meningitis, temporal lobe edema, and epidural hematoma formation are possible. Perioperative antibiotics administered over 48 hr are recommended. Fluid restriction and Decadron are used for the first 3 days postoperatively in order to minimize temporal lobe edema following intraoperative retraction. In addition, our longer craniotomy flap decreases the amount of temporal lobe retraction required for complete exposure of the internal auditory canal and fallopian canal. With adequate intraoperative hemostasis using the bipolar cautery, Oxycel, and dural tacking sutures, we have never had a clinically significant postoperative epidural hematoma develop.

Leakage of cerebrospinal fluid (CSF) must be avoided in order to prevent meningitis. All exposed mastoid air cells must be obstructed with bone wax. A temporalis muscle free graft is placed into the superior aspect of the IAC to separate the posterior fossa from the extradural floor of the middle cranial fossa. Temporalis fascia is then used to provide a second layer of closure between the posterior fossa and the extradural middle fossa. Meticulous care must be taken to assure that there are no dural dehiscences overlying the temporal lobe through which CSF may drain. If these are identified, a temporalis fascia or muscle patch must be used to repair the dural tears to prevent CSF leaks. After a three-layer watertight closure of the temporalis muscle, galea, and scalp, a mastoid-type dressing is applied daily for 5 days postoperatively. Should CSF leakage persist, a temporary lumbar fluid drain is placed and the patient is kept on bed rest. If the CSF leakage does not resolve within 7 days after placement of the lumbar drain, reexploration of the surgical field is indicated to identify and seal the area of CSF egress.

Uncontrolled bleeding or injury to the AICA poses the most serious complication during the operation. The middle cranial fossa approach does not provide adequate access to the entire cerebellopontine angle. The AICA and accompanying veins can loop into the IAC. Control of bleeding of these vessels may require a suboccipital exposure. Injury to the AICA results in brain stem and cerebellar infarction of a variable degree, depending on its size and the area of its terminal arterial supply.

Nerve Repair

Whenever the continuity of the facial nerve has been disrupted by traumatic injury, iatrogenic injury, or tumor invasion, every effort should be made to restore its continuity. In some instances an end-to-end reapproximation can be accomplished, but if any tension occurs at the anastomotic site, an interposition nerve graft has a better chance of providing facial movement. All nerve repair techniques produce synkinesis, but sphincteric function of the mouth and eye are usually restored. Newer microsuture techniques and instrumentation should be employed to enhance return of function.

In general the injured ends of the nerve should be freshened at a 45° angle. Experimental evidence has shown that cutting the nerve at this angle exposes more neural
tubules and improves regrowth of the nerve. In addition, a fresh razor blade induces less crush to the nerve than a scalpel blade or scissors does. We have found that the perineurium of CN VII does not hold 10-0 sutures, and attempting to suture it increases trauma to the neural tubules. Removing a portion of the epineurium before suturing prevents connective tissue growth at the anastomotic site. If the epineurium is cleaned from the end of the nerve for approximately only 0.5 mm, sutures can still be placed in the epineurium for reapproximating the nerve segments. Three or four sutures of 10-0 nylon are placed with jeweler's forceps or longer instruments (19 cm microforceps) for anastomosis in the cerebellopontine angle. At the brain stem two or three sutures are placed.

When an interposition graft is required, the greater auricular nerve and sural nerve are the preferred graft donor sites. The greater auricular nerve is readily available near the operative field if it is not involved in resection of a neoplasm, and has approximately the same diameter as that of the facial nerve. It is easily located midway, perpendicular to a line drawn between the mastoid tip and the angle of the mandible. If a graft of greater than 8-10 cm is required, the sural nerve should be used. The sural nerve has another advantage, in that the peripheral portion of the nerve has many branches that can be used to reconstruct the branching pattern of the facial nerve. There is little discomfort from removing the sural nerve, since it provides only a small area of sensation to the lateral lower leg and foot. The sural nerve is found immediately posterior to the lateral malleolus, along the saphenous vein. The nerve graft should be 10% to 20% larger in diameter than the facial nerve and long enough to ensure a tension-free anastomosis.
Dermatologists and facial plastic and reconstructive surgeons enjoy close collaboration in the evaluation and treatment of patients with cutaneous disorders of the head and neck. Each specialty can benefit from the scientific and therapeutic expertise of the other. Although many of the diseases mentioned in this chapter are covered elsewhere in this book, discussion is repeated here because a different perspective is often valuable. Particular emphasis will be devoted to melanocytic and vascular disorders.

**Melanocytic Lesions**

Disorders of melanocytes cause abnormalities of pigmentation resulting in skin discolorations of tan, brown, bluish-gray, blue, and black. Mixtures and gradations of these colors may be seen, as well as the lack of melanocytic pigment altogether, as in albinism and vitiligo. The vast majority of pigmentary abnormalities seen in children are benign. Malignant melanoma in children is distinctly rare. We will discuss congenital and acquired benign melanocytic tumors, focusing on the precursor lesions of melanoma that the facial plastic surgeon will most likely treat.

**Mongolian Spot**

A mongolian spot is a flat, grayish-blue discoloration with either sharp or ill-defined borders. Histologically it is caused by the presence of melanocytes in the mid- to deep dermis. Lesions are usually seen over the spine or occasionally on the shoulders or neck. Mongolian spots are a normal variant in Asians (90% to 99% of infants), blacks (96% of infants), and in more darkly pigmented races such as Hispanics (46% of infants). Most lesions involute by age 5 years. Histologically, they are similar to the nevus of Ota and nevus of Ito.

**Nevus of Ota and Nevus of Ito**

The nevus of Ota or nevus of Ito is a flat, hyperpigmented lesion that may be bluish-purple to bluish-gray to bluish-brown in color. Discoloration within the area of skin may be diffuse or concentrated in small macules against a background of fainter hyperpigmentation. The spots are typically larger than Mongolian spots and they do persist. Sixty percent are present at birth, and 40% develop in adolescence. The nevus of Ota is located within the dermatomal distribution of the ophthalmic and/or maxillary branch of the trigeminal nerve. Involvement of mucosae is common, including the hard palate, pharynx, and nasal mucosa as well as the auditory canal. In addition, ocular structures are pigmented in around 50% of cases. The nevus of Ito is usually unilateral and in the rough dermatomal distribution of either the lateral brachial or posterior supraclavicular cutaneous nerve branches; thus, the supraclavicular, deltoid, or scapular regions are most often affected.
Camouflaging makeup is the only effective therapy for Mongolian spot or nevus of Ota/Ito, although the Q-switched ruby laser shows promise. Since the melanocytes are deep in the dermis, wounding procedures such as dermabrasion or chemical peels are ineffective. Likewise, the yellow dye pigmented lesion laser will not reach the necessary depth. Surgical intervention is not required unless melanoma supervenes; fortunately, melanoma is only slightly more common in patients with either the nevus of Ota or nevus of Ito than in unaffected subjects.

**Melanocytic Nevi**

Melanocytic nevi (nevocellular nevi; moles) are benign growths composed of nests of melanocytes. Whether they are hamartomas or true neoplasms is controversial. Melanocytes are derived from the neuroectoderm, and the migration of melanoblasts from the skin during development may help explain the different presentations of congenital melanocytic nevi.

Melanocytic nevi may be either congenital or acquired. They are congenital in only 1% of Caucasians. However, most Caucasians will gradually acquire melanocytic nevi, with a peak number of lesions around age 15 in males and in the third decade in females. Following a peak of about 15 to 50 lesions, nevi undergo gradual involution. By the ninth decade, the average Caucasian has only four nevi.

**Acquired Melanocytic Nevi**

Acquired melanocytic nevi undergo a typical life cycle. Lesions start out flat, gradually become raised, then involute completely. There are color changes and histologic appearances corresponding to each of these stages. Flat, dark brown lesions are junctional nevi, in which nevus cell nests are confined to the lowest portions of the epidermis at the dermo-epidermal (DE) junction. Lesions that are medium-brown to flesh-colored and are dome-shaped have nevus cell nests both at the DE junction and in the upper dermis. These are termed compound nevi. Soft, rounded, tan to flesh-colored lesions are known as intradermal nevi, since nevus cell nests are confined to the dermis alone.

Prediction of the histology of the lesion will help determine the optimal mode of excision. If one is suspicious of malignant melanoma, it is desirable to remove the entire lesion, down to subcutaneous tissue, with a 2-mm margin. If the specimen is found to be a melanoma, a wider excision is required. When an entirely banal-appearing elevated nevus is removed for cosmetic reasons, it is axiomatic that cosmesis be maximized. Often "shave" or tangential excisions give the best results. It is easy to see that a junctional nevus is most likely, and an intradermal nevus least likely, to be completely removed by tangential excision. However, even in the case of intradermal nevus, cosmesis is often best served by a tangential excision, with the understanding that reexcision may be required. The lesion is most easily removed after distending it with local anesthetic, making the surrounding skin taut by stretching with the contralateral hand, and then applying either a razor blade (the double-edged variety is sharpest) or a No 15 blade with sweeping motions in a slightly concave arc.
**Congenital Melanocytic Nevi**

By definition, congenital melanocytic nevi (CMN) are present at birth or shortly thereafter; all other nevi are acquired. The CMN is typically medium to dark brown in color. CMNs are typically raised, papillated, verrucous, and often hairy. They are often large; almost any nevus larger than 1.5 cm in diameter is congenital, whereas almost every nevus smaller than 7 mm is acquired.

Some histologic features may be suggestive of CMN, but none of them is definitive. Histologic features suggestive of congenital nevi are the following:

1. nevus cells present in the lower two-thirds of the reticular dermis in almost all cases, and into the subcutaneous layers in more than half of cases;

2. nevus cells disposed between collagen bundles or in "Indian" files;

3. nevus cells commonly involving appendages, nerves, and vessels in the lower two-thirds of the reticular dermis or subcutaneous tissue.

In addition, there may be a fibroblastic stromal response in the dermis, giving CMN a thickened, pachydermatous texture. Horn cysts of the epidermis may be seen, especially in verrucous CMN.

There are two principal considerations in the management of CMN: the increased incidence of melanoma in giant CMN and the cosmetic morbidity of CMN of any size. The larger the lesion, the greater the potential for malignancy. There is no doubt that there is an increased incidence of malignant melanoma in giant CMN. Data are less conclusive for smaller lesions.

The definition of a "giant" CMN in the literature varies. Definitions include lesions greater than 20 cm in the largest dimension, lesions involving the major portion of an anatomic area, lesions larger than 144 square inches, or lesions that cannot be excised and their defect closed primarily in a single setting. Kopf's criteria seem the most practical: giant CMN are 20 or greater, medium CMN are greater than 1.5 cm but less than 20 cm in diameter, and small CMN are 1.5 cm or smaller.

Giant CMN are sometimes termed "garment" or "bathing trunk" nevi because they may encase an anatomic unit such as the trunk or an extremity. Cosmetic morbidity is considerable. Giant CMN are typically hairy (hence, the term "giant hairy nevus"), irregular in texture and shape, and varied in coloration, ranging from tan to brownish-black. They occur in less than 1 in 20,000 births.

Patients with giant CMN have an increased lifetime risk for development of malignant melanoma, variously estimated to be between 2% and 20%. Most series find an incidence of malignant transformation around 8% to 9%.

It is important to note that the majority of melanomas associated with giant CMN appear before the age of 5 years. This fact is especially impressive since melanoma is least
common in the first decade of life. Only 2% of all cases of melanoma occur in childhood.

Another risk of giant CMN is leptomeningeal melanocytosis, in which nevus cells involve the brain, spinal cord, or meninges. It is seen in large CMN involving the head or midline of the back. Proliferation of cells or supervening melanoma may develop and result in a mass effect causing hydrocephalus, nerve damage, or seizures. Prognosis is poor in these patients.

To prevent melanoma and improve cosmesis, complete surgical excision of giant CMN is advised. Excision should be performed as soon as practical, taking into account the patient's general health and risk for complications of general anesthesia. Excision should extend down to and include muscle fascia, since nevus cells may infiltrate to this level. Dermabrasion, cauterization, laser vaporization, cryotherapy, or any other superficial modality does not ablate the majority of nevus cells and should not be used. Nevi may extend too widely or deeply (eg, to periosteum or bone) to make complete excision possible. In these patients the most abnormal areas should be excised, with close follow-up of the CMN that remains. The dermatologist can help identify the most abnormal portions of the nevus.

Data are less conclusive regarding the lifetime risk for developing melanoma in small and medium CMN. Prospective studies have not been done. Retrospective studies using historical data (ie, whether the patient has developed melanoma in a precursor nevus thought to have been present at birth) have shown that patients with small CMN have a 21-fold increase in melanoma incidence. Correspondingly, patients who develop melanoma in nevi with histologic features typical of CMN were found to have a three- to tenfold increase in melanoma. However, retrospective data are not always reliable, especially when relying on historical data. In addition, since congenital and acquired melanocytic nevi may appear similar histologically, lesions cannot be definitively identified as congenital on histologic grounds.

Regardless of the relative risk, melanoma may develop in small or medium CMN; there are, however, important differences from melanoma seen in giant CMN. Melanoma in small or medium CMN usually develops after age 12 and begins with radial growth typical of superficial spreading melanoma that is more easily detected. Melanoma in giant CMN, on the other hand, most commonly develops before 5 years of age and, because of predominantly deep histologic components, is often nodular or metastatic before 5 years of age and, because of predominantly deep histologic components, is often nodular or metastatic before it is detected. In practical terms, this means that the decision whether to excise small or medium CMN can often be delayed until adolescence. Melanoma in these lesions usually will not present before adolescence, and even if it does occur, it can usually be detected in early, more curable stages of growth.

Factors that may favor the excision of small or medium CMN include

1. lesions whose removal would result in minimal cosmetic deformity;

2. lesions that are difficult to follow clinically because of anatomic location, eg, scalp (patient self-examination is difficult);

3. lesions in patients with poor reliability either in self-examination or in regular
physician examinations;

4. lesions whose continued presence is psychologically troubling to the patient.

**Spitz Nevus**

Another variety of acquired melanocytic nevus is Spitz nevus. Another synonym is spindle and epithelioid cell nevus; outmoded terminology includes the term *benign juvenile melanoma*. The latter term arose because the Spitz nevus may be histologically confused with melanoma. However, these lesions are benign, usually arising in childhood, typically on the extremities or face. Growth is rapid; many lesions arise over weeks or months. Lesions typically have a reddish hue and may be pink to brownish-red to bluish-black with a shiny, sometimes slightly scaly surface. They are firm in texture and are usually entirely banal in appearance.

Histologically, most lesions are compound melanocytic nevi with vascular ectasia (hence the reddish color). The nevus cell nuclei are pleomorphic-appearing and may be confused with malignant melanoma. For this reason, it is usually prudent to have biopsy material reviewed by an experienced dermatopathologist. The lesions should be completely excised with 1-2 mm margins since recurrent benign Spitz nevi may be easily confused with melanoma clinically or histologically.

**Blue Nevus**

Blue nevi are deep blue to bluish-black (steel blue) nodules. On the nose, lesions may appear grayish-brown. They are most commonly seen on the dorsa of the hands, feet, forearms, shins, and face. The common blue nevus is a type of benign intradermal melanocytic nevus. Onset is early in life, but lesions grow slowly and rarely measure over 10 mm in diameter. Most are less than 7 mm in size. The melanocytes in these lesions are dendritic and deep, causing the lesion's bluish appearance (Tyndall effect). Because of the depth of the cells, tangential excision will not extirpate the lesions; full-depth excision of epidermis and dermis is required for complete removal.

**Recurrent Melanocytic Nevi**

Melanocytic nevi that are incompletely removed, especially after tangential excision, may result in scars with irregular pigmentation and may clinically resemble superficial spreading melanoma. There may be histological confusion with melanoma as well, but the lesions are benign.

**Vascular Growths and Malformations**

A variety of vascular lesions may be seen in infants and children. Although they all have the common feature of vascularity, their appearance, natural history, histopathology, and treatment differ widely. Some patients may benefit from early surgery, whereas in the majority of cases optimal cosmesis and function are best achieved by allowing natural involution.
Vascular lesions can be classified according to their clinical appearance. Raised lesions usually involute with time, whereas flat lesions tend to persist.

**Flat Vascular Growths**

Salmon patches and port-wine stains are flat lesions that are true vascular malformations. The term *nevus flammeus* should not be used because it has been applied to both lesions and is, therefore, ambiguous.

**Salmon Patches**

Salmon patches are very common; they are seen on the nape of the neck (“stork bites”) in 50% of infants. They may also be seen on the glabella (“angel’s kiss”) or upper eyelids. Lesions are pink or light red in color, and are accentuated by physical exertion or crying. Eyelid lesions generally fade away by age 3 to 6 months and glabellar lesions by age 5 to 6 years, but many nape lesions have a greater tendency to persist and are seen in 25% of adults. The vast majority of lesions do not require treatment, but persistent, cosmetically disturbing lesions may be removed using yellow dye vascular lasers (see discussion under Port-Wine Stains). Histologically, lesions are composed of ectatic capillaries thought to represent persistent fetal circulatory patterns. This is perhaps due to localized dysfunction of nerves of the vascular plexus, such as loss of sympathetic neurons. The etiology is not genetic; discordance among monozygotic twins has been reported.

**Port-Wine Stains**

Port-wine stains (PWS) are flat, congenital, vascular malformations named for their red to reddish-purple color. They may cause a pink discoloration of the skin in early infancy, but eventually assume their characteristic color, which is darker than that seen in salmon patches. A PWS is composed of dilated mature capillaries with no tendency to involute. Unlike juvenile hemangiomas, there is no cellular proliferation and no tendency to grow out of proportion to normal somatic growth. PWSs are usually unilateral, most commonly located on the face or extremities, but any body area may be involved. When the ophthalmic dermatome of the trigeminal nerve is involved, either alone or in conjunction with other areas, the patient should be evaluated for the presence of the Sturge-Wever syndrome, in which there is also a vascular malformation of the ipsilateral meninges and cerebral cortex that may result in seizures, mental retardation, hemiplegia, or glaucoma.

Port-wine stains have a natural history that may favor early intervention. Lesions tend to darken with time, and, although they are initially flat, may eventually develop raised cavernous portions that are easily traumatized. The skin may become thickened, nodular, and irregular. Early intervention may be indicated to forestall cosmetic and hemorrhagic complications.

The treatment of choice for PWS is a yellow light laser. These lasers produce light at 577 or 585 nm, the wavelengths of local peaks of the light absorption curve of oxyhemoglobin. Equally important, these wavelengths are relatively less absorbed by other components of the skin such as melanin or water. Thus, the light energy from the laser is selectively absorbed by hemoglobin, causing coagulation and sludging of red blood cells,
leading to occlusion and fibrosis of the abnormal vessels. Some of these lasers pulse the laser output to bursts shorter than the thermal relaxation time of hemoglobin, thus minimizing the spread of heat beyond the vessel walls, reducing the risk of damaging and scarring surrounding tissues. Some yellow dye lasers lack pulsing. Studies have shown scarring rates less than that seen with the use of the 514-nm argon laser, in which light output is neither as selective for hemoglobin nor as short in duration as the pulsed dye laser. As with any laser modality, scarring can be minimized by using the lowest effective power settings; this is determined by performing test treatments at different power fluences. Different body sites respond to different power fluences. Experience with the argon laser has shown a greater tendency for scarring of the lips and neck than other areas of the head and neck.

With the argon laser, significant scarring occurred in 20% to 25% of patients, with the highest rates seen in infants and young children. For this reason, treatment was often deferred as late as possible, even though the PWS continued to worsen in severity. The pulsed dye laser does not scar as much as the argon laser, and thus can be used in infancy and early childhood before cavernous lesions and psychosocial trauma develop. The yellow dye laser does not work as well with cavernous lesions as it does with flat ones. The copper vapor laser, which also produces yellow light, is a newer modality that shows promise for treatment of the cavernous portions of port-wine stains.

**Raised Vascular Growths**

**Juvenile Hemangiomas**

*Juvenile hemangioma* is a term that includes the capillary hemangioma, the cavernous hemangioma, and the mixed capillary-cavernous hemangioma. Other synonyms include strawberry hemangioma, strawberry nevus, immature capillary hemangioma, and cellular hemangioma. The capillary type of juvenile hemangioma typically has a strawberry-red color and raised surface. Cavernous lesions are deep, bluish-hued nodules with normal overlying skin. An individual lesion may have a mixture of superficial ("capillary") or deep ("cavernous") components. Juvenile hemangiomas are seen in 0.8% to 2.5% of all newborns and in 10% to 12% of Caucasian infants by age 1 year. Most lesions appear by age 2 months and are most prevalent on the head and neck. Thirty percent of patients have multiple lesions. If the lesion is not obvious at birth, the first manifestation may be a pale, circumscribed area of skin with telangiectasias with or without subjacent swelling. Early in life the lesions undergo a proliferative phase that may be alarming in rate. It is essential to inform the patient's parents of the nature of the lesion and its natural history. The most important datum is that at least 95% of these lesions undergo complete resolution. Superficial lesions typically enlarge for 6 to 8 months, achieving the characteristic "strawberry" appearance, whereas deeper, cavernous components may grow for up to 12 months. Fading of color is the first sign of involution, occurring at 12 months of age in the superficial lesions. Flattening occurs by the end of the 2nd year. Involution is seen in 50% to 65% of patients by age 5 years, in 70% to 80% by age 7 years, and in 90% to 95% by age 9 years. Fewer than 2% of lesions have required further treatment.

Untreated, most patients with hemangiomas have no complications. Minor sequelae of cutaneous hemangiomas include erosion, ulceration, bleeding, and secondary bacterial infection. Areas subject to friction, trauma, or poor hygiene (such as the diaper area) are more
susceptible to ulceration or necrosis, which is seen in 15% to 20% of lesions. Raw, oozing surfaces may be treated with compresses of aluminum acetate solution and mupirocin ointment.

More serious sequelae include ocular sequelae, compression of an important anatomic structure, coagulopathy (Kasabach-Merritt syndrome), high-output cardiac failure, and intracranial extension. The exact rate of serious complications in untreated patients is unknown, but is probably close to 5% to 10%, although some studies quote rates as high as 80%. If a periorbital hemangioma blocks vision, amblyopia leading to irreversible visual impairment ranging up to cortical blindness may result, even after a few ways of visual occlusion; cooperation with a pediatric ophthalmologist is imperative. Periorbital hemangiomas may also extend to the retrobulbar area and compress the optic nerve, causing atrophy. Impingement of the globe may cause astigmatism. Strabismus, eyelid ptosis, and proptosis are other ocular sequelae. Infantile subglottic hemangioma causes narrowing of the airway and inspiratory stridor; without intervention, mortality rates approach 45%. Systemic steroids or CO₂ laser ablation cures around 80% of patients. Lesions involving the lower lip may interfere with feeding, have a higher degree of scarring, and tend not to completely resolve. The Kasabach-Merritt syndrome is the sudden, painful, rapid enlargement of a cavernous hemangioma associated with coagulopathy, thought to be due to platelet trapping and local intravascular coagulation. Cardiac failure may result from large arteriovenous shunts present in some very large hemangiomas; patients in which this is a concern should be seen in cooperation with a pediatrician. Some facial hemangiomas have been associated with intracranial extension with arteriovenous malformations, hypoplasia of cerebral vessels, ventricular dilation, and hemicerebral atrophy. Diagnostic imaging of the head may help delineate the extent of the hemangioma. Disfigurement, although not a functional complication per se, may occur to a degree that merits classification as a serious complication.

The decision for early surgical intervention is often difficult. Intervention occurs most commonly with complicated large hemangiomas. High-dosage systemic steroids have an established benefit in reducing the size or rate of growth of a complicated hemangioma if it is treated during the proliferative phase. The latest literature should be consulted regarding dosage, as recommendations have varied. Repeat treatment is to be avoided or delayed as long as possible so that risks for corticosteroid side effects such as growth retardation are minimized. Intrallesional steroid injections (triamcinolone acetonide 40 mg/mL) have also been a mainstay therapy but their use may be accompanied by rapid necrosis or hemorrhage of the lesion. Treatment of periorbital hemangiomas may be associated with inadvertent embolization of retinal vessels by steroid particles, causing blindness that may be permanent; consultation with a pediatric ophthalmologist may be helpful. Interferon-alpha-2a has been used as adjuvant therapy in severe cases with massive lesions, consumptive coagulopathy, or visceral involvement. Finally, intravascular embolization therapy conducted by an interventional radiologist has proven useful in reducing the size of large, cavernous lesions.

Although most hemangiomas resolve spontaneously, and only 6% of them are considered unsightly, some physicians have recommended a greater role for surgical intervention, specifically laser therapy. There are anecdotal reports that yellow dye or argon laser treatment of hemangiomas within the first weeks of life may arrest or slow the growth of a hemangioma while it is still small. If this proves true, not only will the usual morbidities of hemangiomas be reduced, but also the psychosocial morbidity that all hemangiomas, even
small ones, evoke will also be reduced. The yellow dye laser has been helpful especially in healing ulcerated juvenile hemangiomas without adding to the expected amount of scarring. Long-term studies using the yellow dye laser are in progress. The argon laser may be more useful for lesions with a greater thickness, but it also has a greater risk for scarring. There are also anecdotal reports of the use of contact neodymium:yttrium-aluminum-garnet (Nd:YAG) lasers, but this modality may be associated with short-term necrosis and bleeding, as well as a more unsightly, scarred appearance in the long run.

Children with juvenile hemangiomas may benefit from reconstructive facial plastic surgery late in the evolution of the disease. Lesions that remain unchanged for 3 years are unlikely to undergo further involution. Many patients will have loose, atrophic, redundant tissue at the site of an involuted lesion. This tissue may also be scarred if ulceration or infection have occurred. Reconstructive surgery often proves beneficial to these children, especially if surgery is completed before school age. Half of these children will achieve maximal involution by school age; a dermatologic can help determine the degree of involution. In general, the larger the lesion, the more delayed will be the resolution and the more likely it will never completely resolve.

**Spider Angiomas**

Spider angiomas (nevus araneus) are small arteriovenous fistulae named for their characteristic clinical appearance. Lesions have a central red papule from which several fine telangiectatic vessels radiate, similar to the legs of a spider. Spider angiomas are seen in 30% of children by age 4 years and in 40% by age 8 years. Only 10% to 15% of adults have spider angiomas.

Although most spider angiomas appear in normal children and adults, they may be associated with high estrogen states, such as pregnancy, hepatic insufficiency, and exogenous estrogen use, especially when large numbers of lesions are found on the trunk.

In children, the most common sites of involvement are the face, upper trunk, and upper extremities. Although most lesions resolve, some patients or their parents may seek treatment for cosmetic reasons. Lesions may be treated with gentle electrodessication of the central feeding artery; a small depressed scar may result. The pulsed yellow dye vascular laser will also ablate the lesion, almost always without scarring. The argon or CO₂ lasers are also effective, but scarring has been observed accompanying their use.

**Pyogenic Granuloma**

Pyogenic granulomas are benign vascular proliferations similar to granulation tissue. Although the name, a relic of past beliefs, suggests a bacterial etiology, these lesions are most likely an exaggerated and perpetuated healing response to injury, although the injury is usually not remembered and may have been trivial. Most arise in acral skin such as the distal extremities, face, or oral mucous membranes. Lesions are usually dull red, dome-shaped, or pedunculated nodules that may have a moist and eroded or crusted surface. Bleeding may be the presenting symptom; they may be quite friable. Satellite lesions may be seen, but are not malignant.
Treatment is simply ablative; scalpel saucerization excision or scissors snip excision with 1-mm margins is usually sufficient. Alternatively, many lesions can be shelled out with a dermal curette, leaving an epidermis-lined concave space that will heal without scarring; only the remaining central stalk with its feeding vessel need be fulgurated. All tissue removed should be submitted for pathologic examination since amelanotic melanoma or certain benign entities may resemble pyogenic granuloma. Lesions may recur or persist and are simply retreated. Satellite lesions usually resolve within 6 weeks of treatment of the primary lesion.

**Lymphangiomas**

Lymphangiomas are congenitally lymph vessel hamartomas that usually appear at birth or within the first 2 years of life. Like juvenile hemangiomas, lesions range from a circumscribed, more superficial form, corresponding to the strawberry hemangioma, to a deep, cavernous form.

Lymphangioma circumscriptum is the more superficial form, consisting of multiple gelatinous papules and plaques that tend to be grouped. The appearance is likened to frog spawn. There are often interconnections with the venous system, so some lesions may be hemorrhagic.

Cavernous lymphangiomas are skin-covered nodules with a rubbery consistency. Lesions located in the head and neck region are termed cystic hygromas. Like their hemangioma counterpart, cavernous lymphangiomas may become quite large and disfiguring and may have a rapid growth phase in early infancy.

Treatment of either form of lymphangioma is unsatisfactory. Lesions tend to recur after excision, even with wide margins and repair with myocutaneous flaps. Laser ablation is no more effective than cold steel surgery. Small lesions are most likely to be cured. Supportive therapy includes the use of compressive garments and prophylactic antibiotics to prevent cellulitis and scarring lymphangitis and chronic lymphedema.

**Genetic Diseases**

**Nevoid Basal Cell Carcinoma Syndrome**

The nevoid basal cell carcinoma syndrome (NBCCS) is an autosomal dominant disorder of high penetrance and variable expressivity with numerous cutaneous and extracutaneous findings. Although the primary defect is not known, patients appear to have a defect in DNA repair, especially DNA damaged by irradiation.

The key cutaneous feature of the syndrome is the development of numerous (often hundreds) basal cell carcinomas, most predominantly on the head, neck, thorax, and upper extremities. The carcinomas usually begin to develop in adolescence, but have been reported in young children, where they may resemble skin tags or melanocytic nevi. The lesions are usually translucent and pink or pigmented, but any clinical or histologic pattern of basal cell carcinoma may be seen. Left untreated, lesions may become invasive or metastasize. The incidence of basal cell carcinoma is increased in relation to cumulative sunlight or x-ray irradiation. So-called pitting or pinpoint papules may be seen on the hands and feet, especially
the palms and soles, and are considered by some to be a forme fruste expression of basal cell carcinomas; these lesions, however, never become clinically aggressive. Other cutaneous manifestations include epidermal cysts, milia, comedons, and lipomas.

Extracutaneous features are equally characteristic of the syndrome. Seventy percent of patients have keratinizing odontogenic cysts of the maxilla or mandible, some of which develop into ameloblastoma. About 70% to 75% of patients have skeletal abnormalities, which may include bifid or splayed ribs, scoliosis, spina bifida, and bridging of the sella turcica. Shortening of the fourth metacarpal results in dimpling of the overlying skin, known as Albright's sign. By adulthood, craniofacial changes are apparent, consisting of sunken eyes, frontal and temporal bossing, and a broadened nasal root with true or pseudohypertelorism. Central nervous system abnormalities include calcification of the falx cerebri or falx cerebelli, agenesis of the corpus callosum, and medulloblastoma. Following surgery and radiotherapy of the latter, there is an increased incidence of basal cell carcinomas of skin overlying the radiation ports. There have also been case reports of ovarian fibrosarcoma following radiotherapy to the entire neuraxis for medulloblastoma. Many other abnormalities in these and other organ systems have been reported.

As expressivity of the manifestations of the syndrome is variable, treatment must be tailored to the individual. Patients should protect themselves scrupulously from sunlight and avoid x-ray irradiation if possible. Long-term and short-term goals should be kept in mind when planning excision of multiple lesions. A triage approach to tumor management may prove helpful. The most biologically aggressive lesions should be removed first. Because patients are likely to develop scores of lesions during their lifetime, as much normal surrounding tissue as possible should be preserved. Because of the likelihood of the presence of scars from previous treatments as well as the presence of new basal carcinomas in overlying skin, it may be difficult to determine whether a lesion has recurred in an area reconstructed by flaps or grafts. Curettage and electrodesiccation followed by second intention healing is often the most practical modality of treatment for optimal reduction of long-term morbidity and mortality. Microscopically controlled excision (Mohs surgery) should be considered for treatment of recurrent lesions because of tissue sparing and high cure rates. A more recently described therapy is the use of systemic retinoids to prevent formation of new lesions while the drug is being used. Once the drug is discontinued, however, new carcinomas form as frequently as before. Side effects complicate long-term use of the current generation of retinoids. Photodynamic therapy using lasers following the systemic administration of photosensitizing agents is another promising experimental modality.

**Xeroderma Pigmentosum**

Xeroderma pigmentosum is a group of autosomal recessive genodermatoses in which children manifest an increased number of skin carcinomas of the head and neck, including basal cell carcinomas, squamous cell carcinomas, and malignant melanomas. All of the varieties of xeroderma pigmentosum, which number at least eight, have defective capacity for repair of ultraviolet light-induced DNA damage. In addition, there is a variant of xeroderma pigmentosum in which DNA repair is normal but S-phase DNA synthesis is impaired following ultraviolet irradiation (defective postreplication repair). Like the skin, the ocular epithelium has similar hypersensitivity to ultraviolet irradiation. Some forms of xeroderma pigmentosum have varying neurologic abnormalities.
Xeroderma pigmentosum manifests in some populations more than others, but is found worldwide. In European races, the incidence is 1 per 250,000 births, and in Japan the incidence is 1 per 40,000 births.

Onset is congenital; photophobia in infancy is usually the first manifestation. Infants exposed to ultraviolet light, either from natural or, to a lesser extent, artificial sources, will begin to develop dose-related conjunctivitis and sunburn-like erythema and scaling of the skin as evidence of ultraviolet-induced damage. If the acute dose of light is strong enough, blistering may be seen. As damage progresses, freckling and poikiloderma (hyperpigmentation, hypopigmentation, telangiectasia, atrophy, and scarring) of the skin will form and the ocular conjunctiva will become xerotic, hyperpigmented, and scarred. Symblepharon and blepharitis are also seen. As damage accumulates, the skin forms actinic keratoses. Frank carcinomas subsequently develop, usually by the age of 6 years. Basal cell carcinomas, squamous cell carcinomas, and malignant melanomas, in order of decreasing frequency, are commonly seen. Other skin tumors include keratoacanthomas, fibrous tumors, and sarcomas. The conjunctivae and corneas become clouded, hypervascularized, and scarred, and develop malignancies. Vision is correspondingly impaired. Some patients, such as those with the De Sanctis-Cacchione syndrome, also have progressively worsening neurologic abnormalities including mental retardation, cerebellar ataxia, and seizures. Some patients have developmental abnormalities such as microcephaly and dwarfism.

In the past, death usually occurred by the third decade. Patients are now living longer due to better protection from ultraviolet light and better medical care. Strict avoidance of both natural and artificial sources of ultraviolet light have forestalled the progressive damage to eyes and skin. Patients with neurologic abnormalities, however, face progression of neurologic disease despite strict protection from ultraviolet irradiation.

Patients should be monitored closely for the development of cutaneous malignancy, with prompt and judicious biopsy performed on suspicious lesions to allow the earliest possible treatment of cancer. Skillful use of cryosurgery or topical 5-fluorouracil on premalignant lesions such as actinic keratoses or lentigo maligna will help forestall the inevitable onset of malignancy. Use of systemic retinoids such as isotretinoin will also slow the formation of carcinomas, but the effect does not last after the patients discontinue the drug, and long-term side effects may be expected.

Tuberous Sclerosis

Tuberous sclerosis is an autosomal-dominant disease with a wide variety of cutaneous and internal findings. The classic triad is epilepsy, mental retardation, and "adenoma sebaceum", which is a misnomer, since these lesions are actually cutaneous angiofibromas. Angiofibromas are seen in 65% to 95% of patients, almost always over the cheeks, nasolabial folds, and chin; more rarely, they are seen elsewhere on the head. Angiofibromas are shiny, translucent papules usually millimeters in size, with a yellowish-pink to brownish-red hue. Patients typically have dozens of lesions. Some patients have fibrous plaques of the face that lack a significant vascular component.

Patients may have other cutaneous findings besides angiofibromas. The most common are "ash-leaf" macules, which are oval-shaped hypopigmented macules seen on the trunk:
"shagreen" patches, which are slightly raised, slightly textures, leather-like plaques seen on the trunk; periungual fibromas, which are firm, budlike growths around the nail plates, sometimes termed Koenen's tumors. Visceral involvement includes potato-shaped (tuberosous) or flat, sclerotic plaques of the cerebral cortex, cardiac rhabdomyomas, renal hamartomas, retinal hamartomas, and a variety of other ocular tumors.

There are many methods for management of angiofibromas of the face. Lesions may be removed by CO$_2$ laser vaporization, tangential excision, dermabrasion, electrodesiccation, or cryosurgical ablation. Further surgery may be required at intervals, as new lesions continue to form.

Miscellaneous Cutaneous Tumors

Epidermal Cysts

The most common superficial tumor seen in childhood is the epidermal cyst (epithelial cyst; epidermoid cyst). The term sebaceous cyst is a misnomer, since the epithelial lining of the cyst arises from epidermis identical to the infundibulum of the hair follicle, and not sebaceous glands. The cysts are dermal or subcutaneous, and are filled with laminated keratin normally shed from the surface of the skin. Most epidermal cysts arise spontaneously, but rarely may arise traumatically if epidermis inverts into the skin; this variant is termed an epidermal inclusion cyst.

Clinically, epidermal cysts are firm, spherical nodules that vary in size from millimeters to centimeters; most are less than 2 cm in diameter. In children, the most common location is the lateral eyebrow or elsewhere in the periorbital area. Cysts may be superinfected, in which case inflammation with redness and tenderness is seen. Multiple cysts in children should raise the suspicion for Gardner's syndrome, an autosomal dominant disorder in which premalignant intestinal polyposis, muscular desmoid tumors, and jaw osteomas may also be seen.

Epidermal cysts require no therapy as such, as they are harmless. They occasionally regress spontaneously, but may also be subject to repeated infection with cyclical enlargement and drainage. Removal of the entire cyst wall is curative. Cysts may be excised en bloc, but removal through the punctum of the cyst or a small slit incision is often possible and reduces the size of the scar. Using this method, a 2- to 4-mm incision is made over the cyst; if the punctum is large, a tiny 2- to 4-mm ellipse incision is made to include the punctum. The cyst contents are then decompressed, if necessary, and the cyst wall is everted through the opening. Allaying subjacent pressure, scooping out the cyst wall with a small curette, and teasing out the cyst wall using steady traction with forceps are helpful. Removing the cyst wall in this fashion is akin to everting a balloon through its own neck. If the cyst has been subject to repeated infection, adherent scarring around the cyst wall may make this conservative approach impossible. Regardless of the method used, rupture of the cyst during removal has not effect on recurrence. It is retention of any portion of the cyst wall, including its neck or punctum, that causes recurrence.
Dermoid Cysts

Dermoid cysts are round intradermal or subcutaneous nodules most commonly seen on the face or scalp. They are thought to result from the invagination of ectoderm at embryonic fusion planes. Thus, they represent developmental anomalies. Unlike epidermal cysts, which are formed of epidermis alone, dermoid cysts contain epidermis, hair follicle structures, sebaceous glands, and apocrine glands, and thus may form keratin, hair, sebum, and apocrine secretions. The lateral eyebrow is a site of predilection. Lesions located over the midline, such as the nose, glabella, and skull, as well as over skull sutures, should be approached with caution, as they may have sinus tracts that may extend deeply. A typical presentation of such a cyst is a nodule over the nose or glabella that has hairs protruding from a punctum. Some dermoid cysts even connect with the meninges. Magnetic resonance or computed tomography scans should be considered in the preoperative evaluation of midline cysts with neurosurgical consultation as necessary.

Epidermal Nevi

Epidermal nevi are hamartomas that may form from any of the components of skin and its adnexal structures, including keratinocytes, hair follicle cells, sebaceous glands, or sweat glands. Combinations of these cutaneous structures with a variety of reactive patterns such as inflammation, hyperkeratosis, or papillomatosis result in a variety of histologic and clinical patterns. Thus, epidermal nevi may clinically or histologically resemble many other proliferative disorders of the skin including warts, acanthosis nigricans, Darier's disease, ichthyoses, or psoriasis.

For the most part, epidermal nevi are not genetic in etiology and appear sporadically. Although the lesions are composed of epithelium, there appears to be a component of interaction with other tissues of ectodermal as well as mesodermal origin, since patients may also have other developmental abnormalities; permanent destruction of lesions seems to require ablation of underlying papillary (superficial) dermis.

The nosology of epidermal nevi is inconsistent; most lesions are described by clinical features, and there is overlap. The nevus sebaceus (of Jadassohn) will be discussed separately.

The verrucous epidermal nevus is warty, with color ranging from pink to gray to brown. Lesions often have a dirty appearance due to hyperpigmentation and hyperkeratosis. Age of onset is usually at birth or in the first decade, but may range up to the third decade.

Nevus unius lateris is a widespread, generally linear, epidermal nevus that usually appears on the trunk or extremities. The configuration is usually longitudinal on the limbs and transverse on the trunk; Blaschko's lines refer to the pattern these lesions usually follow. Lesions are usually unilateral, stopping at the midsagittal plane. Bilateral distribution is sometimes called ichthyosis hystrix, but this term may be confused with varieties of ichthyosis and should be avoided.

Inflammatory linear verrucous epidermal nevus (ILVEN) refers to a pruritic, inflammatory lesion with a generally later onset. It is usually seen on the extremities in girls.
The epidermal nevus syndrome is a severe condition in which widespread epidermal nevi are seen along with other developmental defects of varying severity. Craniofacial or other skeletal defects involving the spine and extremities occur in two-thirds of patients. Abnormalities of the central nervous system, including seizures and mental retardation, are seen in half of patients. Ocular abnormalities, including extension of the nevus onto the eyelids, ocular dermolioid tumors, and colobomas of the lid, iris, or retina, are seen in one-third of patients. Besides ocular involvement, other challenges for the facial plastic surgeon include widespread involvement of the ear or other facial structures.

Other dermatologic conditions seen in patients with the epidermal nevus syndrome include vascular lesions (eg, nevus flammeus or juvenile hemangiomas), giant congenital melanocytic nevi, and keratoacanthomas. Some patients developed carcinomas in the epidermal nevi, especially those with the nevus sebaceus form of the syndrome.

Treatment, if any, for epidermal nevi depends on the symptomatology, and extent and distribution of the lesions. Topical therapy with tretinoin, aquaglycolic acids, or 5-fluorouracil may be sufficient for some patients, although it is not curative. Simple excision and closure may be practical for smaller lesions. Widespread plaques may be treated with deep tangential excision, electrosurgery, cryosurgery, dermabrasion, or CO₂ laser vaporization; all of these modalities require healing by second intention.

Nevus Sebaceus

The nevus sebaceus (organoid nevus; nevus sebaceus of Jadassohn) is a hairless, raised, yellowish to yellowish-brown plaque most commonly seen on the scalp or face. It is generally present from birth, and grows at the same relative rate as the child. The lesion is comprised of abnormally developed apocrine and sebaceous glands with papillomatosis of the superficial dermis and epidermis. Under hormonal influence at puberty, the glandular elements will proliferate, and the lesion assumes a more thickened, verrucous and papillomatous texture. In one-third of patients, benign or malignant neoplasia will develop. The most common tumor is syringocystadenoma papilliferum, a benign tumor of apocrine origin, seen in 12% to 20% of lesions. A wide variety of other benign adnexal tumors may be seen.

Basal cell carcinoma (BCC), seen in 6% to 16% of lesions, is the most common malignant growth in nevus sebaceous. Fortunately, BCC in this setting is slow-growing and is usually seen before the fourth decade of life. Other malignancies are also reported, some of which have grown aggressively.

Neoplasia in nevus sebaceous is unusual before puberty, so surgical excision can usually be delayed until then unless concerns regarding cosmesis supervene. Since the lesion is benign, narrow margins of excision may be used, with wider excision or microscopically controlled excision (Mohs surgery) being used only if pathologic evaluation discloses malignancy.

Adnexal Tumors

The epidermal adnexae include the hair follicles, sebaceous glands, apocrine sweat glands, and eccrine sweat glands. The first three structures are continuous with the hair
follicle, whereas the eccrine sweat gland empties directly to the skin surface. Some benign neoplasms derived from these structures are discussed elsewhere, eg, the epidermal nevus and nevus sebaceus. Adnexal tumors are more common in adults, but three types, besides epidermal nevi and nevus sebaceus, are more prevalent than other in children or adolescens: pilomatrixomas, trichoepitheliomas, and syringomas.

**Pilomatrixomas**

Pilomatrixomas (calcifying epithelioma of Malherbe) are hard, cutaneous nodules with normal overlying skin. Lesions are usually solitary and arise on the face or limbs. Onset may range from birth, but infancy or early childhood is the usual time of onset. Rarely, lesions are associated in a syndrome with myotonic dystrophy. Lesions measure 2 to 5 mm in size, and arise from cells of hair matrix. Calcification may occur. Lesions may be removed by excision; they usually do not recur.

**Trichoepitheliomas**

Trichoepitheliomas, when multiple, are the result of an autosomal dominant trait. They may also be found in a nonhereditary form as single lesions. Lesions are round, usually measure up to a few millimeters in diameter, are flesh-colored to reddish brown, and are primarily found in the midface around the nose or elsewhere on the head and neck. Onset is usually in adolescence. Histologically, they are comprised of horn cysts and islands of basaloid cells, probably of hair follicle origin. Solitary lesions may be confused clinically or histopathologically with basal cell carcinoma. When the lesions are multiple, with new lesions arising continually, cosmetically acceptable treatment is difficult. Tangential excision, dermabrasion, or CO₂ laser vaporization may be applied, similarly to the treatment of angiofibromas of tuberous sclerosis.

**Syringomas**

Syringomas are derived from sweat gland epithelium. Clinically, they present as small, flat, waxy or translucent papules around the eyes, usually on the eyelids or in an infraorbital or zygomatic distribution. Lesions may also appear on the upper trunk or vulva; there is a female preponderance. Papules are skin-colored with hues of yellow or brown. Onset is usually around adolescence. Since the nests of eccrine epithelium deeply infiltrate the dermis and are not well circumscribed, ablation of the lesions short of full dermal excision is difficult. Lesions are usually too numerous to excise. Sometimes patients are satisfied with tangential or saucerization excision of a few of the larger lesions, with the expectation that new and recurrent lesions will arise and that cosmesis will not be perfect. Laser surgery, including the CO₂ laser, is also used, as well as electrosurgical ablation.

**Juvenile Xanthogranulomas**

Juvenile xanthogranulomas are soft, orange to yellowish-brown nodules appearing on the skin and other tissues, including internal organs and the iris. They usually arise before 1 year of age, are found on the scalp, face, and neck, and occasionally elsewhere, and are often multiple. Although histologically the lesions are comprised of lipid-filled granuloma cells, there is no relation to abnormalities of lipid metabolism. The process is idiopathic and benign,
and lesions usually resolve within 1 year, occasionally requiring several years for complete involution. No treatment is necessary for skin lesions. Biopsy of a single lesion for diagnosis and referral for ophthalmologic evaluation may be indicated. Lesions may be confused with Spitz nevi.
Surgical correction of bony deformities of the maxillofacial region is well established in the skeletally adult population. The timing and sequence of procedures in the growing child are more controversial, since it raises questions as to the effect of surgery on later growth of not only the specific bones that were osteotomized, but also of the maxillofacial complex as a whole. Data is often limited, particularly in terms of long-term follow-up of significant numbers of patients.

Mandibular advancement for example, may be successfully carried out in growing children with true mandibular retrognathia. There seems to be reasonable stability of the maxillomandibular relationship, with both the maxilla and mandible exhibiting harmonious growth. However, treatment of mandibular prognathism during growth has a high rate of relapse and is thus usually done after cessation of growth. In contrast, Le Fort III osteotomy in the growing child does not seem to affect subsequent facial growth, either adversely or favorably.

Of considerable concern is the effect of Le Fort I osteotomies on midfacial or nasal growth. Even groups that have performed "early" (ages 10 to 14) Le Fort osteotomies for vertical maxillary excess advocate consideration of procedures that avoid significant septal resection. The position of unerupted teeth may force alteration of the osteotomy lines. Most surgeons currently advocate waiting until mid- to late adolescence before performing a Le Fort I osteotomy.

Midface osteotomies most likely "do not enhance growth of the underdeveloped midface". If they are done, some overcorrection is thought to be necessary. In general, most orthognathic surgery is still done after cessation of growth. However, the evaluation of and preoperative management with orthodontics is begun earlier.

This chapter presents an outline of common maxillofacial deformities, stressing evaluation, timing considerations, and treatment modalities, with particular attention to how these modalities must be modified for the growing child.

Common Maxillofacial Deformities

The most common maxillofacial deformities in children are mandibular hypoplasia and mandibular hyperplasia. Maxillary size discrepancies are also quite common. These may result in maxillary hypoplasia, hyperplasia, or width discrepancies. Excessive vertical growth of the maxilla also occurs commonly, giving the patient a "gummy" smile. Associated open bite deformities may be quite dramatic and can result in significant speech-related difficulties. These discrepancies may be present in early childhood, but usually become more of a concern after puberty and its associated rapid growth.
Asymmetric facial growth in the nonsyndrome patient most commonly occurs secondary to asymmetric mandibular development. This is generally due to unilateral hyperplasia or hypoplasia of the condyle or ramus. If these occur during somatic growth, there is usually concomitant maxillary compensation, resulting in a broader maxillofacial asymmetry. As in all cases of maxillofacial discrepancies, a thorough evaluation must be made to determine if the deformity is part of a syndrome such as hemifacial microsomia. Sequential studies are usually needed to determine that the asymmetric growth has ceased. These may include asymmetry analysis of a posteroanterior (PA) cephalometric radiograph, lateral cephalometric study, models of dentition, and bone scans to assess differential activity.

Soft tissue changes following orthognathic surgery tend to be only broadly predictable. Most studies show significant individual variation from the mean. This will be elaborated upon in the section on treatment planning and predictions. Variation is compounded when dealing with the growing child who will have not only more bony growth, but also soft tissue modifications associated with secondary sexual changes. In general, secondary soft tissue alterations following osteotomies should be delayed for at least 9 to 12 months.

Special Considerations

Congenital cleft lip and palate deformities represent a significant number of patients requiring secondary maxillofacial surgery. Because of the complexity of this subject as it relates to oral and maxillofacial procedures, this is covered in Chapter 16 by Kuo, Will, and Albert.

Evaluation and Treatment Planning

General Considerations

Volumes have been written in the surgical and orthodontic literature concerning hard and soft tissue evaluation of the maxillofacial region. We will concentrate on the relatively common and simple evaluation methods that usually define most of the problem. Of course, no technical evaluation can replace a careful history, listening to the patient's perception of the problem, and clinical experience. The technical data helps to define the details. Ideally the problem must be evaluated in lateral, anteroposterior (AP), and transverse dimensions.

The most common diagnostic workups for dentofacial deformities include lateral cephalometric radiographs and analysis for both hard and soft tissue, dental radiographs and analysis for both hard and soft tissue, dental radiographs, occlusal evaluation with dental models, and aesthetic facial analysis. These are usually subdivided further to look specifically at the upper, middle, and lower thirds of the face.

Cephalometric Analysis

Cephalometry, particularly the lateral view, is one of the most commonly used diagnostic tools. It allows for reasonable, objective, and reproducible analysis pre- and postoperatively and lends itself to prediction tracing. Most studies of orthognathic surgery have relied heavily on this analysis. A large number of different systems for cephalometric analysis have been devised, the details of which are well beyond the scope of this chapter.
The common features are the bony landmarks of the nose, midface, mandible, and dentition, and their relationships to consistent bony landmarks of the normal skull and to each other. Normative data have been generated. It is important to note that most norms have come from Caucasian or European groups. Anteroposterior, transverse, as well as vertical dimension, can be measured.

A properly exposed cephalometric radiograph will also include the outline of the soft tissues. These can also be analyzed by tracing out the soft tissue contours. Prediction tracings can be generated for any particular procedure. Hard tissue changes are much more predictable than soft tissue, particularly in the adult or nongrowing patient. As discussed previously, relative relationships of bony landmarks in procedures such as mandibular advancement may remain stable; however, absolute positions will change with growth.

Computerized imaging has expanded greatly in the last several years and includes both hard and soft tissue analysis. Video imaging allows for creation of possible soft tissue changes that can be readily visualized by the patient. However, it is important to emphasize again that actual soft tissue changes show a wide individual variation for most parameters. Caution must be used in implying that the actual end result will match the video image.

Orthodontic/Dental Evaluation

Examination of the individual teeth as well as their relationship to one another is critical. Today well over 95% of orthognathic surgery cases are done in combination with orthodontics. Some significant soft tissue changes can be achieved by orthodontic manipulation only. However, changes in tooth position are limited by bony position, ie, the teeth must be moved within the bone itself. Trying to compensate for a true bony disharmony with orthodontics alone gives a less than satisfactory result.

Transverse discrepancies manifested as maxillary constriction are most readily analyzed by examining the dental casts and a PA cephalometric radiograph. In the growing child, rapid palatal expansion reliably produces widening by taking advantage of the nonfused, midpalatal suture. This can usually be achieved with an orthodontic appliance over a period of 2 to 8 weeks. The device can be activated by as much as 0.5 mm per day. Care must be taken to make sure the maxilla itself is expanding at the palatal suture, rather than the teeth being tipped and/or extruded. In rare circumstances, surgically assisted rapid palatal expansion is needed in the older child. This involves performing lateral maxillary corticotomies, usually on an outpatient basis, before activating the expansion device.

The goal of orthodontic evaluation is to determine what is necessary orthodontically to align the arches and allow the teeth to interdigitate after osteotomies are performed. Close consultation between the orthodontist and the surgeon is required.

Timing Considerations

Timing and sequencing of maxillofacial orthognathic surgery is still one of the most controversial questions. As discussed earlier, in the adult population where most of the data exist, there is wide individual variation in the correlation of hard tissue and soft tissue changes. In the growing child there is the additional concern that osteotomies may affect the
growth potential of the particular bone itself or the contiguous bones. Excluding patients with craniofacial syndromes, most patients come to the maxillofacial surgeon's attention in early adolescence. This is probably due to a combination of factors. Most orthodontic evaluation and treatment begins about this time. Major discrepancies elicit more peer pressure for possible change. More permanent teeth are erupted, which allows for better orthodontic evaluation and treatment. The majority of maxillary growth is achieved during this period. Patient cooperation tends to improve as the child becomes older. Technical concerns such as tooth bud position and bony anatomy that may limit the osteotomy tend to diminish toward the end of growth.

**Mandibular Deformities**

Mandibular hypoplasia is usually defined both cephalometrically and dentally. Cephalometric analysis will show the mandible to have below normal AP length in relationship to the cranial base with a normal maxillary AP dimension. Dentally a class II molar relationship exists with the mesiobuccal cusp of the maxillary first molar anterior to the buccal groove of the mandibular first molar.

Vertical height of the mandible, as well as the prominence of the chin button or menton, must be assessed. A retrusive menton will accentuate the aesthetic aspects of the retrusive mandible. As discussed earlier in this chapter, the limited data available concerning mandibular lengthening in the normal, growing child (ages 8 to 12) seem to indicate no adverse effect upon later mandibular growth or upon the maxillofacial complex.

The sagittal split osteotomy is the most common technique utilized for mandibular advancement. This may be used in combination with chin advancement.

Special considerations in the growing child include position of the developing tooth buds of the second and third molars and location of the lingula of the mandibular foramen. The second and third molar tooth buds are often high and lateral, near the buccal cortex, where the vertical cut is usually made. A more posterior buccal cut may significantly reduce the amount of overlapping bone. The tooth bud position is also a consideration in interosseous wire placement or in the use of rigid fixation with plates and screws.

The lingula of the mandibular foramen is located more posteriorly and sometimes more superiorly on the ramus of the growing child. This usually necessitates the medial horizontal cut being made to the posterior border. In addition, there must be adequate bone superior to the cut to allow for coronoid continuity in the proximal segment. Getting the mandibular nerve to stay in the distal segment can be more difficult in the younger patient as the nerve is more central in the mandible.

One of the major potential complications of the sagittal split osteotomy is paresthesia or anesthesia of the mandibular nerve. No studies have been done to show the effect of partial loss of function of the third division on muscle and bone growth. Observations of congenital nerve loss indicate the potential for underdevelopment. A minor complication is that most osteotomies require some period of maxillomandibular fixation. Facial contour changes following mandibular advancement can be dramatic. Much of this is secondary to chin position changes. In patients with retrusive chins, simultaneous chin advancements can be
considered. Stability of sagittal split advancements is correlated with magnitude of advancement. The greater the advancement the greater the likelihood of partial relapse.

Fixation for mandibular osteotomies is usually twofold. The osteotomy site itself may be fixed with wire osteosynthesis, lag screws, or miniplates. Some controversy exists as to the best method. Wire osteosynthesis either at the superior or inferior border requires the use of maxillomandibular fixation for 4 to 6 weeks. Rigid fixation may eliminate or reduce the need for maxillomandibular fixation; however, it may increase the risk of secondary temporomandibular joint changes and nerve injury. An extraoral approach with a trocar is sometimes necessary when applying plates and screws, and carries a small risk of facial nerve injury.

Mandibular Retrognathia and Vertical Ramus Deficiency

A less common finding is that of mandibular retrognathia and vertical ramus deficiency. This is most commonly found in cases of hemifacial microsomia and Goldenhar's syndrome. Vertical osteotomies can be used to lengthen the ramus without advancing the mandible. Inverted L osteotomies with an interpositional bone graft are used to both lengthen the ramus and advance the mandible.

Mandibular Prognathism

True mandibular prognathism is present when the mandible is longer than a normally sized maxilla. Relative mandibular prognathism exists when the mandible is normal length and the maxilla is found to have a diminished anteroposterior dimension as measured cephalometrically.

For true mandibular prognathism the mandible is set back either with a sagittal split osteotomy (with ostectomies) or with a vertical oblique ramus osteotomy. The vertical oblique osteotomy may be modified to preserve coronoid position, or a coronoidectomy may be performed.

Timing considerations are important because the mandible usually continues to grow until the cessation of somatic growth. Treatment before the end of growth usually results in partial relapse requiring a second surgery, and is considered only for extreme cases of mandibular prognathism.

Mandibular growth after the cessation of somatic growth is often associated with a growth hormone secreting pituitary adenoma or with condylar hyperplasia, which may be unilateral or bilateral. Once the etiology has been determined and treated, mandibular osteotomies may be employed.

Genioplasty

Alteration of the chin position via a horizontal osteotomy may be accomplished as a separate procedure or at the same time as a mandibular ramus osteotomy. The bony position is evaluated with a lateral cephalometric radiograph. Chin implants may also be considered. No studies have been done as to the effect of a genioplasty alone on mandibular growth in
In a growing child.

In cases of mandibular asymmetry, the chin position itself is often not in line with the dental midline. A lateral shift should be considered in these cases.

Maxillary Deformities

In the nonsyndrome child, maxillary size and position show wide variation in the most commonly used measurements of vertical height, horizontal position, and width. Washburn et al in their extrapolation of data from multiple studies in facial growth, showed that, on average, most maxillary growth is completed by the time of canine eruption or between the ages of 10 and 13. The different studies, however, showed wide variations among individuals, as well as between the sexes, with boys generally reaching the 95th percentile several years later than girls. Although Washburn et al reported no particular adverse effects of Le Fort I osteotomy for correction of vertical maxillary excess, most of the patients were operated on after the majority of maxillary growth was complete. In addition they recommended using a complete alveolar osteotomy in order to avoid potential facial growth disturbances secondary to resecting the nasal septum. This particular area of concern (effect of resection of nasal cartilage on subsequent facial growth) is not well defined in research or clinical studies.

Timing is also affected by technical considerations such as tooth bud position. Sailer has shown that the buds of developing teeth in young children are positioned nearly at the infraorbital rim. Developing molars are high in the posterior wall of the developing sinus. Kaban, in taking all of these factors into consideration, states, "Le Fort I osteotomy should only be performed after the permanent teeth are fully erupted." The exception to this would be in regard to third molar (wisdom tooth) eruption.

Significant controversy still exists as to whether nasal airway obstruction leads to vertical maxillary excess (VME). There have been no definitive studies to show any indication that early correction of nasal airway obstruction in patients with VME would lead to a diminution of their VME.

The relationship of VME (or maxillary deformities in general) to mandibular growth and position and open bite deformities is also inconclusive. West, in his review of the pertinent literature, found a multiplicity of factors and findings in this regard. Most vertical maxillary dysplasias are dentoalveolar in origin, with the palate varying little in its vertical position. Thus, much of the surgical approach must focus on changes in dentoalveolar position. Depending upon mandibular position, correction of maxillary deformities often requires mandibular osteotomies as well.

Maxillary Evaluation

Lateral skeletal cephalometric analysis is the mainstay in determining horizontal and vertical maxillary position. Normative data must always be reviewed in light of the individual. Width analysis is mainly on the basis of dental casts and occlusal analysis, although this can be misleading if there is a great deal of tipping of teeth. A posteroanterior cephalometric radiograph allows for more reliable bony measurements of width.
Soft tissue evaluation is currently done mainly from lateral soft tissue outlines. Computerized analysis may provide for better three-dimensional evaluation in the future. The real challenge is in the predictability of soft tissue changes secondary to hard tissue movements. So many factors seem to be involved, with so much individual variation, that the conclusion one draws is that there is more art than science in soft tissue predictability. What is known is that changes will occur in absolute and relative lip length, nasal tip, and alar base morphology, and in the commissure of the mouth. Significant patient dissatisfaction can arise if absolute predictions are made. Secondary revisions often need to be considered.

**Maxillary Procedures**

The Le Fort I osteotomy is the most common technique for changing maxillary position. The basic technique may be modified for individual reasons. The maxilla may be divided into multiple pieces to compensate for width or vertical discrepancies. In most cases it is preferable to deal with these orthodontically preoperatively, as division into multiple pieces increases the risk of avascular necrosis and nonunion or periodontal problems.

In patients with infraorbital hypoplasia, a "high" Le Fort I osteotomy may serve to fill out this area. Here the anterior level of the cut is raised. The infraorbital nerve may have to be dissected out and repositioned.

Cases of severe VME may cause significant impingement on the nasal septum and turbinates as the maxilla is shortened in its vertical dimension. A separation of the alveolus from the palate may be a necessary modification along with turbinectomy or turbinotomy.

The alar base is usually widened in vertical shortening of the maxilla. Bone grafting may be necessary in cases where gaps greater than 5 mm occur either vertically, as in vertical elongation, or horizontally, as in an impaction and/or advancement. Rigid fixation is preferred for virtually all Le Fort procedures. It is particularly helpful in vertical elongation. Bone grafts are often fixed to the plates as well. With use of rigid fixation, maxillary advancements rarely require posterior bone grafts. Le Fort osteotomies in patients with cleft deformities require special considerations and is covered in Chapter 16 by Kuo, Will, and Albert.

**Asymmetry**

Asymmetric facial growth may occur for a variety of reasons, either congenital or acquired. The most common congenital etiology is hemifacial microsomia, which has a variable, progressive presentation. It involves soft tissue, skeletal, and neuromuscular aspects of the face. Secondary mandibular and midfacial deformities develop routinely. Reports have shown that intervention during growth can enhance potential growth of the mandible and decrease or prevent secondary deformities.

Acquired abnormalities that can produce asymmetric growth include condylar hyperplasia, trauma, radiation therapy, tumors, juvenile rheumatoid arthritis, and infections. Developing a rational treatment plan is dependent upon determining the etiology. Appropriate studies such as radiographs, bone scans, and orthodontics evaluation are therefore necessary. Symmetry analysis is usually done in the AP mode.
In most cases of asymmetric growth occurring during childhood or before the end of somatic growth, there are compensatory changes in surrounding structures. For example, in cases of condylar hyperplasia, the maxilla will also grow asymmetrically. Treatment must take this into account. It is therefore common to perform corrective orthognathic surgery in both jaws.

In the noncongenital or acquired asymmetries, it is important, after determining the etiology, to also determine that the process has stopped or been corrected without recurrence before proceeding with corrective orthognathic surgery. This usually requires serial bone scans, symmetry analysis, orthodontic models, and cephalometric radiographs.

In general, the key to a satisfactory aesthetic result is to establish the correct maxillary position and then set the mandible to it. Often, the maxilla must be leveled to obtain a normal horizontal occlusal plane. This is facilitated by the use of rigid fixation with miniplates. Large gaps may have to be bone grafted. Preoperative orthodontics is usually required.

Mandibular alignment usually requires asymmetric movement. If the mandible is deviated to the right for example, it may have to be lengthened on the right and retruded or shortened on the left. Even if one side does not require significant lengthening or shortening it is usually advisable to perform an osteotomy to allow for rotation around a vertical axis in the ramus. Attention must also be paid to the width of the mandible, particularly in the posterior. Onlay grafts may be used, often as a secondary procedure.
Pediatric Facial Plastic and Reconstructive Surgery

James D Smith, Robert M Bumsted

Chapter 24: Anesthesia for Pediatric Facial Plastic and Reconstructive Surgery

Harry G G Kingston

The infant with upper airway abnormalities may have unique management problems that are challenging to an anesthesiologist. This chapter describes how anesthesiologists approach these difficulties, and stresses the importance of having a team approach to the management of the pediatric patient.

Anatomical and Physiological Differences Between the Airways of Adults and Children

The neonatal airway differs from that of an adult in a number of important aspects. The tongue is relatively large, the oral aperture small, and the large head is often difficult to stabilize when attempting tracheal intubation. The larynx is more anterior and cranial than that of an adult, the epiglottis is large and leaf-like, and the cricoid cartilage forms the narrowest part of the pediatric airway. A small amount of edema at the cricoid ring can significantly reduce the diameter of the airway, resulting in turbulent gas flow. It can be demonstrated that a 50% reduction in airway radius will increase the pressure gradient required to maintain the same gas flow by a factor of 32! Care should be taken therefore to insure that an appropriate peritubular leak is present after placement of an endotracheal tube, since cricoid edema may result in a large increase in the work of breathing in the immediate postoperative period. This could be critical when trying to assess the wisdom of early extubation relative to the airway compromise that may be present as a result of the surgical procedure.

A child has a small functional residual capacity (FRC), which means that his/her pulmonary oxygen reserves are low. Since the child's oxygen uptake is more than twice that of an adult, this, together with the reduced FRC, results in the rapid occurrence of hypoxia if apnea occurs at any point in the anesthetic management. To satisfy this high oxygen requirement, a child must maintain a high minute ventilation in comparison with an adult, and any sedative agent that depresses respiration will again result in a rapid deterioration in respiratory function.

Children have a high cardiac output in order to meet the increased metabolic requirements of growing tissues. Although they have relatively faster heart rates, they show considerable preponderance of parasympathetic nervous system activity, which results in their responding to stress by a rapid decrease in heart rate rather than the more familiar increase seen in adults. All clinicians who work with anesthetized children are familiar with the sudden decrease in pulse rate and tone of the pulse oximeter that heralds impending trouble!

Another important physiological difference of neonates that complicates their operating room management is the way in which they maintain their body temperature relative to their adult counterparts. A small child has a large body surface area relative to its weight, making him/her vulnerable to hypothermia if prolonged exposure occurs. An adult shivers, dresses
warmly, and complains about his discomfort. A neonate does none of these, but increases the metabolism of "brown fat", producing an exothermic reaction capable of maintaining body temperature for a short while. This is expensive in terms of oxygen needs. There needs to be an increased level of awareness of this when babies are anesthetized in an adult operating room environment, and every effort must be made to carefully regulate the room temperature as well as carefully monitor the child's temperature throughout the case.

**Pharmacological Differences**

There are important pharmacological differences between adults and children with which the surgeon should be familiar, as they become very evident in the operating room. Because of their high minute ventilation (among other things), children have a more rapid inhalation induction of anaesthesia than adults. Although this may well be an advantage, one should always be aware that if the anesthetic agent is left at "induction levels" for a prolonged period of time while the anesthesiologist attempts to intubate the child or place an intravenous infusion, dangerous levels of hypotension can occur. In addition, the distribution of total body water within the intra- and extracellular fluid compartments of children is different from that in an adult in that the compartment into which a drug is distributed may be greater than that of an adult, and may require and adjustment of the drug dosage. Excretory mechanisms show varying degrees of functional maturity, and protein binding of drugs may differ widely from those of an adult, resulting in more free drug being available to produce its desired pharmacological effect. The way in which a child responds to pharmacological agents may not always be predictable or even comparable to the response seen in an adult.

**Challenging Abnormalities Common to Children Requiring Facial Reconstructive Surgery**

Conditions requiring facial plastic and reconstructive surgery may be challenging to the anesthesiologist because of the potential hazards associated with attempted endotracheal intubation. These can be thought of as anomalies involving the cranium, facial skeleton, cervical spine, and the upper airway. A brief summary of some of these, including the reason for their being an anesthetic challenge, is presented in Table 1.

**Assessment of the Child for Anesthesia**

**Common Pediatric Anesthetic Problems**

**Runny Nose**

A decision has to be made as to whether the child has an infectious or noninfectious cause for the nasal discharge. If it is noninfectious and is either allergic or vasomotor, anesthesia can proceed without increased risk. "Infectious" runny noses need to be assessed in more detail and consideration given to whether this is an upper or lower respiratory tract infection. Usually this can be established by a thorough physical examination, taking the child's temperature, and, if necessary, sending off for a white cell count. A child with upper respiratory tract infection will be normal within a few days to a week with no pulmonary sequelae, whereas children with lower respiratory infection should not be electively anesthetized for a few weeks after the infection.
Apnea and the Preterm Infant

Premature infants have been shown to have a high incidence of respiratory problems postoperatively. Each anaesthesia department should have well-publicized guidelines as to when infants are no longer at risk from postoperative apnea. Controversy still exists as to the age at which these children cease to be at risk. Expert opinions range from 44 weeks of gestational age to as late as 60 weeks of gestational age. In our institution we have taken 50 weeks of gestational age to be the age below which we recommend hospital admission, and careful apnea/saturation monitoring is recommended for a period of 24 hr following surgery.

Table 1. Conditions requiring facial surgery under anesthesia

<table>
<thead>
<tr>
<th>Name</th>
<th>Clinical features</th>
<th>Anesthetic challenge</th>
</tr>
</thead>
<tbody>
<tr>
<td>Craniofacial anomalies</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Crouzon syndrome</td>
<td>Variety of skull shapes</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>Hypoplastic maxilla</td>
<td>Raised intracranial pressure</td>
</tr>
<tr>
<td></td>
<td>High arched palate</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hypertelorism</td>
<td></td>
</tr>
<tr>
<td>Apert syndrome</td>
<td>Sphenoethmoidomaxillary hypoplasia</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>Midface hypoplasia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Proptosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Slanting palpebral fissures</td>
<td></td>
</tr>
<tr>
<td>Carpenter's syndrome</td>
<td>Cloverleaf skull</td>
<td>?Omphalocele</td>
</tr>
<tr>
<td>Freeman-Sheldon syndrome</td>
<td>Flat, mask-like face</td>
<td>?ASD/tetralogy of Fallot</td>
</tr>
<tr>
<td></td>
<td>Hypertelorism</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>&quot;Whistling face&quot;</td>
<td>Intubation problems</td>
</tr>
<tr>
<td>Facial abnormalities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pierre Robin anomaly</td>
<td>Micrognathia</td>
<td>?Cardiac anomalies</td>
</tr>
<tr>
<td></td>
<td>Glossophtosis</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>?Cleft palate</td>
<td></td>
</tr>
<tr>
<td>Treacher Collins syndrome</td>
<td>Sloping palpebral fissures</td>
<td>?Cardiac anomalies</td>
</tr>
<tr>
<td></td>
<td>Hypoplastic mandible and zygoma</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>Micrognathia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>High arched palate</td>
<td></td>
</tr>
<tr>
<td>Goldenhar's syndrome</td>
<td>Ear anomalies</td>
<td>?Cardiac anomalies</td>
</tr>
<tr>
<td></td>
<td>Maxillary hypoplasia</td>
<td>Difficult mask fit</td>
</tr>
<tr>
<td></td>
<td>Micrognathia</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>Cardiac defects</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mental retardation</td>
<td></td>
</tr>
<tr>
<td>Neck anomalies</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Klipper-Feil syndrome</td>
<td>Failure of segmentation of cervical spine</td>
<td>Difficult intubation</td>
</tr>
</tbody>
</table>
Jehovah's Witness Patients

There is no easy answer to the problem presented by Jehovah's Witness patients, an issue that is further complicated by differences of interpretation within the religion as to what techniques of blood salvage (if any) are acceptable. Where children are concerned, there is always the possibility of obtaining a court order for an emergency transfusion, but this is seldom authorized for elective cases and is usually only issued when an emergency develops. The surgeon and the anesthesiologist need to develop a patient-specific plan in each instance that allows each of them to manage this difficult situation without being medicolegally or ethically compromised. Must as it is unreasonable for an anesthesiologist to discover that the surgeon has promised the parents that the child will not be transfused, it is irritating for a surgeon to find that a carefully orchestrated admission is canceled because the anesthesiologist does not accept the risk of undertaking the procedure without the assurance that court order will be obtained.

Preparation for Surgery

History and Physical Examination

Since plastic and reconstructive surgical procedures are usually elective, the anesthesiologist is able to assess if the lesion is associated with respiratory distress or stridor prior to surgery and can form an opinion as to whether intubation will or will not be difficult.

Evaluation of the Airway

A history taken from parents will often establish whether the facial anomaly is part of a syndrome and whether this could be of concern to the anesthesiologist. A thorough assessment of preexisting stridor and or hypoxia should be made and it may be necessary to request x-rays, a computed tomography (CT) scan, or magnetic resonance imaging (MRI) to evaluate the anatomy of the upper airway. Useful information can be obtained by awake, fiberoptic assessment of the airway. This may be possible even in small, uncooperative patients.

How Long Should Children be Starved Prior to Surgery?

Until recently, data specific to pediatric patients did not exist and the results of studies undertaken in adults were applied to children. Therefore, recommendations such as "starve from midnight" became part of pediatric practice. But the volume of gastric contents depends not only on the amount of food ingested, but also on the volume of salivary and gastric secretions, the tonicity and pH of the stomach contents, and the rate of gastric emptying. Studies from Sweden and France suggest that the risk of aspiration in the pediatric age group is three to eight times greater than that in adults. Yet when those patients with obvious risk factors are excluded, the aspiration risk in children is exceedingly low. Prolonge fasting in children does not decrease the gastric volume and, as one might predict, too short a fast after the ingestion of food increases the gastric volume and the risk of aspiration. Since the child's stomach is not "empty" after a period of starvation, and since drink of liquid prior to surgery paradoxically has been shown to reduce gastric volume, it has been suggested that it may be advantageous to offer children clear fluid such as dextrose water or apple juice (but no orange
juice or food) 2 to 3 hr preoperatively. Children less than 6 months of age can be fed up to 4 hr preoperatively, whereas children 6 months and older should not receive food for 8 hr prior to surgery. It seems that children offered fluid preoperatively are more comfortable and are less likely to develop intraoperative hypoglycemia.

**Premedication**

No one would disagree that the best premedicant is the trust of the child and the parents. The dilemma that pediatric anesthesiologists face in a busy ambulatory care facility where the order of cases is frequently changed, is the question of how to provide sedation painlessly and at an optimal time. A variety of agents and routes of administration have been used. In our institution, intranasal midazolam is a popular choice. Midazolam is drawn up into a syringe, and the stylette is removed from an angiocath and attached to the syringe. The soft catheter is then gently inserted into the child's nose. The drug has a rapid onset of action, is short acting, and does not require intramuscular injection, but unfortunately has an unpleasant taste and burns for a short while after administration.

"Conscious" Sedation

It is of concern that more patients are being sedated outside of the operating room environment, either because of cost constraints or lack of availability of anesthesia personnel. Drugs commonly used by anesthesiologists as premedicants are being used to sedate patients to facilitate procedures being done under local anesthesia or for diagnostic studies such as CAT scans. Many physicians are skilled at this and may have even received formal training in the care and monitoring of sedated patients. In our institution we follow the recommendations of the American Academy of Pediatrics. In our medical staff bylaws we have defined what is meant by "conscious sedation", "deep sedation", and "anesthesia". We have defined who is responsible for monitoring the patient, what type of anesthesia record must be kept, and how persons who undertake this kind of sedation are to be credentialed.

There is no "recipe" of agents that works best in this situation. This depends more on the experience and skill of the operator, the extent to which monitors such as pulse oximeters and nasal capnography are used, and the nature of the postoperative recovery facility. Drugs commonly used for sedation and local anesthesia are listed in Table 2.

**Table 2. Commonly used sedatives in pediatric anesthetic practice**

<table>
<thead>
<tr>
<th>Agent</th>
<th>Route</th>
<th>Recommended dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midazolam</td>
<td>Oral</td>
<td>0.5-0.75 mg/kg</td>
</tr>
<tr>
<td></td>
<td>Nasal</td>
<td>0.2 mg/kg</td>
</tr>
<tr>
<td></td>
<td>Intramuscular</td>
<td>0.1-0.15 mg/kg</td>
</tr>
<tr>
<td></td>
<td>Rectal</td>
<td>0.3-1.0 mg/kg</td>
</tr>
<tr>
<td>Ketamine</td>
<td>Intramuscular</td>
<td>1.0-2.0 mg/kg</td>
</tr>
<tr>
<td>(stun dose)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Recommended Monitoring Standards for Conscious Sedation

If sedation is going to produce an inability to communicate with the patient, there needs to be predefined, generally accepted monitoring criteria that apply to this situation and that can be enforced and assessed by the hospital quality assurance program. The following are important aspects for consideration:

The person monitoring the patient should have no other duties (ie, not the circulating nurse).

Monitoring should include not only the documentation of vital signs, but should also mandate minimum monitoring standards that need to apply when "conscious sedation" is exceeded either intentionally or unintentionally (ie, when is an EKG, pulse oximeter, automated blood pressure cuff, precordial stethoscope, etc, necessary?).

There needs to be easy access to resuscitation equipment.

Sedated patients need to be recovered in an appropriate facility where published standards of readiness for discharge from hospital are followed.

Anesthetic Management

Induction of Anesthesia

In any situation where airway compromise can occur, there should be a conscientious preparation of alternative techniques by which a patent airway might be secured. There should be the ability to undertake fiberoptic intubation, transtracheal jet ventilation, and urgent tracheostomy, should this become necessary. Usually induction is achieved by an inhalation technique, and once an attempt at visualization of the vocal cords has been made, a decision can be made as to how intubation can best be achieved. These cases should be undertaken preferably by pediatric anesthesiologists with experience in dealing with children with upper airway pathology.

If airway compromise is not an issue, any form of induction of anesthesia can be used (rectal, inhaled, or intravenous). It is important that no matter what technique is used, an intravenous line should be placed before attempting any airway instrumentation. The pain of intravenous needle sticks can be attenuated by the use of a special local anesthetic cream (EMLA cream). If this is applied up to an hour postoperatively, an intravenous infusion can be started painlessly.

Endotracheal Intubation

The operations should have a well-rehearsed plan of approaching a difficult intubation, and alternative means (such as a fiberoptic bronchoscope) should be available in case of unexpected difficulty. Centers specializing in pediatric care should have instruments such as fiberoptic bronchoscopes and jet ventilation attachments that are appropriately small for use in pediatric patients.
In addition to conventional endotracheal tubes, there are a number of special tubes that may be of use to the surgeon. Some of these are summarized in Table 3. Scrupulous attention should be paid to fixation of the tube since secretions and blood may loosen the adhesive on the tape and result in premature or unexpected extubation. This may require nothing more than using an adhesive such as Mastosol or tincture of benzoin, but may require that the surgeon suture the tube to the child's nose.

**Table 3. Special tubes of use to the surgeon**

<table>
<thead>
<tr>
<th>Type of tube</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>RAE tube</td>
<td>Molded nasal or oral tube that facilitates surgical access. Comes in a variety of sizes. Flammable, not reinforced.</td>
</tr>
<tr>
<td>Xomed tube</td>
<td>Silicon based, thin, cuffed laserproof tube that can be used for intraoral/airway laser surgery.</td>
</tr>
<tr>
<td>Armored tubes</td>
<td>Reinforced by a wire or plastic spiral. Come in a variety of sizes, but are not laserproof.</td>
</tr>
<tr>
<td>Porch tubes</td>
<td>Metal tubes, uncuffed, that are laserproof but are stiff, and come in a limited range of sizes.</td>
</tr>
<tr>
<td>Laryngeal mask</td>
<td>A tube developed in Great Britain that allows a cuffed tube to be placed over the laryngeal aperture without having to be inserted into the trachea. This has been used in children and may become a useful alternative to traditional endotracheal intubation.</td>
</tr>
</tbody>
</table>

**Anesthesia Maintenance**

**Monitoring Guidelines**

At the time of writing my recommendation is that all children undergoing upper airway surgery should have at least the following monitoring devices in place: noninvasive blood pressure cuff, EKG, temperature probe, and pulse oximeter. Monitoring should include the ability to analyze inspired oxygen, expired carbon dioxide, and inhaled agents. Since fluid management is critical in these patients, there should be a way of accurately delivering the child's fluid needs. It may even be necessary to place a central line if the anticipated blood loss is going to be significant.

**Readiness for Extubation and Post Anesthesia Care Unit (PACU) Care**

One of the most critical joint decisions that needs to be made by the surgeon and the anesthesiologist is when to extubate the child. Even a procedure such as repair of a cleft palate can be associated with postoperative lingual edema, which, if not recognized, can be potentially life threatening. Often the surgical procedure results in further upper airway edema and even a "trial of extubation" can be very dangerous if reintubation is going to be difficult or impossible. It is our recommendation that were possible, and appropriate, gentle laryngoscopy should be attempted and an assessment made about the ease with which reintubation could be accomplished. If there is any concern, we would recommend the conservative approach of admitting the child to an intensive care unit; then, when airway edema has decreased, planned extubation can be safely accomplished.
If it is possible to extubate the child, we keep the child in the PACU until both the anesthesiologist and the surgeon are content that the child can safely maintain his/her airway. It is much easier to intervene in the operating room environment than in a distant part of the hospital where one is less familiar with the equipment.

Particular attention should be paid to providing analgesia that is titrated to the needs of the patient. Too little is just as troublesome as too much.

The patients may be more comfortable with the judicious use of antiemetic medications, but the benefit of doing this needs to be balanced against the risk of oversedation when analgesics are being used at the same time.

Complications

General

Common general anesthetic complications are those related to the management of pediatric patients in the operating room and include hypothermia, overhydration, hypoglycemia, inadequate assessment of blood loss, inadequate monitoring, and lack of attention to the different pharmacological needs of infants.

Specific

Specific complications refer to the difficulties of either securing the airway in the first place or the preparation needed for expeditious reintubation should the child's trachea become inadvertently extubated. The attending staff should be experienced in the management of these patients and should demonstrate great flexibility in their ability to change techniques rapidly should the situation demand it. They should be familiar and practiced in the use of fiberoptic bronchoscopy for tracheal intubation and should have access to transtracheal jet ventilation as well as the ability to establish rapid surgical access of the airway either by cricothyrotomy or tracheostomy. The best units have a surgical team that undertakes these procedures where there is rapport and mutual understanding between the anesthesiologist, surgeon, and nursing staff such that emergency situations are managed with a minimum of fuss.

Conclusion

The anesthetic management of children for surgery either of the upper airway or surgery that by its nature may impinge on the airway is very stressful and highly specialized. It requires elaborate preparation, extensive experience, and is best managed by a team of physicians who work comfortably together and have a mutual respect and understanding for one another's problems. We are fortunate to have all of these elements.

Alexandra Murray Harrison

The Emotional Impact of Craniofacial Disorders

What is an "Attractive" Child?

What do we mean by "attractiveness"? Suppose we take for an example the case of 4-year-old Molly, with silky brown hair, large brown eyes, and regular features. Molly turns from the mirror to her mother with a sad expression on her face. "I'm ugly", she says. Her shocked mother asks her how she could say such a thing. "I wish I had blond hair and blue eyes like Cinderella", Molly answers. Incidental to this conversation was the birthday party of Molly's 2-year-old brother.

Four-year-old Karen, who has Crouzon's syndrome, comes for a clinical appointment with pastel barrettes carefully placed in her dark hair. Her flowered cotton shirt and shorts outfit includes a junior version of stylish shoulder pads, and her tiny feet are fitted with pink sandals. In spite of the anomalous configurations of Karen's head and face, she seems to feel good about herself and feel proud of her appearance.

These examples illustrate the kinds of observations all of us, but especially those of us who work with "disfigured" children, have struggled to understand. How can we make sense of these contradictions? What conceptual tools do we have? Social psychological theory can help us understand the reactions of people to deviance or stigma; such reactions must contribute to the self-image of a disfigured child. Developmental psychology can help us understand the factors making up self-image in the developing child. Finally, psychoanalytic theory can help us understand the unconscious processes that give symbolic meaning to disfigurement and impairment.

Social Psychology

What kind of responses can children with facial deformities expect from those around them? In his classic work on "stigma", Goffman defines stigma as "undesired differentness" of which physical deformities are one type. He refers to the ancient Greeks' use of the term to signify marks on the body, or brands, identifying the bearers as people to be avoided. Indeed, studies have shown that people attribute negative characteristics such as lack of intelligence, or even dishonesty, to those with facial disfigurements. In a corresponding manner, they are likely to attribute positive characteristics to people with "attractive" faces. Studies have shown that children as young as 3.5 prefer "attractive" peers as friends and associate unattractive children with "antisocial" behavior. These findings are consistent with the relationship between stigma and morality described by social theorists. This notion that
good is associated with beauty and bad with ugliness is demonstrated in myths and literature throughout the ages, for example in many fairy tales and in literary villains such as Shakespeare's Richard III.

**Developmental Psychology**

Developmental psychology, on the other hand, gives us knowledge about the ways a child learns about himself and others and about the preeminence of the face in building human relationships. Infants prefer looking at the human face to looking at other visual patterns. Perhaps one of the reasons for the importance of the human face to the infant has to do with the way emotion is communicated through facial expression. In cross-cultural studies Ekman and Izard have demonstrated there is a universal ability to identify basic emotions such as happiness, sadness, anger, and fear in schematic drawings of a human face.

The fact that the infant registers the configurations of the face in contrast to the facial features alone is revealed by the findings of Field et al, who have shown that 2- to 3-day-old infants can discriminate and imitate smiles, frowns, and surprise expressions in the face of a person interacting with them. The earliest loving attachment between infant and mother is accomplished through facial expressions and vocalizations. This bond has gained prominence recently in the field of "attachment theory", a specialized area of developmental psychology. Attachment theorists relate security in this earliest relationship to successful emotional development and self-esteem throughout life.

Other contributions to our understanding from the field of developmental psychology include ideas about how infants organize perceptions about their environment, and observations about when children begin to recognize themselves through physical appearance. "Amodal perception" is a concept described by Stern, which refers to the process by which an infant associates similar patterns across different types of sensory modalities, such as auditory and visual stimuli delivered at the same frequency, in order to make generalizations about his environment. Could this characteristic of learning relate to the preference infants demonstrate for symmetry in the vertical plane, as in the human face? That children learn to recognize their own visual image between 15 and 24 months of age has been found by a study in which a rouge spot was put on a child's nose and then the child shown his reflection in a mirror. Three-quarters of the infants exhibited mark-specific behavior between 21 and 24 months of age; none of the 9- to 12-month-old group appeared to notice this deviation from their characteristic appearance.

**Psychoanalytic Theory**

Returning to our example of the two 4-year-old girls, we are better prepared to comment on their "attractiveness". We know that Molly and Karen have a clear "facial identity" by now, and have a sense of how their faces compare with the norm. They have used their faces and their mothers' faces to establish an "attachment" relationship and to communicate emotions within this relationship and others. We could guess that cultural stereotypes might in part explain Karen's positive self-regard with respect to her stylish and feminine attire, and Molly's negative evaluation of her dair hair and eyes. However, we are still left with mysteries. Is Molly's lack of resemblance to the Walt Disney image of Cindrella enhanced by her perceived difference from the fairy tale heroine? How do we explain Karen's
apparently positive self-esteem?

Using psychoanalytic theory we can make some hypotheses about Karen's unconscious thoughts. Perhaps her jealousy and anger at her little brother, who has just had a birthday party and who has arrived at an age when he can become a more active and bothersome competitor, has made her feel like a bad girl. Now, a bad girl could not possibly be a Cinderella, and therefore, she could not be beautiful. However, these unconscious ideas can be elaborated even further to help us understand children with disfigurements and functional impairments. Children also associated "defectiveness", which can refer to both disfigurement and functional impairment, with loss.

Regardless of whether or not the defect involves a concrete loss, children with body defects see themselves as missing something or as being vulnerable to missing something. This "have-not" image includes feelings of being "bad" or unlovable, which are organized around the body defect. In other words, the child unconsciously feels there is something bad and unlovable about them that has to do with the defect. This does not mean that children with a body defect cannot have good self-esteem or see themselves as lovable. It only means that whenever they do feel bad about themselves, they are likely to relate that bad feeling to the body defect consciously or unconsciously. For example, a 4-year-old girl with a large facial birthmark told her parents that she could never be the Madonna in the Christmas play because of her birthmark. Like Molly in the first example, this child had a little brother and felt guilty about her hostile feelings toward him. It was not only the Madonna's beauty, but also her goodness that this little girl was (unconsciously) referring to. Another child with a disfigurement due to a traumatic injury, felt guilty about angry, competitive feelings toward her mother, whom she saw as having "everything" (husband, career, baby). She played a game in which she tried to find something missing inside a doll, and drew a picture of herself as, in her wonderful descriptive term, "an ugly stepmaid".

The Emotional Impact of Craniofacial Anomalies on the Developing Child

Now that we have outlined several conceptual models with which to study the emotional impact of craniofacial anomalies, we may proceed to a consideration of the influence of these defects on the child's developmental achievements and sense of mastery. To do this it is helpful to organize the discussion according to different ages of the child and to think of the child not alone, but in relation to his environment: his family, peers, community, and society. Taking a look at the child in this context is important because the effect of the facial anomaly is the result of a reciprocal process. As we shall see, in addition to the stage-specific tasks facing these children and their families at each developmental stage, they are also challenged with the disappointment that comes with having an "imperfection". Although we all have imperfections, facial anomalies are particularly obvious and do not go away. These children and their families go through repeated experiences of mourning.

Infancy

The birth of an infant with a disfigured face requires the parents to confront the loss of their fantasized "ideal child". This fantasy is part of normal family development and carries healing and hope to all families expecting a baby. Instead of the usual slow process of giving up the idealization of the perfect child, which occurs for families of healthy children bit by
bit during the course of childhood, the parents of the defective newborn must precipitously and painfully give up their fantasy of perfection. All family members acquire some of the negative valuation of the stigma just by virtue of the association with the disfigured child.

From the point of view of the infant's development, there is now some evidence to support what has long been hypothesized, that the presence of a craniofacial anomaly in a newborn can interfere with the normal attachment process. Functional difficulties with eating, vision, hearing, or vocalizing can also affect attachment by diminishing the satisfaction of the feeding experience on the part of both baby and mother, by interfering with the powerful mutual activity of gazing, and by affecting the reciprocal vocalizing that forms another important aspect of bonding. It is also difficult for extended family and community, even society at large, to welcome a defective baby. Everyone shares in the hopefulness of the ideal child fantasy, and confrontation with an obviously imperfect infant destroys this fantasy immediately, leaving anxiety in the place of pleasure. Not surprisingly, there is evidence that the birth of a stigmatized child restricts the social life of a family.

Preschool Period

The period from 1 to 5 years of age is an active time of practicing separation from parents and exploring the world in a limited setting. The child often goes to nursery school, an extension of family life and a part of the small community of early childhood that includes new caring adults and a group of peers. There is evidence that nurturing parents and a protective community can foster good self-esteem development in the stigmatized child during this stage of life. However, outside the comforting familiarity of these groups, the child with a facial disfigurement may not be well received by his peers. Even preschool children express awareness of disabilities, and by 3.5 years of age have demonstrated a negative reaction to facial disfigurement.

Pertschuk and Whitaker found parents of young children with craniofacial deformity to exhibit none of the "rejecting and overprotective" behavior suggested in the anecdotal literature. This is consistent with many observations of good adjustment in young children with craniofacial disfigurement and in their parents. However, Pertschuk and Whitaker's study did show much poorer functioning in their older age groups. In addition to the suggested explanation of different societal challenges faced by their older and younger groups, the study may also suffer due to failure to take into account subtle interactional factors in the family that can lay the groundwork for anxious or inhibited behavior in later childhood.

Anxious and controlling behavior on the part of parents, as well as unwelcoming responses from other children, can cause problems in separation, which is the primary developmental task of this age. For example, it may be harder for parents to encourage their disfigured child to go to school against his will than it would be for a less vulnerable child. Functional impairments, especially as they involve speech, vision, or hearing, will clearly put the handicapped child at a disadvantage in school. They will also threaten other children, who will identify with the appearance of incompetence. Finally, they will complicate the efforts of both parents and teachers to make realistic expectations for the child. If the child is clinging and unwilling to participate, or if the child demands special attention, it may be hard to tell when to accommodate the child's expressed needs or when to set limits. Aggressive behavior may also pose a problem from this point of view: How much can the child control
by himself, and how much is the behavior a natural response to provocation?

In contrast with aggressive behavior, children with craniofacial deformities appear to withdraw, inhibit their impulses, and become dependent in response to feeling vulnerable. This pattern of behavior, although adaptive in some ways, may set the course for future problems by restricting the child's repertoire of coping responses and putting a cap on ambition and risk taking, as well as negatively influencing self-esteem. In my experience, the school community and society tend to be quite tolerant of children of this age with a facial deformity. Once the initial threat of the disappointment at birth has passed, and the competitive pressures of the school years have not yet begun, people generally extend a benevolent and protective attitude toward these children, perhaps particularly because they are often compliant.

**School Age**

School years present an even greater challenge to the child with a facial deformity and his parents, as demands for autonomy and performance in social and academic areas increase. Society tends to be less tolerant of school-aged children with a deformity. Societal values stress successful performance and conformity, rather than diversity. Peers continue to register a preference for nondisfigured children as friends, and functional impairments present a greater disadvantage to the child in terms of school performance, and to his teachers and parents in terms of making realistic expectations of him. As one might expect, studies show that children with craniofacial deformities have poorer academic performance, greater separation problems, and lower self-esteem than other children.

**Adolescence**

Adolescence poses particular problems for the facially deformed child. As earlier, the separation task is complicated by the presence of a deformity, which makes the child vulnerable in several important ways. There is a high likelihood of being rejected by peers; adolescents and young adults are progressively more negative about disabled peers. It is hard for the deformed child to fit the group ideal. Conformity to a group ideal facilitates the separation from parents and, through identification with this ideal, allows for the development of one's individual identity. It is harder for the disabled adolescent to succeed in his schoolwork because academic performance is more competitive, in preparation for the demands of adult life in this society. All these factors put pressure on the parents, who worry about their vulnerable child, who is soon to leave the scope of their protection. Setting realistic expectations for behavior and performance at this stage is another major problem, as the stakes get higher and the demands for the child to function autonomously increase.

Sexual development presents a new challenge to the adolescent with a facial disfigurement. Worries about being attractive to the opposite sex and anxiety about changing body image often focus on the deformity. A woman with a facial birthmark reported that as an adolescent she lost interest in clothes because "All people looked at was this (the birthmark) - what did it matter what I wore?" The feelings of rejection and failure she experienced led her to "drown her troubles" in alcohol and delinquent behavior. Anxious about her developing woman's body, another girl with a birthmark joined a fundamentalist religious group that taught "It's what inside that counts".
New demands on parents for limit setting are complicated not only by adolescents' tendency toward action, but by their greater need for independence and responsibility. The need to mourn the "perfect child" presents itself again with a vengeance at this stage. Instead of the young man or woman who is prepared to go out and conquer the world, the parents of the disfigured adolescent see their beloved child as older, but clearly imperfect, still facing harrowing possibilities of rejection and failure. Parents with greater capacity to tolerance the powerful mix of feelings this awareness stirs in them, hate as well as love, are better able to do the requisite mourning and let their child grow up.

The Questin of Surgery

The Surgeon as Consultant

The first step in making decisions about surgery is to clarify the surgeon's role as consultant to the family in making decisions. Staying carefully within this role can avoid grave problems as well as make it possible for the surgeon to work effectively. The role of the consultant is to use special expertise to answer questions. This is different from the role of the surgeon in the operating room, or during most of the time he is taking care of patients. It involves finding out what questions the patient and family want answered, how these questions can be modified to create questions that can realistically be answered, and, finally, answering the latter questions. The answers usually include a recommendation about what action to take. Staying within the role of consultant helps the surgeon avoid (a) making unrealistic promises, (b) attempting to rescue a patient, and (c) giving the patient unwanted advice. It is hard to make a serious mistake if one pays careful attention to one's role as consultant.

Whether or Not to Do Surgery

There are two main issues related to surgery: whether or not to do it, and when to do it. In order to answer either question, the family unit must be assessed. Four questions must be answered. (a) What are the family's goals with respect to surgery? (b) How are decisions made in the family? (c) How does the family deal with frustration and disappointment? (d) What are the family's resources?

What are the Family's Goals?

This question represents an effort to determine whether surgery can realistically satisfy the patient and his family. The surgeon is in a good position to predict what the surgical procedure could offer. Is this consistent with what the family wants? In the best situation, the child and family have a long-standing relationship with the surgeon and have discussed the possibility of surgery for some time. They have talked often about the kind of surgical result that might be realistically expected. In this case, the family's goals for surgery are usually consistent with the average, expectable result.

In other cases, where the situation is not so clear, when the surgeon does not know the patient and the family so well, there may be a hidden agenda. Sometimes the "hidden agenda" is conscious, such as a lawsuit in the case of a traumatic injury or a previous surgery with "unsatisfactory" result.
Other times the motive is unconscious. For example, an adolescent with a Natalie Wood-like nose requested a rhinoplasty to restore her nose to its larger, hooked, original shape, the way it was before a previous rhinoplasty. She explained that she could not recognize herself with her present nose, that she felt "lost" and hopeless. Psychiatric interview revealed an unconscious motive, without which her requests for surgery were incomprehensible. She had requested the original rhinoplasty in an effort to rid herself of, to use her words, her "mother's nose". When this was accomplished, she felt she had lost "her mother in her". Her unconscious feelings about her mother were actually very ambivalent and included a powerful longing and dependency toward her mother mixed in with her conscious anger and rejection. Thus when she "lost her mother", she became depressed and could not rest until she restored her mother within herself in the form of her original nose.

How are Decisions Made in the Family?

If the surgeon inquires about how the family makes other important decisions, he may discover that the family approaches the problem together, includes the child in the decision-making process in a manner appropriate to this age, and comes to a consensus that everyone can support. On the other hand, he may find that the parents make the decision independent of the wishes of the adolescent patient, or expect a 6- or 7-year-old child to decide for himself. He may uncover a marital conflict that centers around the decision, with mother on one side and father on the other. Identifying these problems early may avoid unnecessary complications later on. Generally speaking, the young school-age child is too immature to take on the responsibility of the decision of whether or not to do surgery for facial disfigurement. His opinions should be elicited, but not in the context of whether or not the procedure will actually be done. Around the age of 8 the child begins to be prepared to take a more active role in the decision making. Nevertheless, it is not until mid- to late adolescence that the child becomes the primary decision maker.

How Does the Family Deal with Frustration and Disappointment?

Problems can arise when families are unable to confront their disappointments and frustrations and struggle with the painful realities in order to make peace with them. For example, the parents of a child with a traumatic injury came to the psychiatric consultant with the request for psychotherapy for the child, who was said to be depressed as a result of his injury. After some weeks of evaluation sessions, the parents began to complain about the inconvenience of taking the boy to his sessions. Since the child was indeed depressed, the parents' complaints did not seem consistent with their original request for treatment for the boy's unhappiness.

Through the course of the evaluation the psychiatrist had learned that the father had been out of work for some time, and the couple was having marital difficulties related to the father's work problem. It seemed that the family was ready to break apart. The parents said that the father's previous employer was to blame for his having to leave his job. However, it was clear from other remarks that the mother joined the father's blaming the employer in order to avoid blaming him. Her own father had abandoned the family when she was a child, and she was afraid to focus her rage at her profoundly disappointing father onto her husband, because that might mean that she would have to leave him. Further questioning revealed that the boy's school was being sued for the injury, and that the family's agenda for the
psychiatrist was not psychotherapy, but litigation support. Indeed, it seemed that the family's method of dealing with disappointment and frustration was to blame someone else, and this blaming behavior, actualized in the litigation, got in the way of their attempts to mourn their real disappointments and get on with their lives. The boy's depression reflected his parents' interference with his efforts to come to terms with his injury.

**What are the Family's Resources?**

By "resources" I mean not only the financial means to pay for the procedure. I am also referring to the network of support the family can call on in times of need. Extended family, friends, church, community groups, and agencies are all of potential help during the stressful time of the hospitalization, and in the healing time afterward. All of us have seen hospital rooms decorated with cards, banners, plants and flowers, etc, that testify to the caring community supporting the room's occupant. These are the same communities that take care of the family's other children so that the mother can stay in the hospital with the patient, who do errands for her and help feed the family, and offer emotional support to the family. The family in the example above had alienated its neighbors with its attack on the school and found itself as isolated in the community as the family members were isolated from one another. The vulnerabilities of such a family can be contrasted with a family that has been able to develop strong supportive relationships within the community. These supports can make a real difference to the success of surgery. Exploring the possibilities of such supports with a family in relation to planning a procedure can sometimes help the family identify them and get them into place.

**The Timing of Surgery**

A key issue in the decision about surgery is the timing of the procedure. When I first began consulting to pediatric surgeons, in the early 1970s, the prevailing belief of child psychiatrists and psychologists was that surgery should be delayed as long as possible because of the traumatic consequences of hospitalization and painful procedures to the young child. This belief developed in part from the pioneering work by child psychoanalysts with children who were chronically institutionalized, children who had been separated from their parents during World War II, and children who were institutionalized temporarily for reasons such as the birth of another child. The belief also derived from early psychoanalytic theory, which stressed the long-lasting effects of childhood traumatic experiences on the personality, and emphasized parental separations and abandonment, as well as physical pain imposed on the helpless child, as prototypical traumas of this type. It is generally agreed that hospitalization and surgical procedures are more stressful for the young child, eg, younger than 7, than for older children. This is especially true for children who do not yet have language to organize their experience and to communicate with their environment. Does this mean that surgery should be delayed until adolescence in most cases?

In fact, waiting until adolescence is not usually recommended. The traumatic potential of hospitalization has changed significantly. Even as recently as the early 1970s, it was rare for a parent to be able to stay with the child overnight in the hospital. Now, "rooming in" is frequent. The observation that the child was more compliant in the parents' absence was frequently attributed to a problematic parent-child relationship such as poor parental limit setting, rather than to a worrisome "submissive" defensive behavior on the part of the child.
Now, the child's "fighting back" is recognized as a sign of health, which can be controlled without being crushed. Psychiatric consultation in most hospitals in the 1970s had not yet developed to the point of offering the surgeons the specialized information necessary to understand the effect of the cognitive limitations and defensive distortions on the young child's perception of what was happening to him. Now, many surgeons have expert psychiatric consultation to help them appreciate their young patients' perceptions and capacities for cooperation and taking responsibility.

At the same time, surgical technique and the scientific knowledge supporting it have improved tremendously. It is now possible to improve the appearance and reduce the functional impairment of congenital anomalies much earlier in the child's life, sometimes in early infancy. My experiences as consultant to the plastic and reconstructive surgery team helped me appreciate the benefits of reconstructive surgery early in a child's life, benefits that include an improved aesthetic appearance for a larger portion of the child's development. My consultations also led me to appreciate the complex nature of the human response to facial disfigurement.

There has long been talk of "sensitive" periods in a child's development, when disruptive and potentially traumatic experiences such as surgery should be avoided. Yet an argument can be made for many such "sensitive" periods and there is little "hard" evidence for any of them. Certainly, the support available to the child is a necessary consideration in any evaluation of sensitive periods. Can the factors of abandonment by parents, loss of comforting routine of home, confusion, and frightening fantasies about what is happening to him or her be minimized or mitigated? Parent "rooming in", flexibility of ward routine to suit the needs of the individual child, and hospital social and psychological supports for the child and family can all modify the "sensitivity" of the child to a surgical procedure. There are, however, a few general comments that can be made about "sensitive" times: (a) The second half of the 2nd year of life is a time when the child consolidates his sense of himself, a time when many different developmental paths converge in a burst of integration. It would be better not to disrupt this important developmental moment. (b) At ages 3 to 5, children are particularly vulnerable to anxieties about body injury. They are struggling to manage their aggressive impulses, and are apt to interpret surgical interventions as retaliations for their "bad" out-of-control aggression. This would be a good time to avoid. (c) Preadolescence, which occurs at about 11 to 13 for girls and 12 to 14 for boys is an especially vulnerable time in terms of body image. Children of these ages are very anxious about whether or not their body is all right, and they often feel confused and out of control in relation to their bodies.

Even these statements about sensitive periods should be used only as general guidelines to alert the surgeon to developmental changes occurring at these stages. Many children will go through these changes at ages different from those I listed. This is especially true of children with body defects, who are often somewhat delayed in their emotional development. Psychiatric consultation related to this point may be useful.

The Argument for Early Surgery for the Sake of Psychosocial Development

What about the idea that improving a facial disfigurement spares the child problems with his psychosocial development? Even very good surgical results do not completely erase the deformity, and social psychological theory provides evidence for the fact that slight facial
disfigurement is a powerful stimulus for rejection of the disfigured person. Some observers have noted that objectively minor disfigurements often have powerfully disadvantageous effects on social and psychological adjustment. If the detrimental effects of early hospitalization and surgery are not offset by the advantageous effects of improved aesthetic appearance offered by early surgery, should postponement be the rule?

In search of an answer, let's return to the example of "Karen". Karen came from a middle-class family with a warm, mutually respectful marital relationship and strong social supports. Older children in the family had done quite well in school and provided the parents with a sense of having a successful family. Now the parents's self-esteem focused on doing "the best we can" to help Karen "develop according to her potential" and to "help other people realize what a wonderful person she is". They experienced the surgeon as helping them do their job of being good parents to Karen. First considering and then planning the surgical procedures made them feel less helpless as parents of a child with a problem. They considered the results of her surgery a real improvement, but the objective results were only part of what helped them feel successful. Fortunately, they also struggled with the continuing awareness that they would never achieve a perfect result. There were ups and downs for them as for all parents in comparable situations. Where does this lead us?

Although the development of self-esteem in relation to body image has unfortunately not been studied in a careful, systematic way, some things are clear. Self-esteem in the disfigured child is not solely a function of the objective disfigurement, not even of the social reaction to it. The child's self-esteem is also a function of the family's capacity to deal with the demands created, and the disappointment represented by the deformity. Regardless of the sensitivity of society to subtle signs of disfigurement, the parents' estimation of the improvement, coupled with their sense of mastery over the helplessness they felt in relation to their defective child, may have a powerful impact on the family self-esteem and thus on the self-esteem the child derives from being a valued member of that family. "We did everything we could do", is a major satisfaction to add to even a minor aesthetic improvement. This knowledge on the part of parents marks a turning point that initiates the necessary mourning.
Rules of Thumb in Making Decisions About Surgery

It is helpful, perhaps, to summarize with a few rules of thumb for the use of the surgeon in his role as consultant in recommending surgery:

1. Make an estimation of the successful outcome of surgical intervention for a particular facial disfigurement or functional impairment.

2. Identify the points in time during the physical development of the child or of the deformity when surgical intervention would be optimal from the point of view of the surgical technique.

3. Assess the capacity of the child and the family to make a responsible decision about surgery, to gather the requisite supports, and to deal with frustration and disappointment.

4. Balance the advantages of interventions early in the child's life on improvement of physical appearance and enhanced family self-esteem, against the problems presented by the particular vulnerabilities of the young child, which require special supports during the hospitalization and in the extended recovery period.

5. When possible avoid the "sensitive" periods of 18 to 24 months, 3 to 5 years, and 11 to 13 in girls and 12 to 14 in boys. Remember that these "sensitive" periods are approximations and may be different for different children. When in doubt, a psychiatric evaluation may help in this assessment.
Best Case, Worst Case Examples

Best Case #1

Betsy was a 9-year-old girl with a large facial hemangioma that had faded to the point of being a slightly darker shade of her skin color. A good student with many friends, Betsy was a sensitive child who had problems with separation from her parents, but who generally did well and enjoyed her life. Her parents had some rocky times in their marriage, but they were strongly allied in support of their child's healthy development and worked well together to support her. They had many friends and a helpful extended family. Betsy and her family had a long-term relationship with the same surgeon, who had followed Betsy's problem with them since early infancy. Together, they had anticipated the points at which surgical intervention might be helpful, and had been through the decision-making process before. Twice in the past they had considered surgery and decided to put it off, first because there was not high-enough probability of a good surgical result, and second because the disfigurement did not seem to interfere enough with Betsy's life at the time. Now, in anticipation of adolescence, and with a good surgical result highly probable, the "team" of Betsy, her parents, and the surgeon, decided to schedule the surgery.

Best Case #2

Jody M's parents were both physicians and had long awaited their first child. When Jody was born with a severe cleft lip and palate, both parents were distraught. The surgeon's immediate response to the referral from the neonatologist was a source of great relief in the crisis. Although the reality of chronic disfigurement of some degree and the possibility of multiple interventions could not yet be encompassed, the first surgical repair made it possible for Dr M to bond to Jody, and renewed the hope that had been destroyed in the shock of the birth experience.

Worst Case #1

A 12-year-old girl came with her parents for consultation about a congenital abnormality of her nose and chin. She was obese and unkempt. Her parents said that she had always suffered from her poor appearance and that they had been told that surgery could "fix" it before she went to high school. Since she had already been scapegoated in elementary school, they were worried about her entrance into junior high, which had a reputation in their town for being tough. The parents had made application for her to go to a private school, and although the girl refused to go to private school, the parents said that if she got in, they would make her go, "for her own good".

Worst Case #2

A 4-year-old boy with a diagnosis of "hyperactive" came for consultation about a congenital abnormality of his ears. His mother, a single parent, said that his ears had bothered her since his birth. They reminded her of his father, who had been abusive to her, and they made him look stupid. He had been hospitalized earlier that year for an emergency appendectomy, and he had been a real problem in the hospital, requiring physical restraints to keep him from dislodging his IV. During this hospitalization, the mother had complained
about his ears to the pediatrician, who had made the referral to the surgeon.

Discussion of Cases

These cases are designed to illustrate the main points in the "rules of thumb". The "best cases" illustrate the child and family who are capable of making responsible decisions, who have adequate supports, who have demonstrated capacity to deal with frustration and disappointments, and in which the child is not at a "sensitive" age. The "worst case" illustrate the child and family who are probably not capable of making responsible decisions together, who have inadequate supports, who have demonstrated difficulty dealing with frustration and disappointments, and in which the child is at a "sensitive" age.

Conclusion

Children and their families are helped when the children look as normal as possible as early as possible. It is better for all children to have all the right equipment as soon as they can. Early intervention should be balanced against the negative effects of hospitalization, but it is often possible to minimize these effects through supports in the environment both inside and outside the hospital. Children with facial disorders cannot be evaluated without considering the family context. Surgery should be postponed or avoided if other problems in the child or within the family are being displaced onto the child's disorder. Under good circumstances, evaluation for surgery and surgery itself presents an opportunity for psychological growth that goes beyond the physical result of the surgical procedure. It can become a team effort including the surgeon, the child, and the parents. In replacing helplessness with hope, it can initiate the process of mourning the loss of the ideal child. Making peace with this disappointment can accomplish a step forward in an important maturational process, that of finding value in oneself as one is, and in the related process of learning how to find realistic satisfaction in life.