

Pediatric Facial Plastic and Reconstructive Surgery

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Chapter 8: Pediatric Ophthalmic Plastic and Reconstructive Surgery

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Medicine continues to enjoy an ever-expanding fund of knowledge. The subspecialties of pediatric ophthalmology and ophthalmic plastic surgery each contain a wealth of information that would easily fill a generous-sized textbook. Our discussion will initially focus on the pediatric eye and visual development. There are many potential vision-threatening hazards that await the pediatric ocular adnexal surgeon. Recognition, avoidance, management, and appropriate referral of these problems will be discussed as they relate to the more common eyelid, lacrimal, and orbital problems seen in this age group.

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.

Blepharoptosis 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 *lacrimonal 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 dacryoceles 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 infraction 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

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 canaliculi 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 canaliculi are lacerated, the system is intubated with silicone. The canaliculi 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 dacryoceles. 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*

Cellulitis
Nonspecific orbital inflammation (pseudotumor)
Dermoid
Hemangioma
Lymphangioma
Rhabdomyosarcoma
Glioma of optic nerve
Optic nerve sheath meningioma
Chloroma (leukemia)
Metastatic disease (neuroblastoma)
Retrobulbar hemorrhage.

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 an am cortisol 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 meningotheial 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.