Pediatric Ophthalmology in the Emergency Department

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Examining the young child with a visual or ocular complaint can be a daunting challenge. Understanding the basic concepts of visual and behavioral development will facilitate the examination of the child who presents to the emergency department with eye complaints. Ocular complaints may include pain and visual impairment which may lead to anxiety and interfere with the examination of the child. Keeping the child calm and taking the time to engage the child in a manner he or she is comfortable with will allow a more accurate examination.

Visual development

Vision development is a complex system that requires the development of neuro-ocular pathways and depends on proper visual stimulation of both eyes. The first 3 to 4 months of life are most critical for this development. If significant disruption of a child’s vision occurs during this period and is not quickly corrected, lifelong visual deficit is the likely result despite later treatment. The rate of vision development remains steep until about 2 years of life, at which time three-dimensional binocular depth perception develops. It is not until 9 years of age that the brain’s development of vision is complete.

Full-term newborns do not generally respond well to visual targets. Visual acuity at birth is approximately 20/400. In newborns, the presence of vision may be demonstrated by pupil responses or by aversive behavior to bright lights. Eye position at birth varies greatly. Outward deviation may be normal eye alignment in the newborn period. After birth, the eyes tend
to move to a more convergent position and should be well aligned and stable by 4 months of age [1]. The pupils of newborns are often constricted.

Although fixation is generally present at birth in the full-term newborn, the ability to follow targets is not developed until about 3 months of age. Accommodation, the ability to focus, develops by 4 months [2,3]. Vision improves dramatically during infancy. By 1 year of age, children's vision is 20/50 and by 2 years of age 20/20.

The eye examination in a child

Assessing vision in a child can be difficult but should be evaluated in every child with an eye complaint. To accomplish this it is necessary to adjust the examination to the age and cognitive ability of the child. There are many aspects of the eye examination, but in the emergency department, the practitioner may focus on the skin and the surrounding tissues, light responses, fixation responses, and visual acuity. The discussion herein focuses on those parts of the eye examination which differ in children and adults and describes the examination appropriate for children of different ages [4].

Testing of visual acuity varies markedly depending on the age, verbal skills, and cooperation of the child. Eye alignment is an important part of the evaluation in children. In infants, misalignment or strabismus can lead to severe visual deficits. Misalignment may also be associated with a range of acute processes including orbital cellulitis.

Examination of the newborn and young infant

The first part of any eye examination is to observe the child. In children of all ages, it is best to leave the most invasive part of the examination that will cause the most distress to the child until the end. One should evaluate the lids and the periorbital area for swelling, redness, drainage. One should observe how the child moves their eyes and note the color of the conjunctiva and sclera. The macula in young infants is not fully developed; therefore, the eyes do not fixate well centrally and do not follow objects until about 3 to 4 months of age. To examine infants in this age group some recommend having the parent hold the child in the feeding position and then move the child’s head from side to side. The baby should follow this movement with his or her eyes or head and should also blink when a light is shone into their eyes. Another method of evaluating an infant for fixing is to cradle him or her in one arm upright and facing the examiner while gently rocking the infant side to side. If an infant has the eyes closed, he or she will generally open them when rocked in this manner, allowing for examination.

Of note, young infants may have intermittent downward deviation of the eyes. This finding usually lasts only a few weeks. If this sunsetting is constant or associated with poor feeding, lethargy, a large head size or bulging fontanelle, or occurs in a child in whom it had not been present, it may be...
caused by increased intracranial pressure. Downward eye deviation that persists pasts a few weeks may be a sign of neurologic conditions, and the child should be evaluated by neurology and ophthalmology.

The retina is indirectly evaluated by examining the red reflex. Fundoscopic examination of a young child who is awake and has not had his or her pupils dilated can be extremely difficult. The red reflex allows the examiner to assess light that enters the child’s eye and is reflected off the retina. The examiner should dim the lights in the room and calm the baby by giving the child a pacifier or bottle or by gentle side-to-side rocking. With the child’s eyes open, the direct ophthalmoscope can be used to look at the red reflex. The key to evaluating the red reflex is symmetry and uniformity in the child’s eyes. In light skinned infants, the red reflex appears orange-red. In dark skinned infants, the reflex looks dull orange or whitish orange. This finding should not be confused with leukokoria (Fig. 1), which is a whitish appearance of the pupil that, if present, is not generally found in both eyes. Leukokoria indicates a problem with reflection of light from the retina and may be caused by an array of pathologic entities including cataracts and tumor.

**Older infants and preverbal children**

One should start with the least invasive, least painful parts of the examination in older infants and preverbal children. Some young children are afraid of physicians because stranger anxiety is most pronounced around 10 months of age. One should begin the examination by observing the child from a distance to determine whether there is any swelling or redness to the eyes and surrounding tissues and how the child uses and moves their eyes. Children in this age group usually fix on and follow a toy or other object of interest. With some children, it may be necessary to seat the child in the guardian’s lap and have him or her hold the child’s head still while the examiner moves the object from side to side and up and down.

Fig. 1. Right eye leukokoria from a traumatic cataract. *(From Levine LM. Pediatric ocular trauma and shaken baby. Pediatr Clin North Am 2003;50:145; with permission).*
In preverbal children visual acuity can be difficult to assess. A child who consistently protests to having one eye covered as opposed to the other likely has better visual acuity in the favored eye. To check for such a preference, the examiner should cover one eye of the child and observe whether the child fixes and follows with the uncovered eye. If the child objects to having the eyes covered by the physician, sometimes enlisting the aid of a guardian to hold a hand in front of the child’s eye can be helpful. Again, the child who consistently favors viewing with a particular eye likely has better vision on that side, and more detailed testing is indicated.

A similar technique can be used to evaluate for eye movement and strabismus (Fig. 2). Using a thumb to cover one eye of the child (a patch or parent’s hand can be used as well), the examiner holds a toy or penlight and checks that the child fixes on the object with the uncovered eye and follows the object as it is moved. The examiner then moves the thumb to the other eye to check for strabismus. If the child has been focused on the light or object, in the absence of strabismus, the newly uncovered eye should not move. Strabismus should also be suspected if the light reflex does not fall on the center of both pupils. If one is concerned about the presence of esotropia (eye turning in), a light or lighted toy can be held in front of the child and then brought closer. In some cases esotropia is only revealed with focusing on near objects.

A third test is needed to check for intermittent exotropia (eye turning out). Intermittent exotropia often occurs only with viewing objects from a distance. This condition is brought out by presenting the child with a toy or object at a distance and looking at the corneal light reflex. This reflex can be difficult to assess; if a history of an intermittent out turning of an eye is obtained, the child should be referred to a pediatric ophthalmologist.

In practice, certain parts of the examination may be reasonably omitted in the emergency department setting, because a young child who has eye

Fig. 2. Fixation examination in children 4 months and older. Use of a toy will often help with the examination. Use the thumb to cover each eye in turn to check for fixation. Move the thumb from one eye to the other to check for strabismus (cover testing). (From Drack AV. Pediatric ophthalmology. In: Palay DA, Krachmer JH, editors. Primary care ophthalmology. 2nd edition. Philadelphia: Mosby; 2005. p. 234; with permission).
pain or discomfort may not be cooperative with some or all of the examination. As long as the child is opening both eyes for any length of time, fixation and corneal light reflex can be assessed.

If the child has pain and the history suggests a corneal abrasion or foreign body (and not a ruptured globe), instilling a topical anesthetic may decrease eye pain and allow for an easier examination. The intervention may also be diagnostic as the relief of symptoms isolates the pathology to disruption of the conjunctiva or corneal surface.

In children with a large amount of swelling around the eye, it may be necessary to retract the eyelids to perform the examination. If the history suggests significant trauma and globe rupture is a possibility, it is important to avoid putting pressure directly on the eye by placing the examiner’s thumbs on the infraorbital and supraorbital rims and separating the lids. In some instances, lid retractors may be needed to examine the eye. If only one retractor is being used, it is most helpful to apply it to the upper lid. Cotton swabs can also be used to open the eyes. In this method, one swab is placed on the upper eyelid and one on the lower. The swabs are then rotated toward the eyeball, the upper swab rotated down, and the lower swab rotated up. Simultaneous with rotation, the swabs are moved toward the orbital margins [5]. This method should not be used if trauma is suspected because it places pressure on the eyeball.

**Verbal children**

The examination becomes much easier in children who can talk and allow for objective testing of visual acuity. In young children who do not yet know numbers or letters, the Allen card or other calibrated picture tests may be used (Fig. 3). Before testing, one should have the child identify the objects up close. The Tumbling E chart is also commonly used. With this test, one should make the instructions clear to the child. Some recommend telling the child that the E is a table, sometimes right side up, sometimes on its side, and sometimes upside down [4]. The examiner should have the child point the direction legs of the table are pointing. As in adults, each eye should be tested individually; however, in children, special attention should be paid to whether they are using the covered eye to see. Children tend to look around the hand-held eye covers, and it may be necessary to patch the child. Vision should be 20/50 or better at a distance in children aged less than 5 years but 20/30 or better at near distance in all ages. In the absence of nystagmus, there should be no significant improvement in acuity viewing with both eyes open. One should remember children have short attention spans. There is no need to start on the line with the largest figures or to have the child read every figure in a given line.

With patience, an understanding of children’s development, and a few techniques, it is possible to perform an eye examination on children in the emergency department. The following sections discuss an array of disease
processes that are particular to, or more common in, the pediatric age group.

**Conjunctivitis**

*Ophthalmia neonatorum (neonatal conjunctivitis)*

Ophthalmia neonatorum is defined as conjunctivitis within the first month of life. There are three main types of neonatal conjunctivitis: chemical, bacterial, and viral. Although these entities may present with similar symptoms, the timing of the development of symptoms can often be a useful diagnostic clue. Chemical conjunctivitis secondary to perinatal ocular prophylaxis generally presents within the first 24 to 48 hours of life [6]. Erythromycin ointment is the agent most commonly used today and only rarely causes chemical conjunctivitis. Silver nitrate was used in the past and has been more frequently associated with chemical conjunctivitis. Infants with chemical conjunctivitis typically present with bilaterally inflamed lids and
watery discharge. Gram stain reveals white cells without bacteria. Treatment initially is supportive and involves the discontinuation of any ophthalmic medications and observation, with an expected resolution of symptoms within 48 hours. If no improvement is seen, a culture should be obtained and topical antibiotic therapy initiated, with care to avoid whatever agent was used for initial prophylactic therapy.

The epidemiology of neonatal infections is related to the transmission of organisms at the time of delivery; therefore, pathogens found in the genital tract and enteric system should be suspected. *Chlamydia trachomatis* is more commonly acquired from the birth canal than are *Neisseria gonorrhoeae* and herpes viruses (herpes simplex virus [HSV]) [7]. In addition, gram-negative enteric organisms and several staphylococcus and streptococcus species may also be acquired peri- and postnatally. Gonorrheal infections typically occur 2 to 5 days after birth but can be delayed if neonatal prophylactic therapy provides partial suppression. Chlamydial infections present slightly later, often between 5 and 14 days of life [6].

Physical examination findings can be helpful with diagnosis, but there is tremendous overlap of symptoms from different pathogens. Accurate diagnosis on the basis of physical examination alone is challenging and often requires supplementary laboratory data. Gonorrheal infections are classically characterized by a hyperacute mucopurulent discharge with lid edema, bulbar conjunctivitis, and chemosis. Chlamydial infections can also present with copious discharge but more commonly are characterized by palpebral conjunctival injection and inflammation with less associated lid edema and thick discharge [6,8]. A statim Gram stain and culture, including chocolate agar, should be obtained to aid in the diagnosis but should not delay the initiation of therapy when a high clinical suspicion for disease is present. In addition to Gram stain and culture, Giemsa stain, direct fluorescent antibody, ELISA, and polymerase chain reaction can be used to diagnose chlamydial infections, and laboratory investigation should be guided based on method availability [9]. Intracellular gram-negative diplococci are consistent with gonorrheal infection and constitute an ocular emergency because this organism can penetrate through and ulcerate the cornea, rapidly causing blindness [8]. An ophthalmology consult should be obtained immediately without delay in therapy. Current recommendations for treatment are a single dose of intravenous or intramuscular ceftriaxone with admission and hourly saline eye lavage. The infant should simultaneously be covered for chlamydial disease until cultures are negative using oral erythromycin therapy to treat ophthalmic disease and prevent the late onset of chlamydial pneumonitis [7,8].

*Staphylococcus aureus, Streptococcus epidermis, Haemophilus influenzae, Escherichia coli,* and *Pseudomonas* are other causes of neonatal conjunctivitis and typically present from 5 to 7 days of life. Clinical findings are often indistinguishable from that of other pathogens. Diagnosis is by Gram stain and culture, and polymyxin/bacitracin/neomycin topical ointment is generally accepted as standard treatment. Diagnosis of typeable *Haemophilus*
*Haemophilus influenzae* conjunctivitis is an exception and should be treated with systemic antibiotics, with consideration given to a full septic evaluation before parenteral antibiotic administration.

Neonatal conjunctivitis caused by HSV, typically, although not exclusively, HSV-2, may also be acquired through the birth canal, and ocular manifestations may be the only presenting symptoms of neonatal herpetic infections [6]. Clinical suspicion should be elevated with a maternal history of infection, vesicular blepharitis, or the presence of ocular dendritic ulcers with fluorescein staining. Diagnosis is made by immunofluorescence, smear, or culture. Treatment involves both topical and systemic parenteral acyclovir and the avoidance of steroids. Full septic evaluation should be performed in the neonate with HSV infection [10].

**Childhood conjunctivitis**

Acute conjunctivitis is the most common eye disorder in young children and is the most frequent ophthalmologic complaint seen in the pediatric emergency department [11]. To date, there are no evidence-based guidelines for the diagnosis and empirical treatment of conjunctivitis [12]. Bacterial infections are predominant and are chiefly caused by one of three pathogens—non-typeable *Haemophilus influenzae*, *Streptococcus pneumoniae*, and *Staphylococcus aureus* [7,11]. The clinical course of bacterial conjunctivitis generally has an abrupt unilocular onset, with spread to the opposite eye within 48 hours [13]. Tearing and irritation are the initial symptoms, followed by mucopurulent discharge, typically with a history of crusting or gluing of the eyelashes. Diffuse erythema of the bulbar and palpebral conjunctivae is generally present, whereas preauricular lymphadenopathy is not [14].

Laboratory studies to determine the causative organism are usually reserved for severe cases and those unresponsive to initial treatment. Empiric treatment is commonplace, particularly when a history of sticky eyelids is obtained in conjunction with a physical finding of purulent discharge [12]. Treatment typically involves erythromycin ointment, bacitracin-polymyxin B ointment, or topical fluoroquinolones [7]. Several clinical associations can also help guide diagnosis and subsequent treatment. Conjunctivitis-otitis syndrome is common, occurring about 25% of the time, and is most often associated with non-typeable *Haemophilus influenzae* infections [11,15]. In this scenario, monotherapy with systemic antibiotics is indicated, and a topical agent is not needed [16,17]. Several studies suggest that if *Haemophilus influenzae* is recovered from a culture, or if the patient has a history of recurrent otitis media, systemic treatment should be initiated even in the absence of acute otitis media in the hope of preventing its development [17].

Another common cause of pediatric conjunctivitis is viral illness. The overall frequency of pediatric viral illness is extremely high, but the presence of conjunctivitis in systemic pediatric viral disease varies. The most common
type of viral conjunctivitis in children is adenoviral conjunctivitis, which can present as an isolated condition or as part of a viral syndrome [7]. Adenovirus can cause a nonspecific acute conjunctivitis characterized by red profusely watery eyes, or more severely, epidemic keratoconjunctivitis if corneal involvement is present. Pharyngoconjunctival fever caused by adenovirus is common in children and presents with the triad of pharyngitis, fever, and conjunctivitis, as the name implies. The typical course lasts 2 weeks and often begins with unilateral involvement, becoming bilateral within several days, with preauricular lymphadenopathy [18]. Although the typical course of pharyngoconjunctival fever is self-limited with an excellent prognosis, the same adenovirus types can also cause the rarer but more serious disseminated adenoviral disease which results in multisystem organ failure and death [18,19]. Upper respiratory tract infections caused by rhinovirus, enterovirus, and influenza virus are accompanied by a self-limited conjunctivitis less than 50% of the time, and less than one third of respiratory syncytial virus infections are accompanied by conjunctivitis. Conjunctivitis is also commonly associated with measles, although this pathogen is now rare in the United States [19].

The diagnosis of adenoviral conjunctivitis remains primarily clinical. Conjunctival hemorrhage can occur with adenoviral infection, as can punctate corneal epithelial defects; therefore, the slit lamp examination is an important part of the diagnostic evaluation, although it is often difficult to perform on a young patient. An ideal laboratory study does not yet exist. Viral cultures are epidemiologically useful, but delayed results have little use in the emergency department setting. Enzyme immunoassay and polymerase chain reaction tests are rapid, but the sensitivity varies considerably. Treatment options are also limited and are largely supportive because there is no proven effective treatment for adenoviral conjunctivitis [20,21]; however, topical antibiotics are often prescribed to prevent bacterial superinfection. Corticosteroids should be avoided in treating most cases of pediatric adenoviral conjunctivitis and should only be administered under the care of an ophthalmologist. In fact, the prescription of ophthalmic steroids in general in the emergency department should be limited, because steroids can be devastating in the presence of herpetic infections, which must always be considered and effectively ruled out.

Herpetic ocular infections outside of the neonatal period are typically from HSV-1 [6]. Herpetic keratitis with its classic dendritic pattern with fluorescein staining may be present, is most often unilateral, and is sometimes associated with vesicles in the distribution of the ophthalmic branch of the trigeminal nerve, involving the forehead, periorbital area, and tip of the nose [22]. More commonly, the clinical presentation of HSV conjunctivitis is nonspecific, although always painful, and very similar to other etiologies of conjunctivitis previously discussed. Treatment of HSV ocular infection, most often with a topical antiviral agent, should involve an ophthalmologist.
Orbital and periorbital cellulitis

Orbital and ocular adnexal infections are more common in children than adults and must be accurately distinguished from periorbital infections, because the pathogenesis, treatment, and potential severity of sequelae vary considerably. Knowledge of the region’s anatomy helps one to clinically distinguish orbital from periorbital infections and aids in understanding the pathophysiology and the potential for spread of infection of each of these two entities. Orbital infections are defined by their location relative to the orbital septum, which is a thin membrane that extends from the periosteum and reflects into the upper and lower eyelids [23]. The septum separates the periorbital soft tissues (preseptal region) from the orbital space (septal) and provides a barrier to the spread of infection between the two regions. Preseptal processes do not directly progress into the septal space, nor do septal infections directly spread into the preseptal space [23,24]; however, infection can also travel through the valveless venous drainage system of the midfacial region involving the eye cavity and the ethmoid and maxillary sinuses, thereby allowing for the indirect spread of infection in an anterograde and retrograde fashion [25].

Another important anatomic consideration is the relationship between the sinus cavities and the orbit. The eye is surrounded by paranasal sinuses on three of its four walls. The floor of the frontal sinus is the roof of the orbit, and the roof of the maxillary sinus is the floor of the orbit. The medial border of the eye is formed primarily from the extremely thin lamina papryacea of the ethmoid bone. Infection can spread from the paranasal sinuses to the bone, forming osteitis or subperiosteal abscesses, and into the orbital space, producing an orbital abscess or orbital cellulitis (Fig. 4) [24,26]. These anatomic considerations help to explain the typical pathogens found in orbital cellulitis. The periorbital area is protected from the paranasal sinuses by the orbital septum; therefore, it is far less susceptible to infection by sinus pathogens. Infections in the periorbital area are usually secondary to skin pathogens and are often associated with soft tissue injuries such as insect bites.

Fig. 4. Orbital cellulitis. (From Greenberg MF, Pollard ZF. The red eye in childhood. Pediatr Clin North Am 2003;50:106; with permission).
bites or the spread of local infection (impetigo, hordeolum, chalazion, dacryocystitis) [27].

Periorbital cellulitis is much more common than orbital cellulitis [25]. It presents clinically with erythema, induration, tenderness, or warmth of the periorbital tissues. Signs of systemic illness are often absent, although fever may be present, particularly if bacteremia served as the origin of the cellulitis. Extraocular motion is not affected and should be full. In fact, decreased movement of the eye is one of the cardinal features of orbital cellulitis, along with proptosis, decreased visual acuity, chemosis, and papilledema [8,23]. Orbital cellulitis is also associated with erythema, pain, and swollen eyelids, but the eyelid swelling of orbital cellulitis can be differentiated from that of periorbital cellulitis in that it will not extend beyond the superior orbital rim onto the brow [27]. This limitation of upper eyelid swelling is due to the extension of the orbital septum onto the periosteum of the inferior margin of the superior orbital rim, which effectively provides a structural barrier limiting the degree of upper eyelid swelling in orbital cellulitis.

Distinguishing between these two clinical entities is paramount. If one is unable to do so clinically, a CT scan should be obtained, as should ophthalmology consultation [25]. If CT scanning demonstrates sinus disease as a likely etiology of orbital cellulitis, otorhinolaryngology should also be consulted because surgical drainage may be necessary. Any child with orbital cellulitis must be admitted for parenteral antibiotics and close observation among a multidisciplinary team, with or without surgical intervention. The antibiotic choice should be aimed at the most likely pathogens, typically, respiratory pathogens and anaerobes originating from the paranasal sinuses. Ampicillin/sulbactam or cefuroxime with clindamycin or metronidazole are reasonable choices because they appropriately target the most common organisms, such as Streptococcus pneumoniae, non-typeable Haemophilus influenzae, group A streptococcus, Staphylococcus aureus, and anaerobic organisms [23].

For children with periorbital cellulitis, skin trauma is the most likely etiology. Antibiotics targeted at gram-positive organisms should be administered, because staphylococcus and streptococcus species are the most likely cause of post-traumatic periorbital cellulitis [23]. These patients should be followed up closely for any progression of symptoms. Primary bacteremia is another etiology of periorbital cellulitis but is rare due to effective vaccination against Streptococcus pneumoniae and Haemophilus influenzae [27]. If bacteremia is suspected as a source for infection, particularly in a child aged less than 3 months or in an unvaccinated or newly immigrated patient, Streptococcus pneumoniae and Haemophilus influenzae should be suspected. Any child aged less than 2 years or who has signs of systemic illness should be admitted for parenteral antibiotics and close observation. A full septic evaluation, including lumbar puncture, should be strongly considered before antibiotic administration in any toxic appearing child, or in the presence of any signs or symptoms suggestive of meningitis.
Another special consideration involves immunocompromised children, including children with diabetes. These children should immediately be referred to an ophthalmologist for evaluation because mucormycosis presents with eyelid erythema and is a diagnosis that generally requires surgical debridement [25].

**Lacrimal system infections**

Infections of the lacrimal system are named according to the location of infection. Infection of the nasal lacrimal duct, located between the medial canthus of the eye and the nasal bridge, is known as dacryocystitis and can occur in the setting of acute or chronic obstruction of the duct (Fig. 5). Often, a history of watery or even mucopurulent discharge from the eyes can be elicited, followed by the development of erythema, swelling, and tenderness over the lacrimal sac. The major complication of dacryocystitis is periorbital cellulitis [26] and, less commonly, orbital cellulitis [27] or cavernous sinus thrombosis [23]; meningitis, brain abscesses, and sepsis can also occur, although rarely. Diagnosis must be prompt, and treatment should include oral antibiotics. The most common pathogens in children with acute dacryocystitis are *Staphylococcus epidermidis* and *Staphylococcus aureus* [23]. Any child with dacryocystitis who appears ill or toxic should be admitted for parenteral antibiotic therapy [27].

Another infection of the lacrimal system is dacryoadenitis. This infection of the lacrimal gland is located in the supratemporal orbit. The gland is composed of two lobes. The palpebral lobe is easily visualized with eversion of the superior lid, but the orbital lobe cannot be directly visualized on physical examination. Dacryoadenitis may present as an acute or chronic problem. Acute disease is characterized by the abrupt onset of pain.

Fig. 5. Acute dacryocystitis. Maximal swelling nasally below the medial canthal ligament. (From Greenberg MF, Pollard ZF. The red eye in childhood. Pediatr Clin North Am 2003;50:108; with permission).
swelling, and erythema of the supraorbital region, often associated with chemosis, conjunctivitis, and mucopurulent discharge, and sometimes associated with limited ocular mobility, proptosis, fever, and malaise. More chronic infections typically present with swelling of the superior lid. Mild ptosis may be present secondary to the swelling, but pain, erythema, and fever are not present.

The treatment of dacryoadenitis is dependent on the acuity of the presentation and the most likely etiology. Imaging is often not necessary, although CT scanning can help to make the distinction between orbital cellulitis and dacryoadenitis if the orbital lobe of the gland is involved and clinical distinction is difficult. Acute dacryoadenitis is most commonly associated with viral infections, and treatment is supportive. Bacterial pathogens should be suspected if the discharge is mucopurulent. Cultures should be obtained while initiating treatment to cover the most common pathogens until culture data become available. A first-generation cephalosporin is generally recommended; however, an increasing prevalence of ocular methicillin-resistant Staphylococcus aureus (MRSA) has recently been reported. Choosing an oral antimicrobial agent to best fit the MRSA susceptibility profile within your institution is prudent [28,29].

Congenital

Nasal lacrimal duct obstruction

The most common congenital ophthalmologic finding in newborns is nasal lacrimal duct obstruction. Tears are produced in the lacrimal gland which rests within the temporal portion of the superior lid. They then circulate over the eye toward the punctum located in the nasal corner of the eye where the two lid margins unite. Typically, tears drain through the punctum and canalicular system into the nasolacrimal sac and then into the duct which drains intranasally through the valve of Hasner. When the drainage path is obstructed, most commonly at the level of the valve of Hasner, patients present with watery discharge from the eye, often bilaterally [21]. On further inspection, the tear lake in the inferior portion of the lid is often elevated. If bacterial superinfection exists, a chronic mucopurulent discharge is present, with parents commonly reporting lid adherence. If this adherence persists, symptoms may progress to include conjunctival injection with thickening of the periorbital skin. The examination of any newborn with these complaints should involve carefully applied pressure with a cotton tip to the region of the nasolacrimal sac. If nasal lacrimal duct obstruction is present, reflux of mucopurulent material from the punctum may occur. Careful examination of the skin that overlies the drainage system is also important, because identification of a bluish hued palpable mass is indicative of a mucocele, specifically, a cyst of the nasal lacrimal duct also known as a dacryocele [21]. Simple nasal lacrimal duct obstruction should not be
associated with any photophobia, ocular cloudiness, or abnormal appearance of the red reflex. If any of these findings are present, the diagnosis of nasal lacrimal duct obstruction should be questioned, and congenital glaucoma or cataracts should be considered.

Treatment of nasal lacrimal duct obstruction in the newborn is simply supportive if no superinfection or dacryocele is suspected. Parents should be instructed to apply gentle massage over the nasal lacrimal duct with rapid downward motions three to four times daily to facilitate opening of the valve of Hasner. After the age of 6 months, the patient should be referred to an ophthalmologist, because obstructions rarely resolve on their own beyond the first several months of life and often require surgical probing [21]. If suspicion of a dacryocele exists, the patient should rapidly be referred to a pediatric ophthalmologist and otolaryngologist because obstructive intranasal cysts are often associated and require rapid intervention. The presence of mucopurulent discharge warrants the administration of topical antibiotics for 1 to 2 weeks in conjunction with daily massage as described previously. If this regimen does not clear the discharge, the patient should be referred to an ophthalmologist for further evaluation, independent of age. Continued infection in the lacrimal sac is associated with preseptal cellulitis, a more serious condition that often requires hospitalization in this age group.

**Congenital cataracts**

A cataract is an opacity of the lens of the eye requiring prompt diagnosis and treatment to prevent partial or complete blindness. Congenital cataracts can be present at birth and associated with certain congenital infections such as rubella, toxoplasmosis, HSV, or cytomegalovirus [30]. They can also develop in the first several months of life secondary to several metabolic conditions, such as galactosemia or peroxisomal disorders, or in genetic conditions such as trisomy 21 or Turner syndrome [21].

The clinical presentation of infants with cataracts is dependent on the density of the opacification and the presence in one or both eyes. Leukokoria is caused when the cataract is dense enough to prevent a significant amount of light from penetrating through the cornea to the retina (see Fig. 1). The red reflex is abnormal and may even be absent if the cataract is severe. Nystagmus or strabismus may also be noted if the cataract develops within the first several months of life. Vision may be mildly to severely decreased. In severe cases in which vision is absent, the infant may not even spontaneously open his or her eyes. In moderate cases, the infant may be noted to squint in bright sunlight in an effort to reduce the glare resulting from the reduced ability of the pupil to constrict [21].

Treatment of congenital cataracts should be initiated emergently through a pediatric ophthalmologist, because the first several months of life are critical to the development of the visual axis.
**Congenital glaucoma**

Pediatric glaucoma is divided into primary and secondary types depending on the presence of isolated angle malformations (primary) versus other underlying ocular abnormalities (secondary) [30]. Both types may be present at birth (congenital) or develop at any age (infantile or juvenile). The common finding with any form of glaucoma is increased intraocular pressure, which, if left undiagnosed and untreated, can lead to optic nerve damage and vision loss. Additional damage, such as large refractive errors, astigmatisms, strabismus, and amblyopia, may occur as a result of congenital or infantile glaucoma, because the visual system is undergoing crucial stages of development during infancy, and any disruption to the visual axis may have multiple sequelae [30].

Forty percent of cases are present at birth and 85% by age 1 year; however, the age of diagnosis varies from birth to late childhood. The most common finding in patients who have congenital glaucoma is excessive tearing, also known as epiphora, as well as photophobia and some degree of blepharospasm [30]. Corneal enlargement or asymmetry (when disease is unilateral) is often present, and a corneal diameter of greater than 12 mm in an infant younger than 1 year of age should prompt urgent referral to a pediatric ophthalmologist [30]. Other findings include corneal clouding, conjunctival injection, corneal edema, ocular enlargement, and ocular nerve cupping observed on fundoscopic examination [21].

Treatment of glaucoma in infants and children is almost always primarily surgical, complemented by medical therapy with topical or oral pressure-lowering agents. Prognosis is generally better the later the onset of symptoms, because the structural anomaly is typically less severe [30].

**Misalignment**

Ocular misalignment, generally referred to as strabismus, is not uncommon in newborns and young children and may be of enough concern to the parents to prompt an emergency room visit. It is important to distinguish normal misalignment from more worrisome clinical presentations. Newborns commonly have an ocular instability that is characterized by variable, intermittent ocular misalignment throughout their first several months of life. This misalignment is most commonly secondary to immaturity of the extraocular muscles and self-resolves by 3 to 4 months of life [21]. If the deviation is constant, or if it is bilateral, the patient should be referred to a pediatric ophthalmologist for further investigation, because these patterns may be more consistent with significant pathology such as primary neurologic or oncologic processes.

Patients with congenital strabismus typically have normal eye movements for the first several months of life and then develop the tendency for one or both eyes to deviate [21]. If this deviation is present without interruption of the visual axis, it is referred to as a “manifest strabismus.” More specifically,
it is termed esotropia if there is inward deviation of the eye or exotropia if the deviation is outward. The examination techniques for the evaluation of strabismus described earlier in this article, specifically the “cover uncover test,” may elicit a latent strabismus also known as a “phoria” that is only present when fixation is interrupted by covering one eye [31]. Children who have either manifest or latent strabismus should be evaluated by an ophthalmologist because these conditions can lead to amblyopia, although much less commonly with phorias than tropias [31].

The emergency room physician should always rule out a sixth nerve palsy that could mimic congenital esotropia, particularly if accompanied by other signs of increased intracranial pressure such as nausea, vomiting, lethargy, and sunsetting of the eyes [8]. Similarly, third nerve palsies should be considered when evaluating a child with an exotropia [3,16]. In general, emergent presentations of cranial nerve palsies or mechanical restriction due to orbital fractures, cellulitis, masses, or other intracranial processes can effectively be ruled out by full extraocular muscle movements [8].

Oncology

Retinoblastoma is the most common primary intraocular malignancy of childhood and frequently presents with leukokoria, often detected by a parent who may seek medical evaluation in the emergency department. The white pupil is actually the tumor itself visualized through the pupil and vitreous [21]. The tumor may be unilateral, typically associated with a spontaneous mutation, or bilateral, almost always heritable. These children may also present with a unilateral fixed and dilated pupil, visual changes, a red and painful eye, proptosis, or different colored irises, also known as heterochromia iridis [21]. Any child with a white pupil or any other findings suspicious for retinoblastoma should be immediately referred to an ophthalmologist for a complete ocular examination, typically performed under anesthesia.

Other tumors that may present as orbital masses with proptosis include rhabdomyosarcoma, Langerhan’s cell histiocytosis, acute myeloid leukemia, metastatic Ewing’s sarcoma, Burkitt’s lymphoma, or neuroblastoma [26]. Neuroblastoma can also present with the rare ocular finding of opsoclonus/myoclonus. This condition is often referred to as “dancing eyes,” describing the simultaneous presence of rapid irregular eye movements and involuntary twitching of the eyelids, and is believed to be secondary to an autoimmune reaction. When present, opsoclonus/myoclonus should prompt an immediate evaluation for neuroblastoma, because this is the most commonly associated pediatric tumor.

References


