COMMON PEDIATRIC ANTERIOR SEGMENT DISORDERS AND DISEASES

Diligent follow-up and comanagement with other practitioners is vital for many conditions.

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This article focuses on frequently seen pediatric ocular conditions related to the anterior structures of the eye, reviews secondary complications that can occur with these diagnoses, and revisits the treatment and/or management protocols for each condition.

EYELID

Childhood Ptosis

A common diagnosis that can be found in children and infants is ptosis. It is important to differentiate congenital from acquired ptosis, as the acquired form can be associated with systemic conditions. Fortunately, congenital ptosis is far more prevalent. A study by Griepentrog et al followed 107 children who were diagnosed with childhood ptosis for 40 years and found the incidence of simple congenital ptosis to be 75.7%.[1]

A thorough case history can help guide your diagnosis in differentiating between congenital and acquired ptosis; recent onset of ptosis, changes in lid position, overall health of the child, or association with any additional ocular findings are all concerning issues that may be associated with an acquired ptosis and therefore warrant a referral for further investigation.

In managing patients with ptosis, two secondary complications that can arise are deprivation amblyopia due to lid placement and refractive amblyopia from resulting astigmatism. Watch for a chin-up posture used to help compensate for lid positioning, and use your amblyogenic refractive diagnosis criteria when deciding which patients need refractive correction. Remember that glasses wear is the start of visual rehabilitation and

that some children may need adjunctive amblyopia therapy.

If no amblyopia is present or amblyopia has been successfully treated, surgical correction of mild to moderate amblyopia can be delayed until the patient is 3 to 5 years of age, when long-term surgical outcomes are more favorable.[2]

Nasolacrimal Duct Obstruction

The most common lacrimal system disorder in pediatrics is a nasolacrimal duct obstruction (NLDO); approximately 5% of full-term newborns are affected.[2] This condition occurs due to a persistent membrane at the distal valve of Hasner. Typical signs of an NLDO are epiphora and an increase in mucous discharge within the first few months of life. These symptoms are usually chronic in nature but intermittent from day to day.[2]

Most infants, approximately 90%, have resolution of their symptoms within the first 9 to 12 months. As a result, a conservative nonsurgical approach is generally used in the initial stages of treatment. Digital massage of the lacrimal sac is often recommended, with the goal of forcing fluid through the nasolacrimal duct system to open any existing obstruction.

Topical antibiotics can be recommended if there is significant discharge, but it is important the family understand that the benefit of topical antibiotics is limited and cannot resolve the underlying etiology; this means that the patient will likely need intermittent use of the antibiotic over the course of the condition.

In the event that an infant does not improve with time or conservative treatment, nasolacrimal duct probing is recommended. This procedure has a high success rate and can be done in the office or under general anesthesia, depending on the age of the child and the preference of the ophthalmologist.[2]

TABLE 1. Incidence of Other Ocular Disorders in Children Diagnosed as Having Ocular Coloboma

OCULAR DISORDER	PATIENTS, NO. (%)
Reduced Visual Acuity[a]	9 (27)
Unilaterally reduced visual acuity	
Unilateral coloboma	6
Bilateral coloboma	0
Bilaterally reduced visual acuity	
Unilateral coloboma[b]	1
Bilateral coloboma	2
Amblyopia	11 (33)
Strabismus	10 (30)
Esotropia alone	4
Exotropia alone	1
Hypotropia alone	1
Mixed	4
Structural Abnormalities	5 (15)
Microphthalmia	4
Anophthalmia	1
Other Diagnoses	
Anisometropia	3
Nystagmus	2
Cataract	2
Retinal detachment	2
Hypoplasia of the optic nerve	1

[a]BCVA < 20/60; [b]Patient developed retinitis pigmentosa

Adapted from Nakamura KM, Diehl NN, Mohney BG. Incidence, ocular findings, and systemic associations of ocular coloboma. *Arch Ophthalmology*. 2011;129(1):69-74.

Periocular Hemangiomas

Infantile hemangiomas are composed of proliferating capillary endothelial cells. These tumors are of benign etiology, but it is their location that can be concerning. Due to their rapid growth phase early in infancy, these hemangiomas can grow a considerable amount, occupying the surrounding space and altering the function of a nearby organ. Periocular hemangiomas comprise 12% of all infantile hemangiomas,[2] with 62.5% of these hemangiomas causing a secondary ocular complication.[3]

The most common ocular sequalae are mechanical ptosis and induced irregular astigmatism, either of which can lead to amblyopia if left untreated. Routinely in practice these patients are comanaged with a pediatric dermatologist to monitor the growth and treatment of the hemangioma.

There is evidence to support the use of beta-blockers, either topical or oral, as first-line treatment to halt progression during the rapid growth phase. These infants should be seen frequently to monitor for changes in lid position as well as refractive status.

IRIS AND PUPIL

Iris Coloboma

The term *typical iris coloboma* refers to the classic keyhole shaped pupil. This iris architecture results from incomplete closure of the embryonic fetal fissure during the fifth week of gestation. Typical iris colobomas can be associated with colobomas of the lens, ciliary body, retina, choroid, and optic nerve.[2]

It is important to know that *atypical* iris colobomas exist as well. Atypical colobomas are unrelated to

TABLE 2. Associated Nonophthalmic Disorders Diagnosed in Children With Ocular Coloboma

DISORDER	PATIENTS, NO. (%) TOTAL	DIAGNOSED AS HAVING CHARGE SYNDROME
Abnormal Development	**12 (36)**	
Developmental delay	6	3
Learning disability	4	0
Profound mental retardation	1	1
Moderate mental retardation	1	0
Heart Anomalies	**7 (21)**	
PDA with ASD	2	0
Ventricular septal defect	1	1
ASD	1	1
ASD with pulmonary valve stenosis	1	0
PDA	1	0
PDA with PFO	1	0
Ear Anomalies[b]	**7 (21)**	
Recurrent otitis media[a]	7	4
Hearing loss	4	3
Eustachian tube dysfunction	3	1
External ear deformity	3	1
Deafness	1	0
Seizures	**6 (18)**	2
Skeletal Anomalies	**6 (18)**	
Scoliosis	4	1
Digital malformations[c]	3	0
Kyphoscoliosis	1	1
Kyphosis	1	0
Osteogenesis imperfecta	1	0
Urogenital Anomalies	**5 (15)**	
Vesicoureteral reflux	3	0
Cryptorchidism	1	0
Micropenis	1	0
Other Anomalies		
Cleft palate	2	1
Short stature	2	1
Hypogonadotropic hypogonadism	1	1
Choanal atresia	1	0
Laryngomalacia/tracheomalacia	1	0

[a]Recurrent otitis media is defined as three episodes of acute otitis media within 6 months or four or more episodes within 1 year; [b]Some children have multiple anomalies; [c]Digital malformations included clindodactyly, camptodactyly, and bifid toes.
Abbreviations: ASD, autism spectrum disorder; PDA, pathologic demand avoidance; PFO, patent foramen ovale
Adapted from Nakamura KM, Diehl NN, Mohney BG. Incidence, ocular findings, and systemic associations of ocular coloboma. *Arch Ophthalmology*. 2011;129(1):69-74.

fetal fissure closure; they can occur at any location on the iris and are not associated with additional colobomas of the eye.[2]

A study by Nakamura et al looked at the incidence of typical iris colobomas in children and identified common ocular associations and systemic conditions. The three most common ocular findings associated with iris coloboma (Table 1) were reduced visual acuity (27%), amblyopia (33%), and strabismus (30%).[4] With the prevalence of these associations, infants and children with iris coloboma warrant frequent follow-up to monitor their refractive error and visual development closely.

Systemic conditions can also be associated with iris colobomas (Table 2). Remember that a thorough case history is important, as is communicating with the child's pediatrician so that further testing and comanagement can be completed if warranted.

INFLAMMATORY DISEASE

Vernal Keratoconjunctivitis

Vernal keratoconjunctivitis (VKC) is a chronic bilateral seasonal allergic inflammatory disease of the eye. It is three times more likely to affect males, with onset happening most frequently before age 10 years. This disease is usually self-limiting but of unknown duration. In patients with recurrent disease, it can lead to long-term visual consequences and has been found to cause keratoconus and limbal stem cell deficiency.

One of the main issues with VKC is understanding the etiology. It is more complex than a type 1 hypersensitivity reaction; research is ongoing investigating the cells and mediators that are involved in initiating and perpetuating the inflammation.[5]

Several drugs are available to treat signs and symptoms of this condition, but unfortunately most of these are palliative and do not eliminate the complex immune response that leads to frequent recurrence. Steroids are highly effective at controlling acute and chronic responses, but long-term steroid use has well-known complications. Tacrolimus and cyclosporine are steroid-sparing agents that manage inflammation well, but more research is needed to help determine the appropriate dosage and duration.[6]

The treatment and management of patients with VKC can be complex. Each patient needs a tailored medication plan that will likely be altered throughout the year as signs and symptoms fluctuate.

Blepharokeratoconjunctivitis

Blepharokeratoconjunctivitis (BKC), also known as staphylococcal marginal keratitis, is an

inflammatory condition of the eyelids that progresses to involve the cornea. It is an underdiagnosed disorder that, if left untreated, can cause long-term visual consequences. The top three symptoms identified by Rodriguez-Garcia et al were photophobia, discharge, and grittiness, and the most common signs included blepharitis and hyperemia.[7]

What this manifests as, clinically, is a child with chronic red eyes that vary intermittently in severity. Hammersmith et al conducted a study to identify the incidence, symptoms, clinical signs, and treatment outcomes of BKC. Surprisingly, their study found that the mean age of onset was 4.1 years, with a relatively equal distribution between the sexes. In addition, chalazia were a top clinical finding (73%), and findings were bilateral in 91% of patients.[8]

Treatment for this condition is twofold: (1) decrease the bacterial load and (2) reduce the inflammation.

The most important treatment for patients to maintain throughout this entire process is proper lid hygiene. Good lid hygiene will be most effective at treating the underlying etiology. This includes warm compresses, lid massages, and lid scrubs.

To treat the signs and symptoms, erythromycin works well at targeting the *Staphylococcus* organism, and the ointment form stays on the eyelids for a prolonged period of time. When corneal involvement is present, prednisolone acetate ophthalmic suspension 1% (Pred Forte, Allergan, or generic) is quite effective at removing the current inflammation and eliminating vessel growth onto the cornea. Small dosages work well (once or twice daily), and the medication can be gradually tapered as signs and symptoms improve.

Once corneal involvement is present there is a risk for corneal scarring, which can lead to visual impairment and amblyopia in 7% of children.[8] Close monitoring of these patients is critical to implement treatment as soon as possible in order to prevent lifelong visual consequences.

MANAGE YOUR YOUNG PATIENTS WITH CONFIDENCE

Anterior segment findings in a child or infant can make a difficult examination even more challenging. Remember that the visual development timeframe is critical in pediatrics, which means close monitoring for changes in refractive error and visual acuity are most important. ■

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