Pediatric Ophthalmology Maintenance of Certification for the Retinal Specialist

By Scott A. Larson, MD

As part of the “Road to Recertification” article series in New Retina MD, Scott A. Larson, MD, provides an overview of some of the pediatric ophthalmology topics that retina specialists may want to review in more detail for Maintenance of Certification. As with every article in this series, Dr. Larson’s overview is not meant to take the place of a comprehensive review course; rather, its purpose is to highlight some key areas within the oculoplastics subspecialty and to encourage a more thorough review prior to taking the Demonstration of Ophthalmic Cognitive Knowledge exam.

- Diana V. Do, MD

Diagnostic Tests

Vision testing in the pediatric ophthalmology and strabismus clinic may differ somewhat when compared to the retina clinic. A large part of the examination is devoted to testing ocular motility, alignment, and visual sensory status. It may be helpful to review some of the tests that may not be used on a daily basis by a retinal specialist.

**Cover testing.** This test is used to assess the binocular alignment with suspected strabismus and when combined with a prism can be used to quantitate the amount of ocular misalignment when strabismus is present, both manifest and latent. Cover testing is also useful to determine if the patient has monocular or binocular diplopia and assess for any dissociative phenomena such as dissociated vertical deviation or latent nystagmus.

The cover test is performed after visual acuity and sensory testing, such as testing for stereopsis, so that those who are prone to break down can be accurately assessed before interrupting fusion. The occluder is placed in front of the fixating eye and removed quickly, this is known as the cover-uncover test. The patient should be tested at distance and near. Observing refixation movement in the fellow eye will show manifest strabismus is present or a tropia. If there is no movement of the fellow eye then observe the covered eye as it is uncovered. If there is refixation movement, then a phoria is present. The cover procedure should be repeated on both eyes. If there is a misalignment present, a prism is placed over the eye that is not being covered the instant the occluder is placed over the fixating eye. This is called the simultaneous prism-cover test and measures the amount of tropia present. A prism can be held over one eye and the occluder can be placed over the fellow eye and moved back and forth between the eyes while the patient is fixating on an object. This is called the prism and alternate cover test. This measures the total amount of strabismus present, both phoria and tropia. This measure is most often used for strabismus surgery calculations. Patients with small angles of esotropia on the simultaneous prism-cover test often have larger angles of esotropia on the prism and alternate cover test. This is seen in patients with monofixation syndrome.

Patients with paretic or restrictive forms of strabismus such as in cranial nerve palsy or Graves orbitopathy may have a larger deviation if the prism is held over the eye without paresis or the eye with less muscle restriction. When performing cover testing on these patients, first place
the prism in front of the affected eye. The measurement obtained is called the primary deviation. If the prism is held over the less affected eye, the deviation will increase; this is called the secondary deviation and should not be used for surgical planning. Herring’s law states that equal innervation is sent to either eye to move the eye in the desired direction. When the eye has a paretic or restricted muscle, the involved muscle will receive greater innervation to create the desired movement. This larger innervation will also be sent to the fellow eye and cause the secondary deviation.

Errors can be introduced in cover testing if the patient is not fixing well because of inattention or poor vision. Testing should always be performed with the patient’s best-corrected vision. It is appropriate to repeat this test at a different time to ensure the alignment is stable. Variable alignment measurements over time may indicate a myopathic condition such as myasthenia gravis.

**Worth 4-dot test.** This is a useful test to quickly determine information about the sensory status of your patient. This test requires spectacles with a red filter on 1 side and a green on the other. In order to remember which eye is seeing which color, I place the red filter over the right eye. A light source with lights color-matched to your spectacle-mounted filters is also required. The light should have 1 red light, 2 green lights, and 1 white light. One should be careful with LCD-screen Worth 4-dot tests, as the LCD screen may bleed through other colors in the spectrum and invalidate the test. This test is performed with the patient’s best correction on first. The light is held at one-third of a meter for peripheral visual field testing and at 6 meters for central visual field testing. After showing the patient the lights, ask him or her how many he or she sees. Four lights is a normal response. A response of 5 lights means diplopia. A response of 2 red lights means suppression (of the left eye if you placed the red filter over the right eye). Three green lights also means suppression (of the right eye with the red filter over the right eye) and more than 5 lights is functional disease. A patient with small-angle strabismus may see 4 lights at near (peripheral fusion), but only 2 or 3 lights at distance (central suppression) are seen in the presence of monofixation syndrome. Additionally, if strabismus is present on cover testing, but the patient only sees 4 lights at near, this is anomalous retinal correspondence.

**Three-step test for cyclovertical muscle palsies.** This is a test to determine the presence of monocular cyclovertical muscle palsy, particularly a superior oblique palsy, and identify which is the affected muscle. There is often confusion about this test and how it should be applied. The 3-step test should be used in cases where there is suspected isolated cyclovertical muscle palsy in the presence of hypertropia. This test is invalidated if there has been previous extraocular muscle surgery especially surgery on the obliques, restrictive strabismus, vestibular disease, supranuclear disease, or more than one cyclovertical muscle involved.

For this test, 3 measurements are required. First, it should be determined which eye is hypertropic in primary gaze. This eliminates the elevators in the palsied eye and the depressors in the fellow eye from being considered. Second, the presence of hypertropia should be determined in side gazes. This eliminates the cyclovertical muscles for which action is opposite the side with the greatest vertical deviation. Last, hypertropia should be evaluated in head tilts, also known as the Bielschowsky head-tilt test. Head tilt to the right stimulates incyclotorsion of the right eye (contraction of the right superior rectus and right superior oblique muscles) and excyclotorsion of the left (contraction of the left inferior rectus and left inferior oblique). The opposite is true with the opposite head tilt. The head tilt with the largest hypertropia will have an underacting intorter or extorter, and the ipsilateral normal muscle will be unopposed and become manifest causing a larger deviation. After eliminating from consideration which muscles are not involved, the one muscle that is left should be the affected muscle. In reality, if this test does not reveal a superior oblique palsy most of the time, it is probably being performed incorrectly.

**Conditions**

**Amblyopia.** Amblyopia is one of the most common causes of vision loss in children. Amblyopia is the result of changes in the occipital lobe neural circuitry from an unequal or bilaterally poor visual experience early in life. It can be caused by constant strabismus (particularly esotropia), anisometropia, or any condition that can occlude the visual axis. Bilateral amblyopia can be caused by high refractive error. This typically occurs in the setting of hyperopia greater than 4.00-5.00 D, astigmatism greater than 2.50-3.00 D, or myopia greater than 7.00-10.00 D. Amblyopia associated with strabismus has some abnormal findings that may be helpful for diagnostic testing. Grating visual acuity, such as in teller card testing, is typically better than Snellen visual acuity. Also, a neutral density filter as in the normally sighted eye does not as readily degrade the vision in the amblyopic eye. This is known as the neutral density filter effect. Lastly, there is often a small amount of eccentric fixation in patients with amblyopia, particularly when the vision is worse than 20/200.

Treatment of amblyopia involves correcting anisometropia with spectacles. Occlusion treatment to the better eye or the use of atropine to cause cycloplegia and blur the image in the better eye can be effective. Once treatment for amblyopia is complete, surgical realignment of the eyes is more effective. Any treatment for amblyopia can cause vision loss in the treated (better) eye. This is known as reverse amblyopia and can be treated reverse patching or stopping the patch altogether. A significant minority of children will have regression of vision in the successfully treated eye; therefore, monitoring of vision after discontinuation of the amblyopia treatment is warranted. If the child is compliant with patching, and the vision has not improved as expected, repeating the dilated eye examination is recommended to reassess the refractive
error or re-examine the optic nerve and macula to ensure there are no other explanations for vision loss. Optic atrophy, mild optic nerve hypoplasia, or macular abnormalities can masquerade as amblyopia and may be missed on a cursory fundus examination.

Bilateral amblyopia is treatable with proper spectacle correction. Vision improvement is typically slow and steady, with most children showing substantial improvement in 12-24 months.

Deprivation amblyopia is caused by something occluding the visual axis. Causes may be cataract, corneal opacity, ptosis, pupillary membrane, or vitreous hemorrhage. In these cases, the visual axis must first be cleared during the critical period of visual development in order to obtain good vision. Infants who have significant visual axis opacity that is not corrected in the first 3 months of life are much less likely to gain good vision after treatment. Once the visual axis is clear, patching treatment can be instituted.

Esotropia. Infantile esotropia occurs in the first 6 months of life and has some unique characteristics. The child often has a family history of strabismus. Infantile esotropia is associated with amblyopia in about 50% of children. They often have angles of deviation larger than 30.00 prism D. Dissociated vertical deviation and inferior oblique overaction is also common (50%) but often occurs later in childhood. Typically, children with this condition have a small amount of hyperopia (<2.50 D), and latent nystagmus is common. When examining the child, cross-fixation may be present. This is the phenomenon where the child uses the adducted eye to view objects in the opposite visual field. It is prudent to ensure the eyes can abduct by noticing the child abduct under monocular occlusion or eliciting abduction by the vestibulo-ocular reflex. The most common treatment for this condition is symmetric recessions of the medial rectus muscles. A resection of one or both lateral rectus muscles can be added for deviations greater than 60.00 prism D.

Esotropia acquired after 6 months of life is most often the result of excess accommodation in the setting of high hyperopia (average + 4.50 D). This condition is best treated with prescribing the child’s hyperopic correction. Accommodative esotropia can present as sudden onset of esotropia in a child 2-3 years old. Acute-onset esotropia without significant hyperopia can rarely be associated with neurologic disease such as hydrocephalus, Arnold-Chiari malformation, and rarely brain tumors. If the suspicion of neurologic disease exists, a brain MRI is the most appropriate test in this age group.

Duane syndrome. Duane syndrome is the result of a congenital defect in the fourth cranial nerve or its nucleus with aberrant innervation from branches of the third cranial nerve. Simultaneous contraction of the medial and lateral rectus on attempted adduction causes globe retraction and palpebral fissure narrowing. This condition is usually sporadic but can be an autosomal dominant condition in 10% or fewer cases. There is a predilection for the left eye and females. Depending upon the degree of aberrant innervation, the alignment may be orthotropic, esotropic, or exotropic in primary gaze position. Typically, there is a smaller angle of misalignment than in sixth cranial nerve palsy (<20.00 prism D). Anisometropic amblyopia is not uncommon in the Duane syndrome eye because of astigmatism. The indications for surgery are misalignment in primary gaze with a head turn, upshoots, or downshoots. Esotropic Duane syndrome can be improved with recession of the ipsilateral medial rectus muscle or transposition of the vertical rectus muscles towards the lateral rectus insertion. Exotropic Duane syndrome can be improved with ipsilateral lateral rectus recession and upshoots and downshoots can be improved with a Y-split of the lateral rectus.

Sixth nerve palsy. The most common cause of acquired incomitant esotropia is sixth nerve palsy. The etiologies often include head trauma or increased intracranial pressure. Other causes include meningitis, intracranial mass, inflammation, or microvascular insult involving the abducens nerve. Workup of sixth nerve palsy should be coordinated with the patient’s primary care provider or neurologist. When considering treatment for the palsy, surgical treatment should be delayed for 6 months or longer if spontaneous improvement is anticipated, such as in traumatic, infectious, inflammatory, or microvascular causes. Prisms can be used to alleviate diplopia, and children should be monitored for amblyopia. Botox injection into the medial rectus can help prevent progressive medial rectus tightening while the lateral rectus recovers. If the esotropia persists beyond 6 months without improvement, horizontal rectus recess and resection can be performed if abduction is present but deficient. Transposition of the vertical rectus muscles in the direction of the lateral rectus is preferred if abduction is very poor.

Superior oblique palsy. The most common cause of incomitant vertical deviation is superior oblique palsy. It can be unilateral or bilateral and congenital or acquired. Acquired cases are most often the result of trauma, but microvascular disease and tumors can lead to superior oblique palsy in rare cases. The 3-step test is helpful in determining the palsied muscle. In unilateral cases, the hypertropia will be worse in contralateral gaze and ipsilateral head tilt. The patient will typically be tilting his or her head away from the side of the superior oblique palsy.
In un repaired congenital cases, a facial asymmetry may be present. Ipsilateral inferior oblique over-action and excyclotorsion are common. Torsion can be evaluated with the double Maddox-rod test, Bagolini lenses, or objectively by observing excyclotorsion on an ocular fundus examination. As in sixth nerve palsy, acquired forth nerve palsies may also improve spontaneously especially in the setting of trauma. Treatments are varied but include: recessing overacting inferior obliques, tucking underacting superior obliques, recessing contralateral inferior rectus muscles for deviations out of the field of action of the obliques, and recessing ipsilateral superior rectus muscles if they are contracted and tight. The Harada-Ito procedure is helpful to eliminate excyclotorsion if there is no significant vertical deviation in primary gaze.

**Infantile nystagmus syndrome.** Nystagmus in infancy can be caused by a number of factors or may be idiopathic. Nystagmus results from an abnormal development of the calibration of the oculomotor system during the system’s development. Typical idiopathic infantile nystagmus is conjugate and primarily horizontal. It is more apparent with attempted fixation or fixation at distance. It decreases with convergence. Null zones are common with associated abnormal head positions. There is a reversal of the nystagmus with presentation of a spinning optokinetic drum. The nystagmus is absent during sleep. Eye movement recordings show accelerating slow phases. Nystagmus that falls outside this typical description may require additional testing. A complete eye examination will help to elicit vision loss as a cause of nystagmus. Any vision loss present in the prechiasmal visual system can result in nystagmus. Etiologies include bilateral optic nerve hypoplasia, albinism, achromatopsia, aniridia, bilateral cataract, or other retinal diseases if bilateral. If retinal disease is suspected because the child has poor vision and nystagmus but the retinal appearance is normal, an electoretinogram may be particularly helpful.

**Congenital glaucoma.** Glaucoma in infancy is typically the result of an abnormality of the trabecular meshwork or anterior chamber angle. Typically, the first interventions are surgical as opposed to typical treatment in open-angle glaucoma common in adults. Most cases are bilateral and do not have a family history, but if there is a family history, the inheritance is typically autosomal recessive. The clinical triad consists of episphaeria, photophobia, and blepharospasm. Common findings include corneal edema, Haab striae, buphthalmos and corneal enlargement, poor vision, optic nerve cupping, indistinct landmarks on gonioscopy, and elevated intraocular pressure. Often, the compliance and stretching of the eye is such that the pressure is not as elevated as high as one would guess, and one must consider the entire clinical picture to make the correct diagnosis. Congenital corneal opacities in infants are most commonly the result of congenital glaucoma. If the cornea is too cloudy to see instruments in the anterior chamber, trabeculotomy is typically the procedure of choice. Goniotomy can be used if the cornea is clear. Valve implant surgery is typically reserved for cases that fail angle surgery. Corneal scarring and amblyopia are common causes of irreversible vision loss in this condition.

**Congenital cataracts.** Cataracts in infants and children represent a diagnostic and treatment challenge for ophthalmologists. Most cataracts are idiopathic if they are unilateral; however, pediatric cataracts may be caused by a myriad of causes especially if they are bilateral. Hereditary cataracts are typically passed on in an autosomal dominant manner but there are a few families with autosomal recessive or X-linked pedigrees. Trauma and other ocular abnormalities are frequent causes of cataract in children. Aniridia, persistent fetal vasculature, anterior segment dysgenesis, and posterior lenticous can be to blame. Systemic and infectious etiologies must be considered in bilateral cases. Down syndrome, galactosemia, corticosteroid exposure, rubella and less likely toxoplasmosis, cytomegalovirus, herpes, and syphilis must be considered. If a child has dysmorphic facial features or other organ system or limb abnormalities and bilateral cataracts, a consultation is indicated with a pediatrician trained in genetic diseases.

Surgical treatment for cataract in infants includes management of the high rate of posterior capsule opacity and secondary pupillary membrane formation. The iris should be manipulated as little as possible. A primary posterior capsulotomy and anterior vitrectomy can reduce but not eliminate secondary membranes that occlude the visual axis. An intact posterior capsule in an infant or young child is guaranteed to become opaque. Implantation of intraocular lenses causes intense postoperative inflammation and secondary membrane formation more frequently than when the eye is aphakic. This fact combined with the difficulty in predicting the appropriate intraocular lens power with the rapid growth of the eye has caused many surgeons to avoid implanting intraocular lenses in the first year of life.

Visual rehabilitation of these children requires a long-term commitment on the part of the child’s caregivers. Contact lenses, glasses, and patching are a mainstay of treatment. Good outcomes are possible with vigilant care.

**Shaken baby syndrome/nonaccidental trauma.** Recognizing the ocular signs of shaken baby syndrome can
help guide pediatricians who are caring for children who are suspected to be victims of abuse. Shaken baby syndrome is the combination of intracranial and retinal hemorrhaging. It is caused by sudden acceleration and deceleration in a child with poor head and neck strength and leads to injury to bridging intracranial vessels. The adherent vitreous in these young eyes creates traction forces that translate into hemorrhages in the retina. Typically, this injury occurs before the age of 3 years. The child may lack external signs of trauma but may have subdural or subarachnoid hemorrhages with bone fractures of different ages. The ocular findings include hemorrhages in multiple layers of the retina. The hemorrhages can be flame shaped, dot, blot, white centered, preretinal, subretinal, or even intravitreal. The type and extent of the hemorrhages should be documented. The hemorrhages can be unilateral or bilateral and are not always found on the side with greater intracranial hemorrhage. Full thickness perimacular retinal folds are very specific for shaking injury. Traumatic retinoschisis as well as optic nerve edema can also be seen.

In cases of suspected abuse there is often a history that is inconsistent with the clinical picture. However, care should be taken to rule out hematologic disorders that could lead to retinal and intracranial hemorrhages such as coagulopathies and leukemia.

Retinal hemorrhages can occur in cases of accidental trauma, but the location and extent of the hemorrhages is typically much less dramatic. Serious accidents such as high-speed motor vehicle accidents and falls from windows do not cause retinal hemorrhages anywhere near the number and extent found in a shaken baby. Cardiopulmonary resuscitation does not cause retinal hemorrhages.

**Conclusion**

This was a brief review of some of the common topics that may be touched upon in the maintenance of certification examinations in pediatric ophthalmology. You can find a complete list of topics at the American Academy of Ophthalmology website: [http://one.aao.org/CE/MOC/POCTopics.aspx](http://one.aao.org/CE/MOC/POCTopics.aspx).

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