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C19

Pediatric Neuro-ophthalmology: Dilemmas in clinical practice

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14:15 – 15:45

Room 115

HAND-OUTS

Is this strabismus really harmful?

Karl Golnik, MD, MEd
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Childhood strabismus is often harmless requiring no further diagnostic evaluation. Common causes include congenital idiopathic esotropia, decompensated congenital phoria, overacting inferior oblique muscle (OAIO), dissociated vertical deviation, Duane's syndrome and congenital 4th nerve palsy. However, occasionally neurologic or orbital processes result in childhood ocular misalignment. These can include orbital trauma or tumor, neuromuscular junction disorders (myasthenia gravis), ocular motor cranial nerve palsy (3rd, 4th or 6th nerve) due to tumor or demyelination and internuclear ophthalmoplegia (INO) due to demyelination. Thus, one must know when to further investigate ocular misalignment in children.

Harmless childhood strabismus is usually asymptomatic, there is no pain and there may be a family history of the same. The exact onset is often unclear. Whereas harmful ocular misalignment often causes diplopia, there may be pain, and the onset is usually obvious. The examination can provide differentiating clues as well. Most childhood strabismus is comitant meaning the degree of misalignment is about the same in different gazes. Thus, one must be sure to measure alignment in all gaze positions. Duanes syndrome, OAIO and congenital 4th nerve palsies are exceptions to this rule. All of the harmful causes of ocular misalignment listed above are incomitant.

When ocular misalignment is present, one must closely evaluate other "fellow travelers" (pupils, eyelids, 5th & 7th nerve, and orbital signs) on the examination. Since the iris sphincter muscle is innervated by the 3rd nerve, close attention to subtle or obvious anisocoria and/or poor pupillary reactivity must be observed closely. Eyelid findings such as ptosis (3rd nerve, myasthenia), fatigable ptosis and Cogan's lid twitch should be noted. One should check function of both the 5th nerve (facial sensation) and 7th nerve (facial muscle strength) as both nerves arise and travel in close proximity to the 6th nerve. Subtle proptosis, chemosis and conjunctival injection may indicate the presence of an orbital process.

Finally, congenital causes of vertical misalignment such as 4th nerve palsy often result in supra-normal vertical fusional amplitudes (> 3 prism diopters) which do not occur in acquired ocular misalignment. One note of caution is that myasthenia gravis can occasionally be so variable that large vertical fusional amplitudes may be mimicked.

In summary, harmless childhood strabismus is common and usually asymptomatic, comitant and without fellow travelers. If the childhood strabismus is symptomatic, incomitant or associated with fellow travelers then further evaluation is warranted (with occasional exceptions (Duane's syndrome, congenital 4th).

Is this visual acuity really too low?

Hana Leiba MD
The Hebrew University, Jerusalem, Israel

One of the most challenging problems the pediatric-ophthalmologist faces is the child with unexplained visual loss. Many of those children are referred to a neuro-ophthalmologist when structural ocular abnormalities have been ruled out.
The neuro-ophthalmologist's aim is to differentiate between organic and non-organic visual loss.
Many organic ocular diseases may present initially without visible ocular abnormalities, eg. Leber's hereditary optic neuropathy or Stargardt disease. However, functional visual loss is not uncommon.
Unlike adults children are rarely malingerers. Their main need is attention. At the end of this talk, the audience will have a tool –box that will be helpful to differentiate between organic and non-organic visual loss in children.

Do I worry about this child's anisocoria?

Aki Kawasaki, MD
University of Lausanne, Switzerland

Anisocoria, as an isolated finding, is typically not a cause for worry in children. Below are some of the more common causes of anisocoria in children:

Mechanical anisocoria

A mechanical anisocoria is due to ocular pathology, such as trauma, infection, intraocular surgery or pseudoexfoliation, which can damage the iris. In the event of iritis, there is often pain and redness as accompanying features. Iris damage may lead to a small pupil that does not dilate well, a large pupil that does not constrict well, a misshapen pupil or a fixed, immobile pupil.

Neurologic anisocoria

If mechanical anisocoria is ruled out and pharmacologic exposure is not a consideration, then a neurologic basis of anisocoria is suspected.

Unilateral mydriasis

1. **Oculomotor nerve palsy – this diagnosis is a cause for concern**

The preganglionic pupil fibers travel within the oculomotor nerve from midbrain to ciliary ganglion. Thus when a large, poorly reactive pupil is found, it is important to look very closely at eyelid and ocular motility. With rare exception, if the anisocoria is isolated and NOT accompanied by ptosis or ocular motility disturbance, it is NOT an oculomotor nerve palsy.

2. **Tonic pupil- this diagnosis is benign**

A tonic pupil is due to a lesion at the level of the orbit, either at the ciliary ganglion or the short ciliary nerves. Acutely, patients complain of a dilated pupil, photophobia and blurring of near vision. In young children, the acute loss of accommodation should be corrected with glasses. Over time, the accommodation tends to recover so regular examinations are needed. Sectoral (focal) palsy of the sphincter is highly characteristic and detectable under high slit-lamp magnification.

Over time, the near response improves, leading to a strong and tonic pupillary constriction to accommodative effort and light-near dissociation. Most cases of unilateral tonic pupil in children are idiopathic, occasionally follow chicken pox or vaccination and do not require neuroimaging.

Unilateral miosis

1. **Horner syndrome – this diagnosis is a cause for concern**

Horner syndrome is due to interruption of sympathetic innervation to the head and eye and results in miosis, ptosis and facial anhidrosis on the side of the lesion. The ptosis is typically mild and involves both the upper and lower lid. In occasional patients, an anisocoria is the only clinical manifestation of an oculo-sympathetic defect. The sign that differentiates the anisocoria of Horner syndrome from

physiologic anisocoria is a delayed and slow redilation of the smaller pupil in darkness, called dilation lag.

A Horner syndrome in an infant or child should raise concern for neuroblastoma. An acute and painful Horner syndrome may be the first sign of acute carotid dissection. Complete neuroimaging of Horner syndrome in children is recommended.

2. Physiologic anisocoria – this diagnosis is benign

A physiologic anisocoria is a common occurrence. Its magnitude is typically 1 mm or less in room light and may be more notable in darkness. Both pupils constrict normally to light and dilate equally in darkness and to cocaine. Rarely, the anisocoria can change sides.

Bilateral poorly reactive pupils – this diagnosis is a cause for concern

If the pupil light reflex is insufficient on both sides, then check the near reflex. If the pupil contraction to near effort is greater than the pupil contraction to light stimulation , this is called light-near dissociation. The presence of bilateral light-near dissociation in a non-blind patient suggests a lesion at the dorsal midbrain and neuroimaging is mandatory.

Is This Disc Really Swollen?

Klara Landau, MD, FEBO
University of Zurich, Switzerland

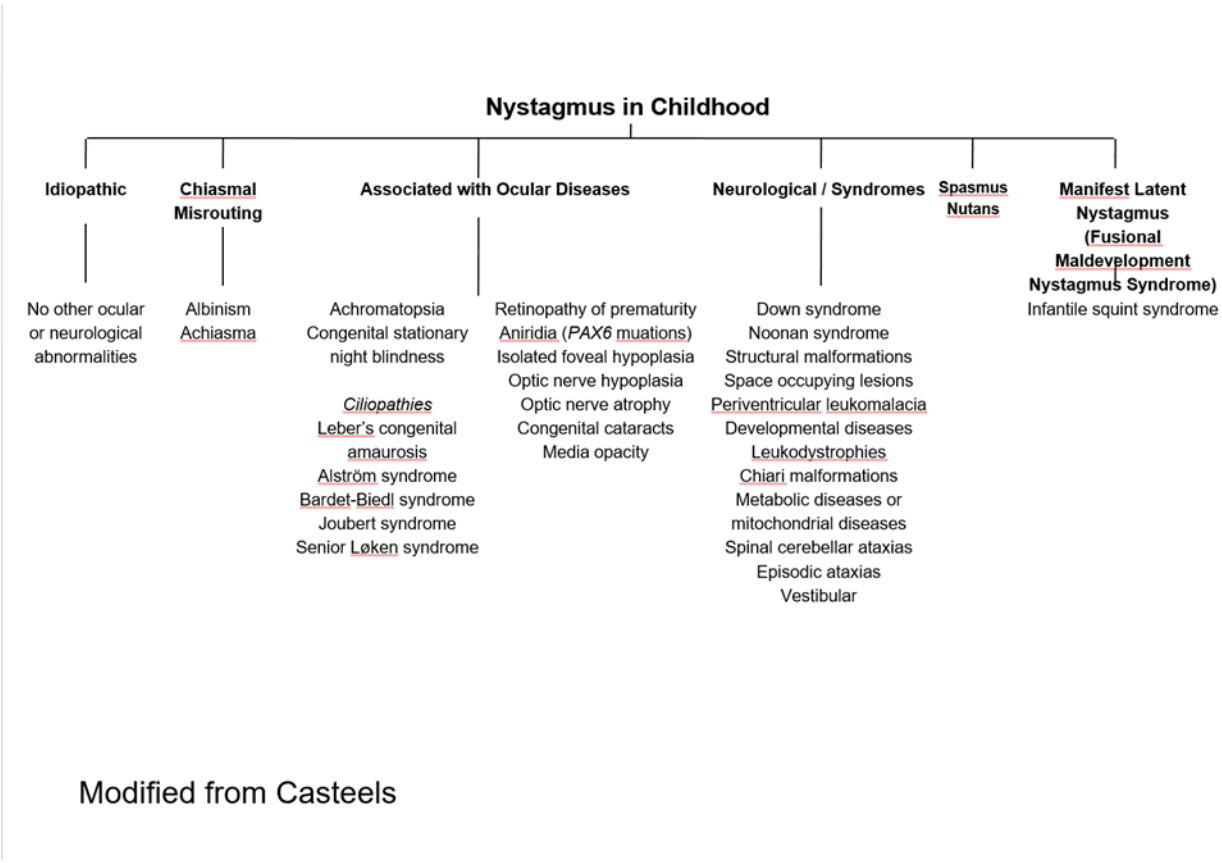
In routine ophthalmological practice, seeing a patient with suspected disc swelling is a challenge that is even more demanding if the patient is a child. Differentiating between true and pseudo-swelling is crucial as management differs greatly: Bilateral disc swelling with good visual function (true papilledema) could be caused by increased intracranial pressure (ICP) due to an intracranial mass lesion, impaired cerebrospinal fluid flow and/or CNS inflammation. Such a suspicion requires urgent referral to a neuro-pediatric center with immediate neuroimaging.
As a rule, a healthy appearing child seen by an ophthalmologist for a routine eye examination without symptoms of increased ICP or some other obvious neurological deficit will have either congenitally crowded discs or buried disc drusen and further management can be performed in a regular way.
The following criteria and signs are helpful in distinguishing between true and pseudo- disc swelling:

Pseudo- disc swelling:	**True disc swelling:**
Commonly an incidental finding	Symptoms of raised ICP are present
Disc does not show definite signs of true swelling, such as obscuration of vessels on the disc margin, cotton-wool spots, hemorrhages	Disc shows signs of nerve fiber layer swelling, such as obscuration of disc vessels, cotton-wool spots, hemorrhages
Typical signs of buried drusen are present such as crowded disc without excavation, multiple branching of disc vessels, prominent disc margins with a few barely visible drusen (use slightly oblique slit lamp beam)	Cave: In long standing chronic papilledema there may be drusen-like bodies present on the disc that should not be confused with disc drusen
Perform B-scan echography, looking for calcification. Perform OCT looking for buried drusen	Look for spontaneous venous pulsation – if present, increased ICP is ruled out
Examine the parents (drusen are inherited as an autosomal dominant trait)	

Is This Nystagmus Really Congenital?

Irene Gottlob, MD
University of Leicester, United Kingdom

Forms of Nystagmus in Childhood



Modified from Casteels

Typical Features	
Infantile Nystagmus	**Acquired Nystagmus**
Visual acuity variable	often good
Nystagmus pendular of jerk mostly horizontal and conjugate can be vertical and/or disconjugate in retinal disorders latent nystagmus with increasing amplitude upon covering one eye	pendular or jerk Conjugate or disconjugate Horizontal or vertical Often gaze evoked
Eye movement recordings idiopathic nystagmus and nystagmus associated with retinal diseases: increasing slow phase velocities Latent nystagmus: decreasing slow phase velocities	constant slow phase velocities
Anterior segment normal or iris transillumination, aniridia, dygenesis	no specific changes
Fundus normal or foveal hypoplasia, retinal dystrophy	no specific changes
OCT Normal or foveal hypoplasia with reduced foveal pit and persistent retinal layers in fovea and reduced size of outer retinal layers, retinal dystrophy with changes of lamination of outer retinal layers (retinal dystrophy) or hyporeflective zone (achromatopsia)	no specific changes
ERG Normal or reduced (extinguished photopic in achromatopsia, negative in congenital stationary night blindness and reduced in other retinal dystrophies)	normal
VEP (lateralisation) Normal or abnormal lateralisation in albinism and FHONDA (foveal hypoplasia, optic-nerve-decussation defects, and anterior segment dysgenesis)	normal
MRI No specific changes	Often specific changes such as demyelination in MS