Neuro-Ophthalmic Disorders SIG
The Lawrence Gray Symposium: “Neuro-Ophthalmic Top 10 Lists”

Program Details/Speakers:

Moderators: Joseph Sowka, OD (jsowka@nova.edu), Patricia Modica, OD (pmodica@sunyopt.edu), Kelly Malloy, OD (kmalloy@salus.edu)

Speaker 1: Jonathan Trobe, MD (jdtrobe@med.umich.edu): “The Top 10 Mistakes in Evaluating Diplopia”

Speaker 2: Cherie Farkash, OD (farkashc@med.umich.edu): “The Top 10 Mistakes in Evaluating Visual Fields”

Speaker 3: Valerie Purvin, MD (vpurvin@iupui.edu): “The Top 10 Mistakes in Evaluating Unexplained Visual Loss”

Overview/Summary:
Evaluating neuro-ophthalmic disorders can be perceived as being difficult and intimidating. This symposium helps you learn how to avoid the most common mistakes when evaluating diplopia, visual fields, and unexplained visual loss. In this fast-paced program focusing on Neuro-Ophthalmic Top-10 lists, world-renowned neuro-ophthalmologists provide clinically applicable tips to increase your comfort level in dealing with these concerning presentations.

Neuro-ophthalmic Disorders in Optometry SIG: The Lawrence Gray Symposium

Top 10 Mistakes in Evaluating Diplopia
Jonathan Trobe, MD
Thursday, October 12, 2017

Optometrists are often the first caregivers to encounter patients complaining of diplopia. In some cases, diplopia is caused by potentially life-threatening conditions that require urgent disposition. This lecture will present the 10 most common—and critical—mistakes that are made in the evaluation of diplopia with the hope that having them highlighted and explained will lead to improved diagnosis and management.

I will divide the errors into those that involve problems with technique and those that involve problem of interpretation of findings.

10. Failure to measure, when appropriate, alignment in horizontal, vertical, and torsional planes in primary gaze position and pertinent eccentric positions of gaze.
Too often, eye care professionals only do cover testing in primary gaze. It is critical to do cover testing in multiple positions of gaze in order to determine whether or not the deviation is comitant and if there is any recognizable pattern of the deviation to suggest a cranial nerve palsy.

9. Failure to recognize when the Maddox rod is a critical adjunct to the cover test in assessing alignment.

We have available to us both objective and subjective means of assessing ocular motility alignment. We must realize the value of both cover testing and Maddox rod testing, both in isolation and in combination with each other.

8. Failure to recognize the significance of a hyperdeviation that reverses direction between upgaze and downgaze as a means of distinguishing third nerve palsy from skew deviation.

It is important to know the critical ocular motility patterns that suggest cranial nerve palsies. Due to the possible emergent association with aneurysm, it is especially important to be able to recognize the pattern of a cranial nerve III palsy, even if it is subtle. This requires having a good differential diagnosis of vertical ocular misalignment, and being able to distinguish a cranial nerve III palsy from other causes of vertical misalignment, including skew deviation.

7. Failure to acknowledge that preservation of iris sphincter function in third nerve palsy does not provide much diagnostic value.

We must remember that not all aneurysmal cranial nerve III palsies have pupillary involvement. Therefore, we still need to rule out an aneurysm in all partial or painful cranial nerve III palsies, regardless of the pupil findings.

6. Failure to detect aberrant regeneration in suspected third nerve palsy.

While #7 above cautions us not to rely too heavily on the pupil findings in a cranial nerve III palsy, we must realize that there is one pupillary finding that does provide significant diagnostic value. The finding of aberrant regeneration in a cranial nerve III palsy indicates that the palsy is NOT from a vasculopathic process. Therefore, the concern for aneurysm or tumor is heightened.
5. Failure to realize that patients with small misalignments, especially vertical misalignments, may report blurred vision rather than double vision.

It is important to measure the ocular alignment in multiple positions of gaze even in patients who do not specifically report diplopia. Some patients may not realize that they are seeing double, and may instead complain of blur rather than diplopia.

4. Failure to realize that diplopia may be secondary to impaired sensory fusion.

We must realize that every complaint of diplopia is not related to an ocular misalignment. There may be other causes of reduced sensory fusion, which must be assessed in order to determine the reason for the complaint of diplopia.

3. Failure to hunt for vertical gaze abnormalities or nystagmus in what looks like a unilateral third nerve palsy.

Don’t look at the ocular motility findings or cover test results in isolation. We need to factor in all of our observations and findings when localizing a problem. Nystagmus is one such important finding that can help in localization. A unilateral abducting nystagmus can suggest an internuclear ophthalmoplegia, whereas convergence retraction nystagmus can suggest dorsal midbrain syndrome.

2. Failure to use a suction duction device to help diagnose restrictive extraocular muscle conditions.

We must keep in mind that we have the ability in office to help distinguish between restrictive and neurogenic ocular motility limitations. When there is a ductional limitation, we must remember that the forced duction test is critical to make this distinction. A positive forced duction, indicating a restrictive process, helps us to realize that we are dealing with an orbital process, and we can then streamline our work-up to determine the exact nature of the problem.

1. Failure to realize that myasthenia gravis can mimic cranial nerve palsies and brainstem disorders.

Myasthenia gravis can cause ANY pattern of ocular misalignment. Therefore, it can have a pattern that looks exactly like many other things, including cranial nerve III, IV, and VI palsies, as well as internuclear ophthalmoplegias and skew deviations. Therefore, even when the patient presents with what appears to be a cranial nerve palsy of brainstem
motility finding, we must still consider the possibility of myasthenia gravis. In office testing on initial evaluation, assessing for interval change on follow-up appointments, and laboratory testing are all very helpful in determining whether there is a suspicion for myasthenia gravis, despite what appears to be a recognizable motility pattern of another disease process.

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Top 10 Mistakes in Evaluating Neurologic Visual Fields
Cherie Farkash, OD, FAAO
Thursday, October 12, 2017

10) Failure to do a visual field
When a patient has subnormal visual acuity or progressive vision loss, having information about the visual field can be key in making the diagnosis. Visual fields can also be important for patients with persistent photopsia in an otherwise normal eye exam. Cases will be presented in which vision loss was mistakenly attributed to cataract, maculopathy, or retinal disease causing a delay in diagnosis.

9) Failure to do a visual field correctly
Technical errors are common, especially in teaching settings where new students are rotating into the practice. Mistakes such as performing a visual field with the room lights on, selecting the wrong trial lens, not positioning the trial lens correctly, testing the wrong eye, forgetting to patch an eye, not making the patient comfortable, and not explaining the test can all give misleading results. Other aspects will be discussed including why it is recommended to "turn on" the foveal threshold part of the test.

8) Failure to recognize when patients cannot do a visual field
Some patients are not good candidates for visual fields due to a variety reasons including dementia and inattention. In these cases, it is important to
make treatment and management decisions based on other criteria. It becomes important to consider confrontation fields and/or patient visual behavior. Cases of “blackout” or “tunnel vision” fields will prompt a discussion about the differential diagnosis and when to suspect that the visual fields are misleading.

7) Assuming there has been progression based on just one visual field
In patients with idiopathic intracranial hypertension or sellar/suprasellar masses, treatment and management decisions are sometimes based on visual function. Cases will demonstrate the importance of repeating the visual field before assuming progression.

6) Failure to look for tilted optic discs in what appears to be bitemporal hemianopia
A patient with congenitally dysplastic or “tilted” optic discs can have temporal wedge field defects in each eye, mimicking a bitemporal hemianopia. A case will demonstrate this potential pitfall, and clinical pearls will be presented in analyzing the disc and the field.

5) Failure to look for an afferent pupillary defect in homonymous hemianopia
The presence of an afferent pupillary defect in a patient with homonymous hemianopia (assuming no other ocular pathology) is highly localizing to the optic tract. The anatomical basis will be reviewed. A case will demonstrate that being able to direct a radiologist’s attention to the optic tract region can be helpful in this tricky area.

4) Failure to localize a monocular temporal hemianopic defect
Most of us would have little trouble localizing a bitemporal hemianopic defect and arranging for the appropriate referral and/or neuroimaging study. However, less well appreciated is the fact that a monocular temporal
hemianopic defect also localizes to the same area. Case(s) will be presented to demonstrate this.

3) Mistaking hemifield neglect for homonymous hemianopia

A case will be presented about a patient with an old pre-existing right homonymous hemianopia who suddenly developed apparent “blindness” and was found to have a lesion in the right parietal region. She had no light perception, a right gaze deviation, and gestured only into right hemispace. The case will demonstrate the challenges in distinguishing left hemifield neglect and left homonymous hemianopia.

2) Assuming that all field loss in a glaucoma patient is due to glaucoma, especially when it is mainly temporal.

A case will be presented to demonstrate clinical pearls in identifying sellar/suprasellar visual field defects that can mimic glaucoma.

1) Failure to recognize that respect for the HORIZONTAL and VERTICAL is OCCIPITAL

Mild lesions to the optic radiations (temporal, parietal lobes) do not cause quadrantic defects. When a homonymous hemianopia respects both the horizontal and the vertical, the localization is to the occipital lobe. A case will be presented about a monocular patient whereby this clinical pearl was critical in the determining the appropriate treatment and management.

Valerie Purvin, MD
Indianapolis, 2017

The Top Ten Mistakes in Evaluating Neurologic Visual Loss

HISTORY

1. Mistaking sudden onset for sudden discovery
The 3 most common causes of optic neuropathy are optic neuritis, anterior ischemic optic neuropathy (AION) and compressive lesions. The physical findings in these cases are generally non-specific. We distinguish them by their time course. In optic neuritis the vision declines over several days, remains stable for a couple of weeks, and then slowly improves over several months. In AION the onset is more abrupt, often found upon awakening, and recovery is modest if at all. Compressive lesions, in contrast, are marked by relentlessly progressive visual loss.

Thus, diagnosis is based largely on these distinctive temporal features. Occasionally a patient reports sudden onset of loss but in fact the visual loss was gradual but discovered abruptly, typically on the occasion of covering the good eye. One tip-off that this is the case is the discovery of optic disc pallor (or thinning of the retinal nerve fiber layer) in an eye with reportedly recent acute loss. It is sometimes helpful to ask the patient what he/she was doing when the visual loss occurred.

2. Mistaking ocular for neurologic disease

The distinction between ocular and neurologic causes of visual disturbance is important, especially early in the course of evaluation. In many cases we can make this distinction based on the clinical features, and even more specifically on the history, rather than after obtaining extensive (and negative) testing for neurologic disease.

The hallmark of visual loss due to optic neuropathy is loss of brightness. Patients report that things look “dimmer” or “darker” in that eye. Desaturation of color perception is often present as well. In contrast, aberrations of the ocular media (refractive, corneal, lenticular) produce “blurring” but not dimming, often accompanied by seeing haloes around lights. Distortion of shape or size in one eye is always retinal, never due to optic nerve disease. Degradation of vision in bright or dim illumination is also characteristic of retinal disease.

3. Believing the patient that visual loss is monocular rather than hemi-field.

Patients with homonymous hemianopic visual loss frequently experience and therefore describe their defect as affecting one eye. This is even true in conditions that are persistent, such as the loss following an occipital stroke. This distinction is even more challenging when the visual loss is episodic. The key to making this distinction is to ask the patient to describe what they could see during the episode. For example, seeing only one half of the person one is conversing with, or seeing only one half of the dinner plate, is not compatible with monocular loss. Only hemi-field loss produces this appearance.

4. Failing to ask about non-visual symptoms

In the discipline of neuro-ophtalmology the details of the visual symptoms are often the key to the correct diagnosis. We thus tend to spend more time on the patient’s description of these symptoms than for typical ocular disorders. However,
in this focused attention, other non-visual symptoms are sometimes neglected. For example, the average patient with giant cell arteritis has a two-month history of systemic symptoms prior to the onset of visual loss. Most patients won’t think to tell their eye doctor about their jaw pain or their decreased appetite. They’re not supposed to know that – we are. Another example is the characteristic pulsatile tinnitus in patients with increased intracranial pressure (ICP). They are not likely to volunteer that they just saw an ear doctor for this pulsing sound, only to be told that their ears are fine.

EXAM
5. Over-reliance on visual acuity

For many practical purposes, such as for obtaining a drivers license or passing a school vision screening exam, visual acuity is taken to mean vision. When we say ‘the vision was normal’ we often mean that the acuity was 20/20. The afferent visual system has a number of skills, of which the ability to read small high contrast letters is a relatively low level one. It is estimated that the system can lose 44% of its retinal ganglion cells and still read the 20/20 line on the Snellen chart. Part of this is because the optimally refracted eye of a healthy young individual should be able to do better than that – more like 20/15 or even 20/10. But it is also because the system is over-determined; reading small black letters on a white background does not engage the entire system. Moreover, it is possible to lose half of the vision in an eye, whether from a hemianopic defect or a bi-temporal one, and still read normally in the remaining hemi-field.

For these reasons, a patient with an afferent “complaint” may be dismissed as overly sensitive or even neurotic, in the face of normal acuity. Any patient with an unexplained afferent symptom should have other tests of optic nerve function, such as color vision and contrast sensitivity and most importantly visual field testing.

6. Failing to obtain visual field testing

Tests of visual acuity, color vision and contrast sensitivity only tell us about the central field. Lots of mischief can happen in surrounding areas of vision without showing up in the center. For this reason, in some conditions serial assessment of optic nerve function must include measurement of visual fields.

One common example of this is idiopathic intracranial hypertension (IIH). In patients with IIH who lose vision, early defects usually involve the inferior nasal field and generalized constriction. Loss of acuity is typically a late finding, similar to what we see in glaucoma.

In most cases the serial examination of a patient with IIH includes observing optic disc appearance – is the papilledema improving or not? However, when an optic nerve dies from persistent edema, the resulting loss of the nerve fiber layer results in a decrease in the papilledema. The only way to know that decreasing papilledema is a sign of improvement is to couple tests of optic nerve function with measurements of optic nerve structure. The visual field is the most important of these tests of function.
7. Failing to recognize a bi-temporal visual field pattern

Loss of temporal field in each eye is a highly localizing neuro-ophthalmic finding, indicating a lesion involving the optic chiasm. Because most such lesions are tumors, failure to appreciate the significance of this finding is an especially grievous miss.

Schematic representations of these defects suggest that the entire temporal half of vision is lost in each eye but in fact this degree of severity is rare. More often what we see is lesser variations on this theme. In general, central field is affected before and more prominently than peripheral. The use of screening perimetry that employs only a bright test object may therefore miss the defect at this stage. Because most compressive lesions approach the chiasm from below, defects usually affect the upper field first. Thus, the most common form of early chiasmal loss is a bilateral superior temporal quadrant defect. Note that this is reminiscent but the opposite of what we see in glaucoma, which also tends to affect superior field early but on the nasal side of fixation. In some cases of chiasmal and pre-chiasmal compression, the visual loss is very asymmetric or even unilateral but with that same distinctive temporal hemianopic pattern.

8. Missing a relative afferent pupillary defect

A relative afferent pupillary defect (RAPD) is both highly sensitive and highly specific for neurologic visual loss. (The caveat here is that the RAPD represents a comparison between the two eyes: disease that is bilateral and symmetric will not result in an RAPD.) For example, after recovery from an attack of optic neuritis, visual acuity typically recovers to 20/20 and the visual field may return to normal but in most cases there is still an RAPD. Similarly, at the leading edge of disease, in compressive optic neuropathies, an RAPD is the earliest finding, along with subjective loss of color saturation. A decrease in visual acuity only occurs later in the course.

Detection of a small RAPD (also termed a Marcus Gunn pupil) demands meticulous technique. Low ambient lighting and a bright test light are important, as is distance fixation (to prevent an accommodative pupil response). Step to one side so that the patient can view a distance target. Observe the pupil response to light in each eye and then alternate the light from one eye to the other. Move as quickly as possible across the bridge of the nose, to avoid allowing the system a chance to escape, but slowly enough to maintain precision. While older textbooks sometimes state that an initial constriction upon illumination constitutes a negative test, this is not correct. In a small or subtle RAPD, there is usually an initial constriction followed by more expansion when the system is illuminated via that optic nerve than via the other. It is this resting state of both pupils when illuminated from one side vs the other that is important. After several passes, bleaching of the retinal makes subsequent observations less reliable. If you’re not sure what you saw after a few tries, give the system a rest for a minute or two and then try again.
9. Diagnosing migraine based on a first episode

By definition, the diagnosis of migraine consists of a tendency to have recurrent attacks of characteristic neurologic and frequently visual symptoms. We can guess that some day we will look back at this attack and identify it as the first of the disorder but the diagnosis cannot actually be made during the first event. The reason this caveat is important is that the characteristic features of migraine, namely headache with light sensitivity, nausea and vomiting are also the cardinal features of subarachnoid hemorrhage, meningitis and increased ICP. How much testing is needed to rule out (or in) these disorders in a given case is a judgment call, but they must be considered. Beware of the first migraine event. Prompt referral to a neurologist in such cases is appropriate.

10. Moving too slowly

There are not many genuine neuro-ophthalmic emergencies but there are a few in which the outcome is in part dependent upon prompt diagnosis and treatment. Giant cell arteritis (GCA) is an important one. When one eye is affected by GCA, usually in the form of AION, the chances of the fellow eye similarly losing vision is high, and when this occurs it is usually within one to two weeks; hence the emergent nature of appropriate testing and treatment. Corticosteroids usually prevent second eye involvement but blood tests should always be obtained prior initiating treatment.

Internal carotid artery dissection, manifest in about one-third of patients as an acute Horner’s syndrome, is another emergency. The risk of stroke after such an event is high, usually within two weeks of the initial event. Such patients should be sent to a local emergency department for anti-coagulation. If more than 3 weeks has elapsed without a stroke, the subsequent risk is very low; these patients can be managed with anti-platelet therapy on an outpatient basis.

Pituitary apoplexy is another important neuro-ophthalmic emergency. This condition occurs due to hemorrhage of a pituitary tumor. Patients present with acute headache and light sensitivity and variable neuro-ophthalmic findings, including unilateral or bilateral visual loss and double vision often due to 3rd nerve palsy which may be bilateral. In most cases the pituitary tumor was not known prior to hemorrhage. These patients should be sent directly to a local ED.

Papilledema, meaning disc edema due to increased ICP, is variably urgent. Occasional cases are due to a brain tumor or other expanding mass or to hydrocephalus, and in these patients prompt diagnosis is crucial. In cases due to IIH, the degree of urgency depends on the severity of the papilledema and the status of optic nerve function. The best course is prompt referral to a neurologist or neurosurgeon for further management.