Competencies in Neuro-ophthalmology: Diagnostic Tests, Headaches, and Optic Neuropathies

By Marc J. Dinkin, MD; and Cristiano Oliveira, MD

As part of the Road to Recertification article series in New Retina MD, Marc J. Dinkin, MD, and Cristiano Oliveira, MD, provide an overview of some of the neuro-ophthalmology topics that retina specialists may want to review in more detail for maintenance of certification. As with every article in this series, their overview is not meant to take the place of a comprehensive review course; rather, its purpose is to highlight some key areas within the neuro-ophthalmology subspecialty and to encourage a more thorough review to prepare for the Demonstration of Ophthalmic Cognitive Knowledge examination.

—Diana V. Do, MD

The maintenance of certification exam for neuro-ophthalmology/orbit covers a large amount of material. Here we focus on 3 specific areas: diagnostic methods, headaches, and optic neuropathies.

Diagnostic Methods

There are a few routine diagnostic methods in neuro-ophthalmology. One common starting point with our patients is visual-field testing. It can be conducted at the patient’s bedside, does not require any special tools, and is a good first assessment to identify certain defects. By conducting a visual-field test in all new patients, you can clarify an afferent vs efferent diagnosis, and the test will guide you toward any other screening that patients may require.

Automated static perimetry is a helpful diagnostic method to use with all patients in our specialty, as it can give you a larger sense of the problem and help distinguish an optic neuropathy from another diagnosis such as glaucoma or a retinal defect.

Computed tomography (CT) is a useful scanning device in neuro-ophthalmology. It is used in patients who are contraindicated for magnetic resonance imaging (MRI) because of metal in their body, and it is superior to other imaging technologies for viewing hemorrhages. CT can also assist with viewing tumors and signs of a stroke. However, if there are no contraindications for MRI, we recommend performing MRI in most situations because of its superior imaging capabilities. Imaging after gadolinium administration increases the sensitivity for orbital and intracranial tumors.

Diagnosing and Treating Headaches

Headaches are a common complaint treated in neuro-ophthalmology. It is important to characterize headaches with as much detail as possible so you have a better idea of their cause.

A common headache type that we see is intracranial hypertension. The patient may wake up with a headache, and the headache may be worse when he or she is lying down. The patient may also describe a “whooshing” sound in his or her ears, and there can be transient visual obscurations. Patients with suspected intracranial hypertension headaches need a full workup, including MRI, MR venogram to rule out a venous sinus thrombosis, and lumbar puncture to search for signs of infection or a tumor.

—Diana V. Do, MD

Patients with suspected intracranial hypertension headaches need a full workup, including MRI, MR venogram, and lumbar puncture to search for signs of infection or a tumor.

—Diana V. Do, MD
Obese women of childbearing age may suffer from idiopathic intracranial hypertension, in which there is no visible cause for the headaches. These patients are treated with acetazolamide or with surgery. Although the cause is not always evident, treatment is important to avoid permanent vision loss.

Migraines are another type of common headache. They are usually moderate to severe, unilateral, and throbbing. Some patients also experience visual auras or nausea. Patients feel better when they lie down in the dark, and they can usually identify triggers that set off migraines. Although we can treat migraines with triptans and nonsteroidal anti-inflammatory drugs, it is helpful to also involve neurologists in patient care.

Optic Neuropathies

Neuro-ophthalmologists manage optic neuropathies on a daily basis. One of the most common neuropathies is optic neuritis, which is most common in younger women. Optic neuritis is an inflammatory demyelinating condition of the optic nerve in which patients present with fairly acute vision loss in 1 eye, usually with pain on eye movement. As with most optic neuropathies, there is a loss of visual acuity, color vision and central visual loss, but in the majority of cases, the inflammation is retro-orbital, and there is no visible swelling of the optic nerve head.

Generally, vision loss continues for several days, stabilizes after a week or 2, and begins to improve over 3 to 6 months. By 1 year, it is thought that most patients return to 20/40 or better.

In a number of cases, the cause of optic neuritis is multiple sclerosis (MS)—or the patient may already have MS when he or she presents with optic neuritis. The chance of developing MS after optic neuritis is approximately 33%; for this reason, it is critical to perform an MRI of the brain and orbits to look for lesions that could indicate a greater risk for MS. If a patient is considered high risk for MS, we’ll recommend that he or she uses an MS disease-modifying agent, which is thought to lower the chance of developing MS.

Lack of blood flow to the optic nerve head, known as anterior ischemic optic neuropathy (AION), causes disc edema, accompanied by loss of visual acuity and color and visual-field defects, which are frequently altitudinal. In patients over 55 years of age, the cause may be giant cell arteritis, which is a vasculitis affecting the branches of the external and internal carotid arteries. Headache and jaw claudication frequently accompany this disorder. It is critical to treat with steroids immediately, to avoid AION to the fellow eye. It is one of the true emergencies in our field, and we treat immediately even if the diagnosis is not yet confirmed. Steroids are then tapered over 1 year. Of note to retinal specialists is that giant cell arteritis can also cause retinal occlusion.

Neuro-ophthalmologists also treat nonarteritic AION, which appears to be the result of poor autoregulation of the arterioles that feed the optic nerve head. The use of steroids in this condition remains controversial, and there remains no proven treatment to reverse or prevent further vision loss.

Two other optic neuropathies with which to be familiar are toxic and nutritional optic neuropathies. The flagship for the latter is vitamin B₁₂ deficiency. Toxic optic neuropathy is caused by exposure to a toxic substance, such as certain systemic medications.

Eye Movements

On a final note, 1 additional emergency in neuro-ophthalmology is a pupil-involving third nerve palsy. The mydriatic pupil may be accompanied by ptosis and an eye which is laterally and inferiorly deviated (down and out). Although there are many possible causes, any pupil-involving third nerve palsy is aneurysmal until proven otherwise, and an MR or CT angiogram should be performed urgently to ensure prompt diagnosis in order to prevent a subarachnoid hemorrhage.

Conclusion

These articles provide a brief review of some of the common topics that may be touched upon in the maintenance of certification examination in neuro-ophthalmology. You can find a complete list of topics and more study materials and resources at the American Academy of Ophthalmology website: http://one.aao.org/POCTopics.

Marc J. Dinkin, MD, and Cristiano Oliveira, MD, are both Assistant Professors of Ophthalmology at Weill Cornell Medical College in New York. They both specialize in neuro-ophthalmology. Dr. Dinkin may be reached at (646) 962-2020 or mjd2004@med.cornell.edu. Dr. Oliveira may be reached at (646) 962-2020 or cro9004@med.cornell.edu.

Diana V. Do, MD, is an Associate Professor of Ophthalmology and Vice Chair for Education in the Department of Ophthalmology and Visual Sciences at the University of Nebraska Medical Center in Omaha. Dr. Do is also the Director of the Carl Camras Center for Innovative Clinical Trials in Ophthalmology and the Director of the Ophthalmology Residency Training Program. Dr. Do is a member of the New Retina MD Editorial Board. She may be reached at diana.do@unmc.edu.