



# Ophthalmology Update

Richmond Eye Associates, P.C.

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## Ophthalmic Topics of Interest to the Medical Physician

### Neuro – ophthalmology Update

This issue discusses recent developments in neuro-ophthalmology as well as important clinical conditions and findings for the medical physician.

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### CHAMPS study Indicates Early Treatment is Protective in High Risk Multiple Sclerosis Patients

The recently published CHAMPS study<sup>1</sup> (Controlled High-Risk Subjects Avonex Multiple Sclerosis Prevention Study – Sept. 28, 2000) indicates that early treatment with interferon beta-1a can be protective in the progression or development of multiple sclerosis after an initial demyelinating event. In this study, about half of the patients had as the qualifying event optic neuritis, with the other patients experiencing either incomplete transverse myelitis or a brain-stem / cerebellar syndrome.

CHAMPS was a randomized, prospective, double-blind study where 383 patients were either given placebo or weekly injections of 30 micrograms of Avonex after and initial treatment with corticosteroids. Study end points included the development of clinically definite MS (CDMS) and changes in the MRI findings.

Results of CHAMPS showed that during 3 years of follow-up, the probability of developing CDMS (with the development of a second demyelinating event) was 44% lower in the treated group vs. the placebo group.

Significantly superior MRI findings were also found, as shown in the box.

#### Results of CHAMPS

- 44% reduction in the risk of developing a second demyelinating event in the treated group
- 57% reduction in the development of new T2 lesions on MRI in the treated group
- 91% reduction in T2 lesion volume in the treated group
- 82% of those in the placebo group developed ongoing, new lesions on MRI

The importance in these findings is that they change the way that initial demyelinating events such as optic neuritis are handled. The Optic Neuritis Treatment Trial (ONTT)<sup>2</sup> showed that patients with optic neuritis and evidence of demyelination on MRI had only a transient delayed in the development of MS after receiving IV steroids. More importantly, this study showed that oral steroids

were detrimental in the development of MS, and that they should be avoided.

With the CHAMPS study results, the need for MRI at the time of an initial demyelinat-

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#### **In the Next**

#### **Issue of**

#### **Ophthalmology Update:**

#### **Controversies in Ophthalmology**

- Does Night Light Use Cause Myopia?
- Does Plaquenil Cause Retinopathy?
- Roth Spots and Endocarditis

### Causes of Simultaneous Bilateral Visual Loss

There are numerous causes of acute unilateral loss of vision, including retinal detachment, retinal artery occlusion, ischemic optic neuropathy, wet macular degeneration, acute angle closure glaucoma, and many others. Similarly, there are many causes of chronic

bilateral visual loss, such as chronic glaucoma, dry macular degeneration, diabetic retinopathy, and cataract. However, there are relatively few causes of sudden, bilateral, and simultaneous loss of vision. Due to inde-

*Continued on page 3 . . .*

## Visual Loss During Cardiac Surgery

Cardiac surgery is commonly and successfully performed for many reasons, including ischemic heart disease, cardiac valve abnormalities, cardiomyopathies and arrhythmias. However, there are several mechanisms for visual loss as a complication of cardiac surgery.

### Ischemic Optic Neuropathy:

Through several mechanisms, unilateral or bilateral ischemic optic neuropathy can occur during cardiac surgery. An individual with this complication would notice an immediate reduction in visual acuity after surgery, often with a horizontally oriented loss of peripheral visual field in one or both eyes. If unilateral, a Relative Afferent Pupillary Defect would be present, and the optic nerve on the affected side would appear pale and swollen (unless the vascular insult was retrobulbar). Risk factors for ischemic optic neuropathy after cardiac surgery include:

- History of pre-existing optic nerve damage or significant glaucoma which may make the optic nerve more susceptible to damage during surgery.
- Systemic hypotension, hypothermia, or significant systemic hypertension.
- Prolonged cardiopulmonary bypass time with myocardial ischemia
- Generalized edema during the bypass.

### Cerebral Ischemia or Infarction:

Hemodynamic factors during cardiopulmonary bypass can result in cerebral infarct causing visual changes as well as other neurological deficits.

- Loss of peripheral vision, manifested as a bilateral visual field cut oriented vertically (homonymous hemianopia).
- Severe ischemia to the occipital cortex could lead to cortical blindness, with bilateral loss of central and peripheral vision.
- Other more unusual visual symptoms related to cerebral ischemia during surgery include impaired ocular movements in maintaining fixation on an object, slowness of visual fixation, inability to read, visual hallucinations, and persistence or repetition of a visual image (palinopsia).

### Embollic Disease:

Manifestations of embolic retinal disease have been reported after cardiac surgery. These can lead to branch retinal artery occlusions or central retinal artery occlusion, with retinal cotton wool spots, attenuated retinal arterioles, and ischemic retinopathy being present. Typically a complete loss of vision in the area served by the ischemic retina would be present.

## Clinical Pearl: Diagnosis of Ocular Myasthenia Causing Ptosis

Myasthenia Gravis is an autoimmune disorder which often has ophthalmic manifestations, such as intermittent ptosis and double vision. The ptosis often affects one eyelid more than another, and the eyelids are often "fatigable", where prolonged upgaze will lead to the development of ptosis. Myasthenia is also known for causing variable fatigability of the extraocular muscles, leading to a variable and intermittent diplopia.

Testing for myasthenia has included serum acetylcholine receptor antibody levels, electromyography, the Tensilon test, and more clinically oriented tests such as the sleep test, rest test, and ice test. Generally it has been noted that the ptosis of myasthenia improves following sleep, rest, or the application of ice to the eyelids.

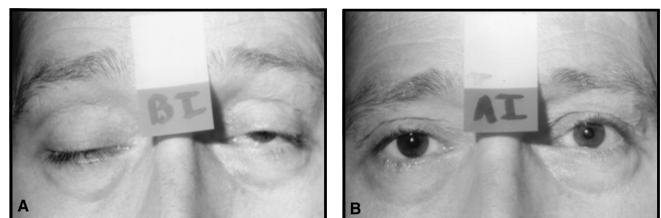
A recently study published in the November issue of Ophthalmology<sup>1</sup> sought to determine the effectiveness of the ice test and the rest test on the ptosis of newly diagnosed myasthenia patients, and on patients with ptosis from other causes (levator dehiscence, oculomotor palsy, Horner's syndrome, and congenital ptosis).

The ice test was performed by placing a surgical glove filled with crushed ice on the more ptotic eyelid for 2 minutes. The rest test was performed by placing a surgical glove filled with cotton on the more ptotic eyelid for 2 minutes. Each test was performed on each

patient on the same day, with a period of eyelid fatigue in between.

It was found that eyelid elevation improved with the rest test (median elevation of eyelid of 2 mm), however the ice test cause a more dramatic elevation (median elevation of 4.5 mm). None of the non-myasthenic cases of ptosis were improved by rest or ice. Overall, 50% of myasthenic eyelids improved at least 2 mm after the rest test, and 90% improved at least 2 mm after the ice test.

The rest and ice tests appear to be simple, non-invasive, and reliable tests to establish myasthenia as a cause of ptosis.



**Before Ice**

**After Ice**

As shown above, rest reduces ptosis for both eyes, but ice over the right eye gives a more impressive improvement.

<sup>1</sup>Kubis KC, et al. The ice test versus the rest test in myasthenia gravis. Ophthalmology 2000;107: 1995-1998.

## Update on Pseudotumor Cerebri and Optic Nerve Decompression

Pseudotumor cerebri, or idiopathic intracranial hypertension, frequently presents with visual symptoms of visual obscurations, peripheral visual field defects (in up to 90% of cases), and diplopia. Other symptoms and findings include headache (often severe), pulsatile tinnitus, papilledema, and elevated intracranial pressure with normal CSF studies. Medical treatment options include weight loss, diuretics, serial lumbar puncture, and steroids. Acetazolamide is the most commonly used and effective medical treatment, but is fraught with side effects. In up to 10% of cases, severe and progressive visual loss can occur.

Traditionally, it has been felt that neuroimaging studies are unremarkable in cases of pseudotumor, except for the presence of an empty sella. A 1998 study by Brodsky<sup>1</sup> identified specific MRI findings in cases of papilledema with elevated intracranial pressure. Posterior scleral flattening was present in 80%, empty sella in 70%, periorbitic subarachnoid space distention in 45%, prelaminar optic nerve enhancement in 50%, along with other findings that gave an overall 90% prediction of elevated intracranial pressure when certain MRI criteria were present.

In cases of progressive visual loss in spite of maximum tolerated

medical therapy, optic nerve sheath decompression has been found to be relatively safe and effective. An October 2000 study by Banta<sup>2</sup> demonstrated the effectiveness of optic nerve sheath decompression in these cases. 158 eyes of 86 patients experiencing progressive visual loss were treated with optic nerve sheath decompression. After surgery, visual acuity stabilized or improved in 94% of eyes (148 or 158), and peripheral visual fields stabilized or improved in 88% of eyes (71 or 81). In patients with headaches as a presenting symptom, only 13% had subjective improvement of headaches after optic nerve sheath decompression in spite of stable visual parameters. Surgical complications were transient, with the most common being temporary double vision. One optic nerve had a presumed traumatic optic neuropathy with loss of vision.

Weight loss may be a successful method of treatment in pseudotumor, but a study by Johnson et al found that a 6.2% reduction in body weight was required for papilledema to resolve. Visual field defects were also found to improve with weight loss.

<sup>1</sup>Brodsky MC et al. *Ophthalmology* 1998;105:1686-1693.

<sup>2</sup>Banta JT et al. *Ophthalmology* 2000;107:1907-1912.

## Simultaneous Visual Loss (from page 1)

pendent vascular supplies to each eye and visual pathway, it is relatively uncommon to have both visual pathways simultaneously involved by a disease process. Discussed below is a selection of disorders capable of causing a sudden, simultaneous or nearly simultaneous, bilateral severe loss of vision.

### Pituitary Apoplexy:

Pituitary Apoplexy is an acute change in the volume of a pituitary adenoma as a result of spontaneous hemorrhage, edema, or necrosis. Infarction or hemorrhage into a non-tumorous gland can occur, as during childbirth ("Sheehan's Syndrome"). Symptoms include an acute severe headache, bilateral visual loss due to the crossing of the optic nerves over the expanding gland, bilateral or unilateral ocular motility disturbances due to expansion into the adjacent cavernous sinuses, and epistaxis. Enlargement of the sella is seen on plain radiological films, and fresh hemorrhage is seen on CT or MRI. Corticosteroid replacement and supportive measures are critical, and surgical decompression through the sphenoid sinus should proceed promptly.

### Leber's Optic Neuropathy:

Leber's optic neuropathy primarily affects young males in the second and third decades of life. While some females are affected, the disorder primarily conforms to a mitochondrial inheritance pattern. In this disorder, the central vision is suddenly lost bilaterally (within days or weeks of each other), with visual acuity dropping to the 20/200 level or worse rapidly and usually permanently. Systemic findings include progressive muscular dystonia and cardiac "pre-excitation syndrome".

### Arteritic Ischemic Optic Neuropathy:

While Giant Cell Arteritis (Temporal Arteritis) does not typically lead to an immediately simultaneous visual loss, the risk of bilateral loss is significant and preventable. Typically a disease of elderly individuals over the age of 70, symptoms include headache,

weight loss, fatigue, polymyalgia rheumatica, jaw claudication, fever, and severe visual loss due to ischemic optic neuropathy or central retinal artery occlusion. Sed rate levels are typically markedly elevated, and temporal artery biopsy is diagnostic. Due to the risk of bilateral visual loss, systemic steroids should be initiated immediately, as long as the biopsy is performed within two weeks.

### Cortical Blindness:

Cortical or cerebral blindness refers to visual loss following bilateral occipital cortex lesions. (Hemispheric lesions do not reduce visual acuity, but instead lead to peripheral vision defects.) Causes of cortical blindness include stroke, post-angiography, post-cardiopulmonary bypass, hypotension, meningitis, trauma, Creutzfeldt-Jacob disease, and others. The degree of visual loss and peripheral visual loss are usually symmetrical between the two eyes. Pupillary reaction and optic nerve appearance appear normal. In some cases, it may be difficult to distinguish cortical visual loss from non-physiological (hysterical) visual loss. In Anton's Syndrome, cortical blindness is accompanied by a denial of blindness. The patient is often able to confabulate details about his surroundings that give the appearance of accurate visual abilities.

### Infiltrative Lesions of the Optic Nerve and Meninges:

Leptomeningeal infiltration by metastatic cancer can lead to acute or subacute bilateral visual loss. Involvement of the second optic nerve is usually rapid and relentless. Fundoscopic findings are normal.

Infiltration of the optic nerves can occur in sarcoidosis, acute forms of lymphocytic leukemia, histiocytosis, and in opportunistic infections of the CNS such as cryptococcus and toxoplasmosis. Visual loss may be profound and rapid in these cases. In some noncarcinomatous infiltrations (such as leukemia, lymphoma), radiation therapy may allow recovery of useful vision.

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- Extensive patient information, including discussion of over 80 eye conditions and a section discussing risks and benefits of laser vision correction.
- Interactive Clinical Section concerning eye disease and physical findings
- Clinical Trials Database

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*Ophthalmology Update*

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**The CHAMPS study (page 1)**

ing event becomes more critical, since the treatment that can be offered shows definite longer-term benefit. Immediate treatment with IV steroids (which speed visual recovery) followed by Avonex appears to be indicated in these cases to delay the onset of MS, rather than just simply observing the patient for new findings. The major side-effect of the injections of Avonex was a flu-like illness that developed within the first six months of injections.

Neuro-ophthalmic findings of Multiple Sclerosis are most often those of optic neuritis. Overall, visual difficulties are the presenting complaints of 30-40% of patients diagnosed with MS. Other visual disturbances can occur, such as double vision, as well as cerebellar dysfunction.

**Optic Neuritis:**

Optic neuritis typically presents as an acute, unilateral loss of vision often with pain on eye movement. Important findings include:

- Acute unilateral loss of vision or visual field. Color vision abnormalities often occur, with colors such as red appearing washed out compared to the other eye.
- Patient is typically under 40 years of age.
- 90% have periocular pain, worsened by eye movement.
- A Relative Afferent Pupillary Defect (RAPD) is an extremely important physical finding, and is present even in cases of mild visual loss. The RAPD may not be present if the other eye has experienced an episode of optic neuritis in the past.

- 35% have optic nerve head edema, while in 65% the optic nerve inflammation is retrobulbar, and thus not visible on ophthalmoscopy.
- Good visual acuity recovers over time (within 3 weeks).

It is important to distinguish optic neuritis from Ischemic Optic Neuropathy, more commonly seen in older patients. Typically vascular risk factors are present, there is a lack of pain, and the optic nerve shows a pale edema. Giant cell arteritis should be ruled out in appropriate cases.

**Double Vision:**

Double vision can present as a visual disturbance in early MS. A palsy of the sixth cranial nerve is the most common isolated nerve paralysis, leading to an intorted eye, and an inability to turn the eye outward beyond the midline. Double vision is usually severe and constant, unless the eye is patched.

Internuclear ophthalmoplegia is a more commonly found cause for double vision in MS, occurring in 35-50% of patients. This lesion in the medical longitudinal fasciculus blocks the inturning of one eye when the other eye turns out. The eyes appear to move at different speeds when looking from side to side, and nystagmus may be present.

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<sup>1</sup>Jacobs LD et al. Controlled High-Risk Subjects Avonex MS Prevention Study. N Engl J Med 2000;343:989-904.

<sup>2</sup>Optic Neuritis Study Group. Optic Neuritis Treatment Trial. Arch Ophthalmol 1991;109:1673-1678.