# WriteClick: Editor's Choice

**Editors' Note:** In reference to the article "In vivo identification of morphologic retinal abnormalities in neuromyelitis optica," Dr. Borruat reports his own experience in diagnosing microcystic macular edema in patients with noninflammatory optic neuropathies, supporting the hypothesis of retrograde transsynaptic degeneration of inner retinal cells as etiology. Dr. Khoo espouses the benefits of Sniffin' Sticks, an inexpensive, noninvasive test of olfactory dysfunction, in diagnosing idiopathic intracranial hypertension. Authors Kunte et al. explain the drawbacks of this approach.

*—Megan Alcauskas, MD, and Robert C. Griggs, MD*

## IN VIVO IDENTIFICATION OF MORPHOLOGIC RETINAL ABNORMALITIES IN NEUROMYELITIS OPTICA

**Francois-Xavier Borruat, Lausanne, Switzerland:** Sotirchos et al.[1] used optical coherence tomography (OCT) and found microcystic macular edema (MME) in a cohort of patients with neuromyelitis optica (NMO). The authors considered the possible mechanisms leading to MME in NMO: a primary retinal inflammatory process, a retinopathy related to the high expression of aquaporin-4 in the inner retina, possible unrecognized episodes of uveitis, or a retrograde transsynaptic degeneration of inner retinal cells.

The authors recommended further OCT studies in other types of optic neuropathies to exclude or confirm a transsynaptic etiology of MME. During the past year, I diagnosed MME in 3 patients who presented with noninflammatory optic neuropathies: optic disc drusen, optic nerve compression from craniopharyngioma, and unilateral traumatic optic neuropathy. Similar to others,[2,3] I believe that MME does not reflect a specific underlying inflammatory process but rather I consider it a marker of the severity of optic neuropathies. MME probably reflects a certain stage of retrograde transsynaptic degeneration,[4,5] which can occur in optic neuropathies whatever the etiology.

**Author Response: Elias S. Sotirchos, Peter A. Calabresi, Baltimore:** We thank Dr. Borruat for his comments on our study, in which we reported MME of the inner nuclear layer in 30% of eyes that had experienced NMO-associated optic neuritis.[1] MME was associated with severe retinal axonal and neuronal loss and visual disability. We proposed that MME may develop as a consequence of either retrograde transsynaptic degeneration or a retinal inflammatory process. MME has also been reported in various noninflammatory optic neuropathies,[2,6] and Dr. Borruat suggests this provides evidence supporting a transsynaptic etiology. We agree that this is a possibility, but this hypothesis does not satisfactorily explain why MME may fluctuate over time[7,8] or why MME develops in only a subset of patients with severe optic neuropathies.[1,5,6]

We believe that severe optic atrophy is necessary but not sufficient to cause MME and that the possibility that secondary factors contribute to the development of MME must be considered. Secondary factors could include retinal inflammation[1,7,8] with a humoral or cell-mediated immune response gaining access to the immune-privileged retina after breakdown of the blood–retinal barrier, a mechanical process such as vitreoretinal traction due to severe retinal atrophy resulting in schesis,[6] or a genetic susceptibility to more extensive neurodegeneration.



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3. Balk LJ, Killestein J, Polman CH, Uitdehaag BMJ, Petzold A. Microcystic macular oedema confirmed, but not specific for multiple sclerosis. Brain 2012;135:e226.
4. Van Buren JM. Trans-synaptic retrograde degeneration in the visual system of primates. J Neurol Neurosurg Psychiatry 1963;26:402–409.
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8. Saidha S, Sotirchos ES, Ibrahim MA, et al. Microcystic macular oedema, thickness of the inner nuclear layer of the retina, and disease characteristics in multiple sclerosis: a retrospective study. Lancet Neurol 2012;11:963–972.

## OLFACTORY DYSFUNCTION IN PATIENTS WITH IDIOPATHIC INTRACRANIAL HYPERTENSION

**Kah Fang Khoo, Bayan Lepas, Malaysia:** This interesting pilot study[1] provides new evidence that patients with idiopathic intracranial hypertension (IIH) may have olfactory dysfunction. This feature is added to more conventional symptoms like headache, nausea, vomiting, and double vision. This is helpful as the Sniffin' Sticks procedure is less invasive than lumbar puncture and less expensive than MRI. Schmidt et al.[2] demonstrated reduction of olfactory bulb volume using MRI. A future study should include reduction of olfactory function as one of the minor criteria for diagnosing IIH and rule out space-occupying lesions and high intracranial pressure.

**Author Response: Hagen Kunte, Felix Schmidt, Jan Hoffmann, Lutz Harms, Golo Kronenberg, Berlin:** We appreciate the suggestion that an assessment of olfactory function by the extended Sniffin' Sticks procedure may be helpful in the diagnosis and follow-up of IIH. The prevalence of absolute hyposmia in patients with a recent first diagnosis of IIH or patients with a clinically significant worsening of IIH within the last 3 months reached 80% in our sample.[1] We agree that the presence of olfactory dysfunction should be considered as one of the minor criteria for diagnosing IIH. However, other potential causes of reduced olfactory function must first be ruled out. In contrast, olfactory testing is less appropriate as a follow-up measure in patients with diagnosed IIH because olfactory function typically trails changes in intracranial pressure (ICP) by several weeks. Emerging evidence suggests that 3D spectral-domain optical coherence tomography of the optic nerve head might be used to noninvasively monitor IIH and possibly other disorders with increased ICP.[3]



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2. Schmidt C, Wiener E, Hoffmann J, et al. Structural olfactory nerve changes in patients suffering from idiopathic intracranial hypertension. PLoS One 2012;7:e35221.
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## CORRECTION

**Teaching Neuro*Images*: Dyspnea as a presenting manifestation of amyloid myopathy**

In regard to the article "Teaching Neuro*Images*: Dyspnea as a presenting manifestation of amyloid myopathy" by P.S. Ghosh et al. (*Neurology*® 2013;81:e184), there is an error in the cover image text found at the end of the Table of Contents. It should have read: "Trichrome stain of a deltoid muscle showing a necrotic muscle fiber in a patient with amyloid myopathy." The authors regret the error.

*Author disclosures are available upon request (journal@neurology.org).*

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