Editors' Note: This week's WriteClick submitters offer their own data to either support or question the conclusions of 2 studies. In response to the study by Dr. Defazio et al. linking ocular pathology and the development of blepharospasm, Dr. Kurlan proposes, as a possible mechanism, that chronic ocular irritation could lead to abnormal sensory feedback that disturbs central motor control signals, thereby causing blepharospasm. He provides supportive examples from his own research into patients with Meige syndrome. Dr. Kashyape, in response to the study by Dr. Sharma et al. into temporal lobe pathology in epilepsy of infancy with migrating focal seizures, presents data from one of his patients with this epilepsy syndrome who had a nonfocal brain MRI but hippocampal sclerosis on autopsy. The authors respond that they continue to contend that this disease is not a structural epilepsy but that more research should be pursued.

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

ENVIRONMENTAL RISK FACTORS AND CLINICAL PHENOTYPE IN FAMILIAL AND SPORADIC PRIMARY BLEPHAROSPASM

Roger Kurlan, Summit, NJ: I read with interest in the article by Defazio et al.¹ Their study revealed an evident relationship between eye symptoms and diseases (dry eyes, blepharitis, keratoconjunctivitis) and the occurrence of the dystonic condition primary blepharospasm, both familial and sporadic forms. Although not discussed in their article, I wonder if this link provides insights into the potential underlying mechanisms of blepharospasm.

We previously described a series of patients demonstrating coexistent Meige syndrome (blepharospasm/oromandibular dystonia) and myasthenia gravis.² We raised the possibilities that 1) eyelid/extraocular muscle fatigue might lead to blepharospasm, perhaps by an abnormal sensory feedback process, and 2) there might be a shared autoimmune basis. The results of the Defazio et al. study may support these possibilities. The ocular symptoms identified in the report by Defazio et al. are typically manifest by dry, irritated eyes which could potentially lead to abnormal sensory feedback that influences the development of disturbed central motor

control signals, causing dystonia. A similar abnormal sensory feedback process might result from weak eyes of myasthenia gravis. Autoimmune diseases commonly result in dry, irritated eyes, which might explain a link to blepharospasm.

Although controversial,³ localized dystonia following peripheral trauma has been described, again suggesting the possibility of an abnormal sensory-motor loop. In Tourette syndrome it is the form of tics termed dystonic tics⁴ (motor tics resembling dystonia, such as torticollis-like head/neck twisting or blepharospasm) that are most associated with premonitory sensations, such as an irritation in the eyes preceding eyelid closure tics, causing patients to feel like they need to tic in order to relieve the uncomfortable sensations. In this situation, a centrally generated sensory disturbance (compared to peripherally generated with irritated or weak eyes) might produce the abnormal feedback to the motor system resulting in dystonia.

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CHILD NEUROLOGY: EPILEPSY OF INFANCY WITH MIGRATING FOCAL SEIZURES

Pawan S. Kashyape, Katharine Forrest, Southampton, UK: In their review, Sharma et al.¹ stated that neuroimaging was normal in all reported patients. We wish to draw attention to 4 reported cases and our experience of a single case of hippocampal sclerosis in epileptic encephalopathy. Caraballo et al.² reported 3 cases with radiologic evidence of hippocampal sclerosis on MRI scans yet the exact timing

of these scans in relation to seizure onset was unclear. Coppola et al.³ found left temporal dual pathology on MRI in one case, including hippocampal sclerosis and cortical-subcortical blurring with left temporal lobe atrophy. Three case reports of autopsy have also been published: one was normal and the other two showed gliosis of the CA1 sector of the pyramidal layer of the hippocampus.^{4,5} Our case had no focal signs on MRI brain scan but revealed bilateral hippocampal segmental neuronal loss and gliosis affecting CA1 and CA4 sectors with patchy cortical damage consisting of astrocyte gliosis and relatively good neuronal preservation. The relationship between hippocampal sclerosis and this epilepsy syndrome needs to be further elucidated by high-resolution neuroimaging and neuropathologic evidence from autopsy in fatal cases.

Author Response: Suvasini Sharma, Naveen Sankhyan, Konanki Ramesh, Sheffali Gulati, New Delhi: Drs. Kashayape and Forrest correctly point out that neuroimaging findings have been reported in a few patients with epilepsy of infancy with migrating focal seizures.^{2,3} However, we contend that “epilepsy of infancy with migrating focal seizures” is not a structural epilepsy.⁵ Coppola et al.³ emphasize that hippocampal sclerosis in their patient was unlikely to contribute to the pattern of migrating partial seizures and fulfilled the diagnostic criteria for mi-

grating partial seizures in infancy independent of the temporal lobe lesion. In the report by Caraballo et al.,² the exact timing of the MRI scans in relation to seizure onset is unclear, so the cause for the hippocampal sclerosis is unknown. We thank Drs. Kashayape and Forrest for sharing these cases and their experience with autopsy evidence of hippocampal segmental neuronal loss and gliosis in children with this syndrome. We agree that the relationship between hippocampal sclerosis and this epilepsy syndrome warrants further study.

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