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Jeavons Syndrome: An Overlooked Epilepsy Syndrome

To the Editor:

We read with great interest the study by Smith et al.[1] on Jeavons syndrome (JS). JS is characterized by a triad of eyelid myoclonia with or without absence, eye-closure–induced electroencephalography (EEG) paroxysms, and photosensitivity. Smith et al. have highlighted frequent under-recognition, female preponderance, and antiepileptic drug refractoriness in JS. We wish to share our experience with this electroclinical diagnosis over recent few years. We encountered three patients (two girls and one boy) with this syndrome and the diagnosis was missed on mutiple outpatient visits in all three children. Median delay in diagnosis was 30 months (range four months to nine years).

Two of these children presented with complaints of poor scholastic performance and intermittent eyelid flickering. They were misdiagnosed with intellectual disability with tics. The third child, a 30-month-old girl, presented with hyperactivity and inattentiveness. Hence, the subtle symptoms and nonidentification of events by parents may have misled the child's physician. Two patients who were misdiagnosed even by an attending neurologist had been evaluated for epilepsy but their brain magnetic resonance imaging and sleep EEG were normal. Therefore the diagnosis was again missed in these two children. Later, an awake EEG with photic stimulation clinched the diagnosis. All three individuals developed refractory epilepsy. Antiepileptic drugs included sodium valproate, levetiracetam, lamotrigine, and clobazam. The ketogenic diet was tried in two patients and one was responsive.

On the basis of our experience, we concur with the observation of Wang et al. that JS is an under-recognized and difficult-to-treat epilepsy. Causes for under-recognition include misdiagnosis of events such as tics and the poor diagnostic yield of sleep EEG.[2] Seizure semiology and EEG with unique signatures provoked by eye-closure are usually diagnostic. In relevant children, it is important to obtain an awake EEG with photic stimulation (especially in younger children) lest both parents and physicians miss the diagnosis.

The recent International League Against Epilepsy classification has renamed JS as "epilepsy with eyelid myoclonias." This rare condition may be "out of sight and out of mind." Recognition of this entity as a separate epilepsy syndrome by International League Against Epilepsy is a laudable step which might bolster its early recognition.

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