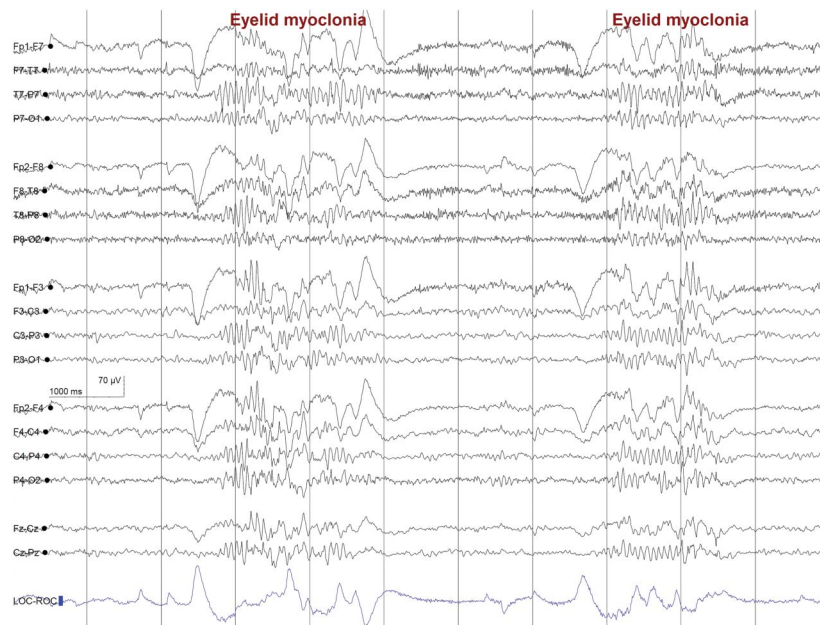


# Mystery Case: Eyelid myoclonia with absences in an adult patient



**Figure 1** Eyelid myoclonia without absence



With levetiracetam: posterior-predominant spiky alpha frequency activity that rapidly spreads to the frontal regions coinciding with eyelid myoclonia.

A 28-year-old man presented to the epilepsy monitoring unit (EMU) with frequent “eye fluttering” episodes since he was 3 years old (video on the *Neurology*<sup>®</sup> Web site at [www.neurology.org](http://www.neurology.org)). He was diagnosed with epilepsy as a teenager after he developed generalized convulsions at age 12. His convulsions were well-controlled with antiepileptic drug therapy. His neurological examination was normal.

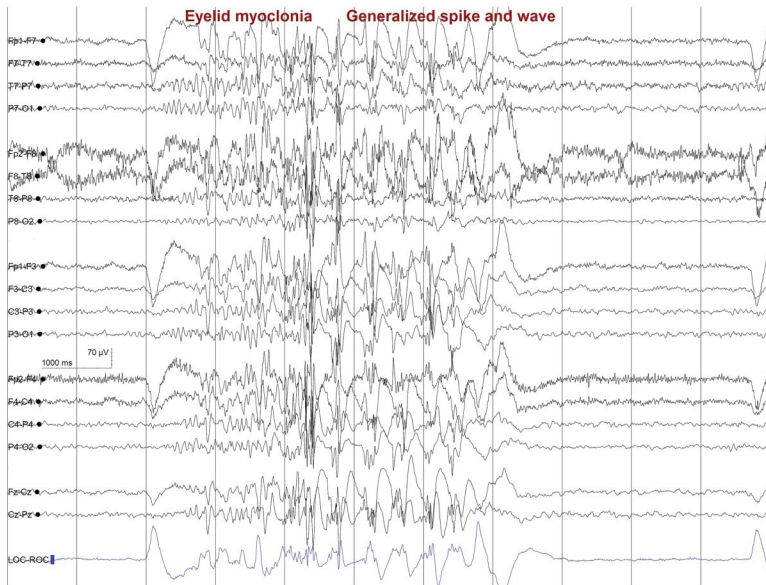
His EMU study revealed brief episodes of eyelid myoclonia (video) coinciding with a paroxysmal “spiky” posterior alpha activity, which rapidly spread to the frontal regions while he was on levetiracetam (figure 1). These episodes were more prominent in light compared to dark and were often triggered by eye closure. Interictally, the posterior dominant rhythm appeared sharply contoured. Discontinuation of levetiracetam for 3 days resulted in 2 to 3 Hz generalized polyspike and wave activity associated with some of the episodes of eyelid myoclonia, within seconds of eyelid closure (figure 2). Additionally, in sleep there

were fragmentary bursts of anteriorly predominant 3 Hz abortive spike and wave discharges. Prior records indicated a photoparoxysmal EEG response.

The patient’s clinical and electrographic picture is consistent with eyelid myoclonia with absences (EMA), also known as Jeavons syndrome.

EMA is an underrecognized syndrome of unknown etiology defined by the triad of childhood onset, photosensitivity, and eyelid myoclonia with or without absence seizures.<sup>1,2</sup> Patients often develop generalized tonic-clonic seizures in adolescence.<sup>1,3</sup> The photosensitivity component may decrease as the patients get older.<sup>1,4</sup> Classic EEG features include a sharply contoured or spiky alpha frequency activity that rapidly spreads to the frontal regions with sustained eye closure. Eyelid myoclonia often coincides as well with occipital epileptic discharges or occipital polyspike and wave discharges, with or without generalized polyspike and wave discharges.<sup>3</sup> The overall prognosis of this syndrome is often good as the generalized tonic-clonic and absence

**Figure 2** Eyelid myoclonia with absence



After discontinuation of levetiracetam: generalized polyspike and wave discharges within seconds of eyelid myoclonia onset.

seizures become controlled with antiepileptic drug therapy. Despite the sensitivity of the absences to antiepileptic drugs, the eyelid myoclonia often persists, as seen in our patient.<sup>4</sup> A small proportion of patients may continue to have uncontrolled generalized convulsions.<sup>4</sup> Valproic acid, lamotrigine, and levetiracetam are good treatment options,<sup>1,5</sup> whereas sodium channel agents such as carbamazepine may exacerbate the seizures.<sup>6</sup> Our patient initially responded to valproic acid, but he developed a reaction concerning for Stevens-Johnson syndrome. With levetiracetam treatment, his generalized tonic-clonic and absence seizures resolved.

Most adult patients with EMA are diagnosed in childhood, but making the diagnosis in adults for the first time may be challenging. Neurology residents and fellows should be aware of the characteristic clinical and electrographic features of this underrecognized syndrome.

#### AUTHOR CONTRIBUTIONS

Dr. Hannawi analyzed and interpreted the data, drafted and revised the manuscript. Dr. Satpute analyzed and interpreted the data. Dr. Maheshwari analyzed and interpreted the data, reviewed and revised the manuscript.

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#### DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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#### MYSTERY CASE RESPONSES

The Mystery Case series was initiated by the *Neurology*<sup>®</sup> Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool. Responses were solicited through a group e-mail sent to the American Academy of Neurology Consortium of Neurology Residents and Fellows and through social media. All the answers that we received came through social media, from individuals rather than groups.

Most of the respondents (66%) correctly indicated Jeavons syndrome as the most likely diagnosis. The other preferred response was absence epilepsy. The most complete answer came from Dr. Felipe Borlot (Clinical Fellow, Toronto Western Hospital and University of Toronto, Canada). In his response, he pointed out that the key element for this Mystery Case is the fact that these reflex seizures are induced by eye closure and that the epileptiform abnormalities disappear with eye opening. The eyelid myoclonia in this patient is characteristic. Jeavons syndrome can be misdiagnosed as childhood or juvenile absence epilepsy, other forms of genetic or idiopathic generalized epilepsies, or even facial tics.

This Mystery Case illustrates a classic epilepsy syndrome, usually refractory to treatment, which persists throughout life.

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