Teaching Video NeuroImages: Epilepsy with myoclonic absences
A distinct electroclinical syndrome

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A 10-year-old girl presented with recurrent absence spells of 6 years’ duration. Video-EEG revealed absences with rhythmic unilateral shoulder jerks, classic of epilepsy with myoclonic absences (EMA) (videos 1 and 2 on the Neurology® Web site at www.neurology.org; figure).

The average age at onset is 7 years.1 EMA may be associated with trisomy 12p and Angelman syndrome.2 It has a variable prognosis; cognitive deterioration occurs proportionate to duration of intractable epilepsy. Seizures persist into adulthood in approximately 50% of cases. While a valproate–ethosuximide combination is best, alternatives include valproate with benzodiazepines, phenobarbital, and lamotrigine.1 Video-EEG correlation is recommended to differentiate it from childhood absences and eyelid/perioral myoclonia with absences. Presence of focal semiology should not deter the diagnosis of this distinct generalized epilepsy syndrome.

REFERENCES
Teaching Video NeuroImages: Epilepsy with myoclonic absences: A distinct electroclinical syndrome
Ramshekhar Menon, Neeraj N. Baheti, Ajith Cherian, et al.
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