Teaching Video NeuroImages:

Epilepsy with myoclonic absences

A distinct electroclinical syndrome

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Figure Interictal EEG

Interictal record showed generalized and multifocal spike and wave discharges on a normal background of 8–9 Hz.

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.

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