

# Teaching Video NeuroImages:

## Epilepsy with myoclonic absences

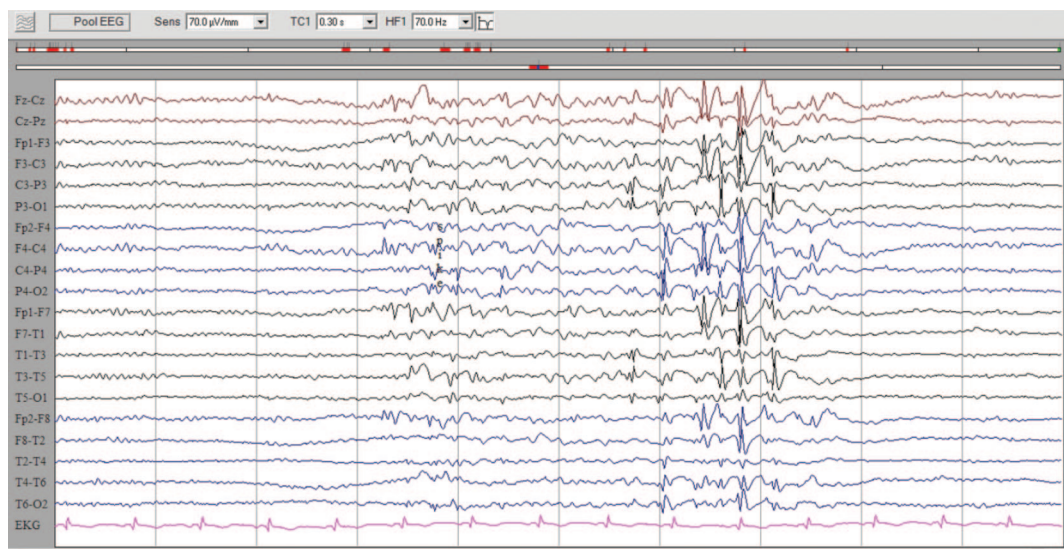
A distinct electroclinical syndrome



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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*<sup>®</sup> Web site at [www.neurology.org](http://www.neurology.org); figure).

The average age at onset is 7 years.<sup>1</sup> EMA may be associated with trisomy 12p and Angelman syndrome.<sup>2</sup> 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.<sup>1</sup> 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

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2. Elia M, Guerrini R, Musumeci SA, Bonanni P, Gambardella A, Aguglia U. Myoclonic absence-like seizures and chromosome abnormality syndromes. *Epilepsia* 1998;39:660–663.

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

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*Neurology* 2011;76:e113

DOI 10.1212/WNL.0b013e31821e54c9

**This information is current as of June 6, 2011**

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