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Teaching Video NeuroImages: Epilepsy with myoclonic absences

A distinct electroclinical syndrome

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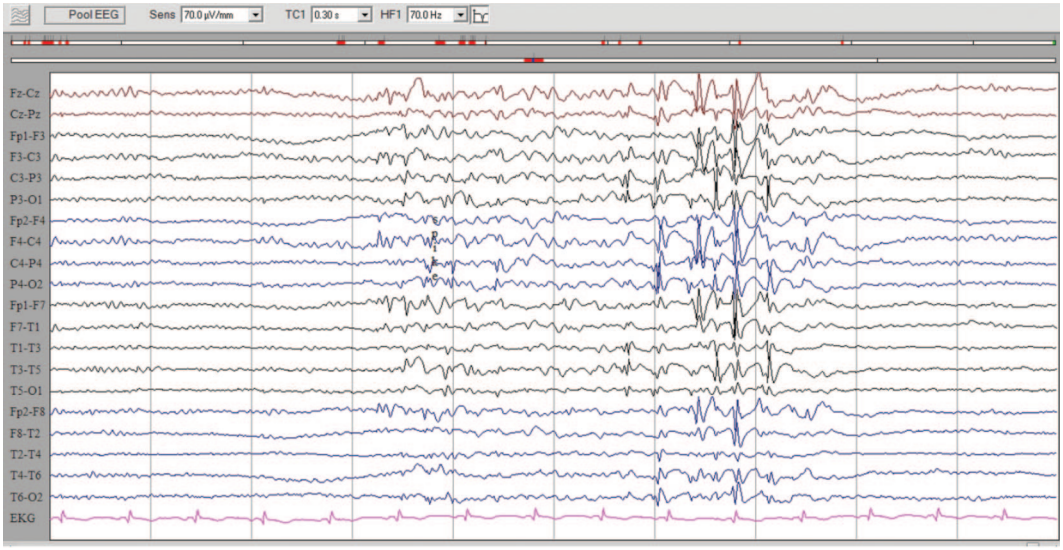
Figure Interictal EEG



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.

REFERENCES

1. Bureau M, Tassinari CA. Epilepsy with myoclonic absences. Brain Dev 2005;27:178–184.
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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