

## Eyelid myoclonia with absences (Jeavons syndrome): still an overlooked epilepsy syndrome. Comments to Covanis review in this issue of *Journal of Epileptology*

Salvatore Striano<sup>1</sup>, Pasquale Striano<sup>2</sup>

<sup>1</sup> Epilepsy Center, Department of Neurological Sciences, Federico II University, Napoli, Italy

<sup>2</sup> Pediatric Neurology and Muscular Diseases Unit, Institute G. Gaslini and University of Genova, Genova, Italy

Received November 5, 2015

Accepted for publication on-line November 6, 2015

Published on-line November 17, 2015

### Correspondence

Prof. Salvatore Striano, MD  
Epilepsy Center  
Federico II University  
Via Pansini 5, 80131, Napoli, Italy  
Phone/fax: 39 81 746 35 09  
e-mail: sstriano1@gmail.com

The epileptic phenotype featuring eyelid myoclonia (EM) and absences at eye closure plus photosensitivity (EMA or ELMA) was first reported more than 80 years ago by Radovici et al. (1932) and then fully described by Jeavons in 1977. EMA typically starts in childhood with long-lasting and often drug-resistant course with high risk of recurrence of generalized tonic-clonic seizures (GTCS).

In fact, EM as well as eye closure sensitivity and photosensitivity may also occur in other forms of generalized or focal epilepsies. Likewise, different seizure types observed in genetic generalized epilepsies (GGE), namely, absences, myoclonic jerks and tonic-clonic seizures can occasionally occur in EMA (Striano et al., 2009).

However, if we put together: a) the specific seizure type (i.e., EM followed by a short absence on closing the eyes); b) the ictal EEG pattern (i.e. oculo-palpebral artefact related to eye closure immediately followed by a generalized discharge of multiple spikes, then by a short sequence of 2- max 6 sec of generalized spike and waves about 3/sec) (Fig. 1); c) the almost constant photosensitivity, with the possibility of induction of seizures of the same type, but also of massive myoclonic jerks or GTCS, in some case originating in occipital cortex (Fig. 2); d) the age-related onset and long-lasting duration, this condition shape clearly the well-defined syndrome described by Dr. Peter Jeavons. Obviously, the full electroclinical picture may modify over

the time, in consequence of antiepileptic treatment or in relation to age. Indeed, some patients may show persistence of eyelid fluttering even without concomitant EEG abnormalities (Fig. 3), probably as behavioural habit in subjects with a previous history of self-induced seizures. In this issue of *Journal of Epileptology*, Covanis presents a comprehensive and useful review which includes different aspects of the syndrome and points again to the need for a correct recognition and definition of the syndrome. This is the first step to address further neurophysiological (Viravan et al., 2001), neuroimaging (Liu et al., 2008; Vaudano et al., 2014) as well as genetic studies (Galizia et al., 2015) aimed to understand better this intriguing condition. Nevertheless, EMA so far remains an overlooked epilepsy syndrome.

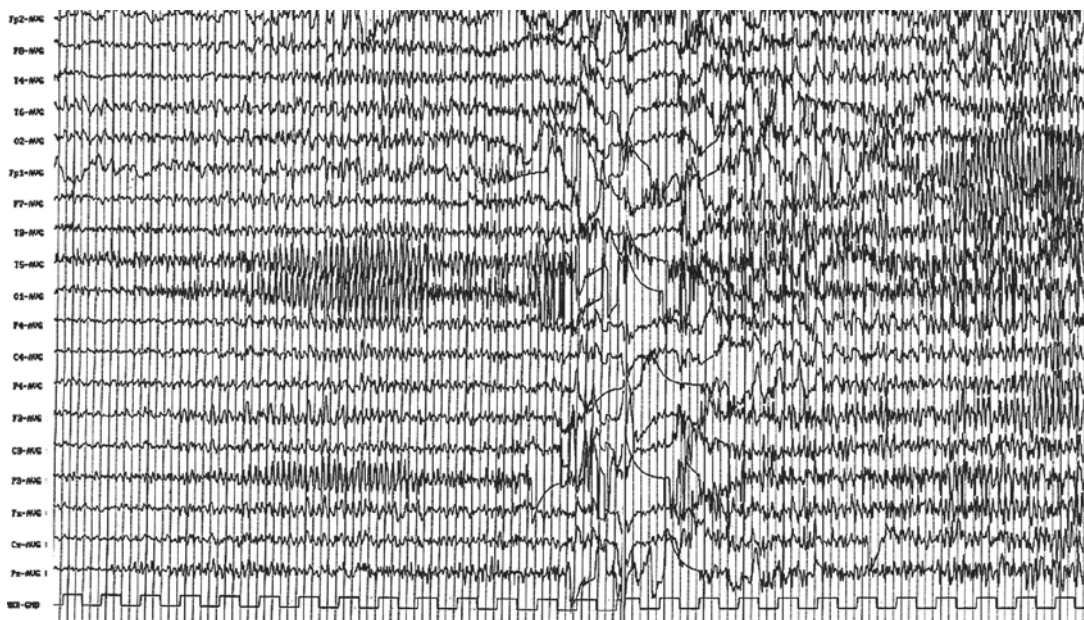
**Key words:** eyelid myoclonia absences • EMA • ELMA • syndrome

### REFERENCES

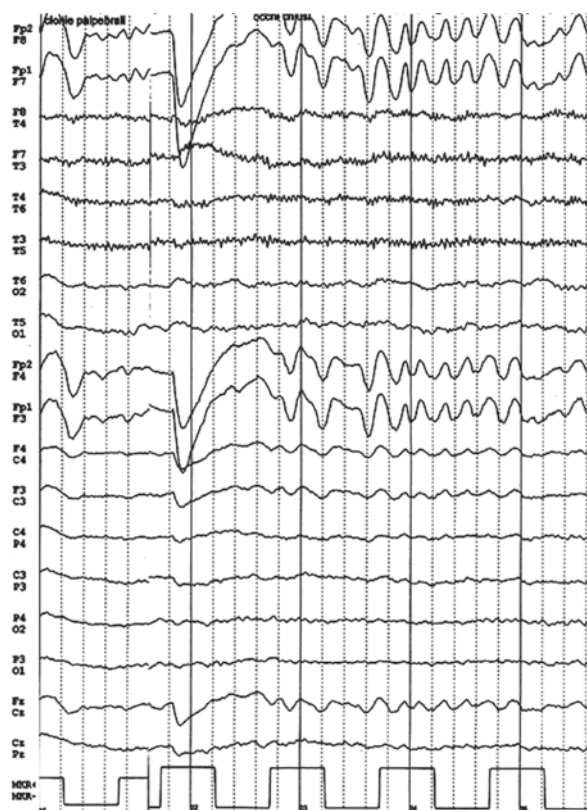
- Covanis A.: *Jeavons syndrome-Updated review*. Journal of Epileptology, 2015, this issue.
- Galizia E.C., Myers C.T, Leu C., de Kovel C.G., Afrikanova T., Cordero-Maldonado M.L. et al.: *CHD2 variants are a risk factor for photosensitivity in epilepsy*. Brain, 2015, 138 (Pt 5): 1198–1207.
- Jeavons P.M.: *Nosological problems of myoclonic epilepsies in childhood and adolescence*. Dev. Syndrom Med., 1977, 19: 3–8.
- Liu Y., Yang T., Liao W., Yang X., Liu I., Yan B.: *EEG-fMRI*



**Figure 1.** Eyelid myoclonia followed by a short absence on closing the eyes. Ictal recording shows the oculo-palpebral artifact followed by generalized multiple spikes and then spike and waves at  $>3$  c/sec. The square horizontal bars at the bottom of the record mark 1 s; vertical bars =  $100 \mu\text{V}$ ; filter: LF 0.2 Hz, HF 40 Hz.



**Figure 2.** Ictal recording of a tonic-clonic seizure in a teenager with a Jeavons syndrome. EEG shows a recruiting rhythm in left occipital cortex rapidly spreading to the whole brain. The square horizontal bars at the bottom of the record mark 1 s; vertical bars =  $100 \mu\text{V}$ ; filter: LF 0.5 Hz, HF 40 Hz.



**Figure 3.** Eyelid fluttering without paroxysmal activity on closing the eyes in a EMA patient. The square horizontal bars at the bottom of the record mark 1 s; vertical bars = 100  $\mu$ V; filter: LF 0.5 Hz, HF 40 Hz.

study of the ictal and interictal epileptic activity in patients with eyelid myoclonia with absences. *Epilepsia*, 2008, 49: 2078–2086.

**Panayiotopoulos C.P.:** *Jeavons syndrome*. In: C.P. Panayiotopoulos (ed.), *A clinical guide to Epileptic Syndromes and their Treatment*. Springer Healthcare Ltd 2010, 513–517.

**Radovici M.M.A., Misrliou V.L., Gluckman M.:** *Reflex Epilepsie réflexe provoquée par excitations optiques des rayons solaires*. *Rev. Neurol.*, 1932, 1: 1305–1308.

**Striano S, Striano P., Nocerino C., Boccella P., Bilo L., Meo R., Ruosi P.:** *Eyelid myoclonia with absences: an overlooked epileptic syndrome?* *Neurophysiol. Clin.*, 2002, 32: 287–296.

**Striano S., Capovilla G., Sofia V., Romeo A., Rubboli G., Striano P.:** *Eyelid myoclonia with absences (Jeavons syndrome): a well-defined idiopathic generalized epilepsy syndrome or a spectrum of photosensitive conditions?* *Epilepsia*, 2009, 50 (suppl. 5): 15–19.

**Vaudano A.E., Ruggieri A., Tondelli M., Avanzini P., Benuzzi F., Gessaroli G.:** [The visual system in eyelid myoclonia with absences](#). *Ann. Neurol.*, 2014, 76: 412–427.

**Viravan S., Go C., Ochi A., Akiyama T., Carter Snead III O., Otsubo H.:** *Jeavons syndrome existing as occipital cortex initiating generalized epilepsy*. *Epilepsia*, 2011, 52: 1273–1279.