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

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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 and tonic-clonic seizures can occasionally occur in EMA (Striano et al., 2009).

Key words: eyelid myoclonia absences - EMA - ELMA - syndrome

REFERENCES


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 µV; 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 µV; 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 µV; filter: LF 0.5 Hz, HF 40 Hz.