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Blinking and eyelid myoclonia: Characteristics and correlations of eyelid movements

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ABSTRACT

Purpose: Eyelid myoclonia (EM) is considered a seizure type and has been described in several epileptic conditions. Previous studies reported that EM are precipitated only by slow eye closure, but little is known about the characteristics of blinking in patients with EM seizures and differences in precipitation of EM by different kinds of eye closure. We analyzed by video-EEG the characteristics of blinking and eye closure in these patients.
Method: Twenty patients with EM had a video-EEG protocol with eyelid sensors. Semiology and rate of blinking and EM were analyzed. Statistical analysis was performed and p-values <0.05 were considered significant.
Results: Seventeen cases (mean age 20.7, range 3–35) were women, 10 had EM as the main seizure type and between the others, all, but one, had criteria to Juvenile Myoclonic Epilepsy. All patients presented EM, 18 spontaneously during awake, 10 on eye closure and one only during intermittent photic stimulation. EM assumed the form of flicker, flutter or jerk, accompanied by generalized discharges, spiky posterior alpha, theta rhythm or absence of any EEG abnormality. Analysis of the characteristics of blinking had no statistics differences between patients and healthy subjects. The rate of blinks and EM increased during speech and decreased during reading. EM never occurred during blinking or in the dark.
Conclusions: Despite normal physiology of blinking, EM can manifest as jerk, flicker or flutter, with or without EEG abnormalities and independently of IPS, suggesting that eye closure sensitivity seems to include both, a motor and a visual component.

1. Introduction

Eyelid myoclonia (EM) is considered by ILAE Commission as a seizure type[1,2] and has been described in several epileptic conditions.[3] Some authors have applied the eponym Jeavons syndrome (JS) when dealing with a homogeneous condition with childhood onset, frequently resistant to medical treatment, characterized by EM with absences (EMA), eye closure-induced seizures or electrographic paroxysms and photosensitivity (PS).[4–7] Similar electroclinical features of EM in JS with different degrees of association of generalized tonic-clonic seizures (GTCS), myoclonic jerks other than EM, intellectual disability (ID), presence or absence of PS, age of onset and treatment response have also been reported, defining a clinical heterogeneity in patients with EM.[7,8] Most of these have been referred as a variant of JS.[9,10] Some of these characteristics also overlap with other epileptic syndromes, including Juvenile Myoclonic Epilepsy (JME).[11]

In recent clinical and neuroimaging studies, it has been suggested that the occipital visual cortex play a pivotal role in EMA generation in the presence of light, during eye closure.[3,12–14] Activation of brainstem structures has also been proposed in the epileptic neural network of EM[3,14] and some of these structures, such as the nuclei of the oculomotor and facial nerves, are likewise involved in normal blinking.[15]

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Previous studies have demonstrated that cognitive tasks such as speaking, reading and praxis may precipitate discharges or myoclonic seizures in patients with idiopathic generalized epilepsies (IGE) and some of these tasks also modulate the frequency of blinks in normal individuals.[16–19] However, little is known about the characteristics of blinking in patients with EM seizures and differences in precipitation of EM by cognitive tasks and different kinds of eye closure. According to Covanis[8] EM are precipitated only by slow eye closure, not by rapid blinking, but to our knowledge this difference has not been subjected to a specific research.

The objectives of this study were to analyze by video-EEG the characteristics of EM and blinking in patients with EM/EMA and to compare the effect of habitual and cognitive methods of seizure activation or inhibition in these two types of eyelid movements.

2. Materials and methods

2.1. Subjects

Twenty patients with EM/EMA were submitted to a video-EEG protocol, approved by the Ethics Committee, at the Epilepsy Unit of the Hospital São Paulo, Universidade Federal de São Paulo, São Paulo, Brazil, between January 2011 and June 2012.

Inclusion criteria were patients with documented seizures of EM with or without absences and generalized paroxysmal EEG activity triggered by eye closure, with age of epilepsy onset between birth and 25 years. As PS is a highly age-dependent phenomenon, even in JS its absence was not considered exclusion criteria.[20]

Patients with brain lesions, and progressive myoclonic epilepsies were not included in the study.

Twenty healthy individuals were also included in the study to provide information regarding normal characteristics of the blinking.

2.2. Video-EEG monitoring

After having slept at most 4 h, all subjects were submitted to a specific video-EEG protocol (Fig. 1) using a 32-channel digital equipment (Ceegraph software, Bio-Logic Systems Corp., Mundelein, IL, U.S.A.) with electrodes placement according to the 10–20 International Electrode System, in addition to supraorbital, infraorbital and deltoid electrodes, besides palpebral electrodes (a piezoelectric polyvinylidenefluoride film transducer, length 2.0 cm, width 0.5 cm and weight 4.5 g).[21] Medications were maintained in treated patients. Nineteen were using antiepileptic drugs (AEDs) at the time of the video-EEG. Therapeutic scheme was considered inappropriate in one patient taking carbamazepine.

Recording of background activity, awake silently, with eyes open, for 50 min

5 min taking breakfast

5 min of reading a Portuguese text silently
5 min of reading a Portuguese text aloud
5 min of reading an English or a known language text silently
5 min of reading an English or a known language text aloud

5 min of speaking aloud (about their seizures, a very stressful task)

Closing of the eyes: the eyelids are closed and opened each 5 seconds during 5 min
. 5 min by command
. 5 min voluntary
. 5 min passive (the eyelids are opened and closed by the fingers of the examiner)
. 5 min in the dark (using diving goggles sealed to light)

5 min of blink reflex (by threatening about 0.2Hz)

Hyperventilation for 5 min

Photic stimulation on eye closure (?ashes 1, 2, 3, 4, 6, 8, 10, 12, 14, 16, 18, 20,25, 30, 50, 60)[a]
Photic stimulation with eyes opened (?ashes 1, 2, 3, 4, 6, 8, 10, 12, 14, 16, 18, 20,25, 30, 50, 60)[a]
Photic stimulation with eyes closed (?ashes 1, 2, 3, 4, 6, 8, 10, 12, 14, 16, 18, 20,25, 30, 50, 60)[a]

Fig. 1. Video-EEG protocol. [a]If a generalized response occurred at a given frequency, the series would begin again going down and up to 60 Hz in frequency until a new generalized response happened.

In healthy individuals, a 5-min recording while awake followed by reading and speaking tasks of the video-EEG protocol was performed.

All eyelid movements were analyzed with the video-EEG camera focusing the eyes of the subjects.

The semiology (flicker, flutter or jerk)[8] and rate of EM (number of EM/EMA per minute) was evaluated during all tasks of the video-EEG protocol.

Spontaneous blinks were analyzed during the first 5 min of awake recording, measured in milliseconds (ms), the downward deflection (closing phase), the upward deflection (opening phase) and total duration (downward + upward deflections). The blinking rate was also analyzed during reading, speaking aloud, hyperventilation (HV) and breakfast.

A single neuropsychologist (K.C.) conducted the neuropsychological examination at the time of video-EEG recording.

2.3. Statistical method

Statistical analysis was performed using Kruskal–Wallis, Mann Whitney, Wilcoxon and Dixon tests. The criterion for statistical significance was $p < 0.05$.

3. Results

3.1. Subjects

Among the patients, 17 were females, aged 3–35 years (mean 20.7) while among healthy individuals 14 were females, mean age 31.9, range 24–42.

In ten patients EM/EMA was the main seizure type, with onset between birth and 10 years (mean 2.8). In these cases, two patients presented all criteria to JS and the others were diagnosed as variants of JS, characterized by ID associated or not with early onset (<2 years) and negative history of PS. In the remaining patients, limb myoclonia seizures were predominant, with EMA onset between 7 and 18 years (mean 12.1). All these patients, but one, fulfilled diagnostic criteria to JME,[22] with limb myoclonia occurring predominantly in the morning.

Induction of the EM with or without absences when exposed to light of the intensity encountered in daily life was reported by 60% of the patients.

The clinical features of patients are summarized in Table 1.

3.2. Interictal EEG

During video-EEG protocol the background EEG was normal in 15 patients and slow in 5/9 with a variant of JS. The interictal EEG showed generalized spike, polyspike, spike-wave and polyspike-and-wave discharges, predominantly in the anterior regions. Two patients had also focal discharges, bilateral temporal in one and occipital in the other. No patient admitted to self-inducing seizures in themselves and self-induction was not demonstrated during or following video-EEG protocol. Three patients had upper limbs myoclonia, isolated during praxis tasks in two (patients 11 and 19) and associated with a cluster of EM during breakfast in one (patient 7).