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Eyelid myoclonia with absences: main features and prognostic factors

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Objectives

Eyelid myoclonia with absences (EMA) is a Genetic Generalized Epileptic syndrome characterized by eyelid myoclonia with or without absences, eye-closure-induced EEG paroxysms and photosensitivity.
The main objectives of our study were to describe the clinical and EEG features of a homogeneous group of patients with diagnosis of EMA and to evaluate the presence of possible prognostic factors.

Materials and methods

We retrospectively selected a cohort of patients with diagnosis of EMA evaluated in the Epilepsy Centre of the Neurological Clinic of Catania.
We considered as T1 the clinical and EEG features of the patients during the first year of disease and as T2 the last follow-up visit and we stratified the patients in two groups: "seizure-free" defined as the absence of any type of seizure for at least two years, and "not seizure-free". We evaluated their electro-clinical characteristics at T1 and at T2.

Results

We enrolled in the study 38 patients with the diagnosis of EMA [5 men (13.2%); median age 29.5 (19-79) years]. The mean follow-up time was 8.2 ± 5.6 years. 7.3 ± 4.8 EEG were evaluated for each patient .

Table 1: Baseline clinical characteristics of the population of study at onset.

	Total n= 38 (%)
Sex (Males)	5 (13.2)
Median age at seizure onset(years)	**12 (range 2-21)**
Median age at follow-up (years)	29.5 (range 19-79)
Median disease duration (years)	14.5 (range 0-69)
Diagnostic delay (years) (mean ± SD)	4.5 ± 10.1
Family history of epilepsy	20 (52.6)
Febrile convulsions	4 (10.5)
Psychomotor delay	4 (10.5)
Status epilepticus	**8 (21.0)**
First seizure type	
•GTCS	**16 (42.1)**
•Absences	6 (15.7)
•Eyelid myoclonia	5 (13.2)
•Absences with eyelid myoclonia	9 (23.7)
•Limbs myoclonia	1 (2.6)
•Other	1 (2.6)
Seizure time of occurrence at onset	
•Wakefulness	36 (94.7)
•Awekening	10 (26.3)
•Sleep	8 (21.1)
Precipitating factors	
•Sleep deprivation	17 (44.7)
•Menstrual cycle	4 (10.5)
•Stress	16 (42.1)

Table 2: Features statistically significant of the study population based on seizure outcome.

Variables	Seizure free n= 8 (%)	Not seizure free n= 30 (%)	p
EEG photosensitivity at T2	0 (0)	14 (46.7)	**0.01***
EEG eye closure sensitivity at T2	2 (25.0)	21(70.0)	**0.02***
Focal interictal EEG features	2 (25)	19 (63.3)	**0.06**

*** p<0.05**

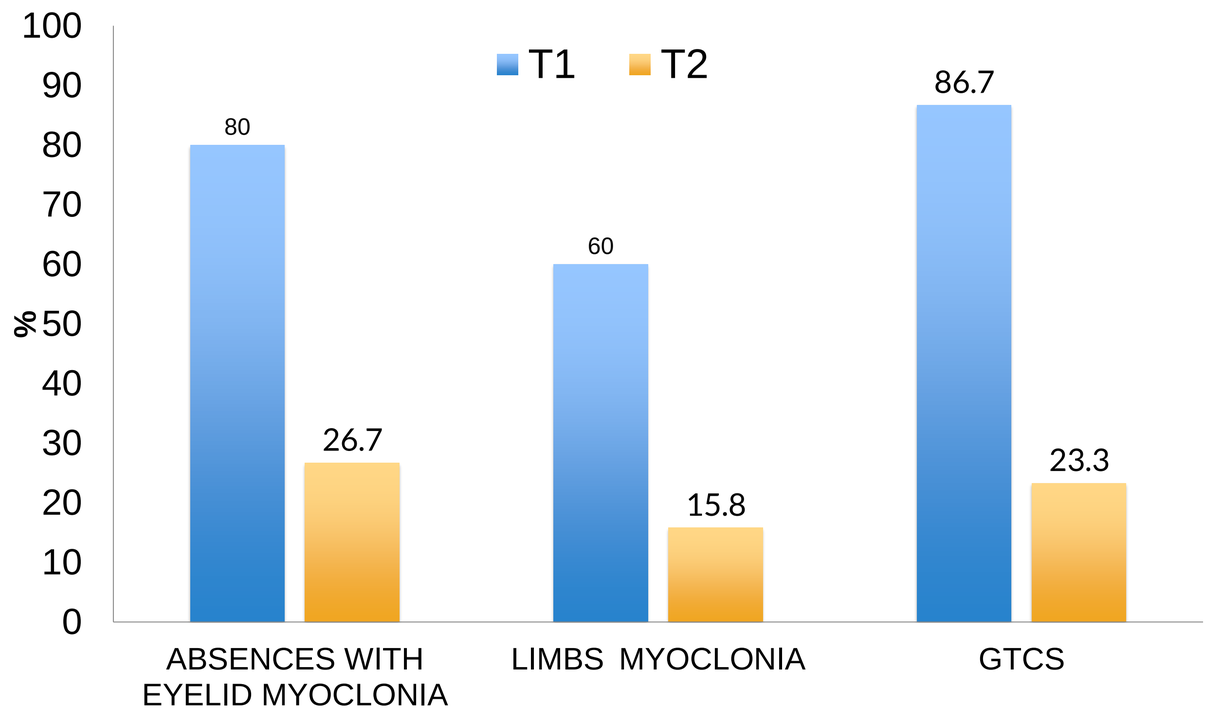

Figure 1: Significant reduction of patients presenting different seizure types between T1 and T2 among the "not seizure-free" group

Conclusions

The findings of our study confirm the presence of definite clinical features of EMA, contributing to the definition of EMA as a separated syndrome. Moreover, our study shows that merely EEG features of EMA can be used as indicators of the long-term outcome of these patients.

References
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