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Status epilepticus in patients with juvenile myoclonic epilepsy: Frequency, precipitating factors and outcome

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ABSTRACT

Status epilepticus (SE) is rarely described in patients with juvenile myoclonic epilepsy (JME), and little is known about its frequency, subtypes, and predictors and the prognosis of these patients. In this retrospective study, we aimed to analyze the incidence of SE in patients with JME and emphasize the risk factors and long-term outcome of SE in an epilepsy outpatient-based cohort.

We included patients with JME with a history of predominant myoclonic seizures and at least one diagnostic EEG with normal background activity and bursts of typical ≥3-Hz generalized spike–polyspike and waves. We investigated the electroclinical features of patients with JME who had experienced SE and compared them with patients with JME without SE.

Of the 133 patients with JME, only 5 patients were diagnosed with SE (3.8%) according to new criteria. The most common SE subtype was myoclonic SE (MSE, 4 patients), followed by generalized clonic–tonic–clonic SE (1 patient) and nonconvulsive SE (1 patient). One patient had both MSE and generalized clonic–tonic–clonic SE. In three out of five patients, recurrent episodes of SE were observed. Same seizure precipitants including sleep deprivation, inappropriate antiepileptic drug treatment, and noncompliance were identified in patients with JME with and without SE, not reaching a significant difference between the groups. Myoclonia limited to specific body parts (one arm, face, or head) were significantly more common in patients with JME with SE (p: 0.002). We did not find any significant correlation with drug-resistant course and SE.

Status epilepticus is rarely observed in patients with JME, and MSE appears to be the most common subtype. Local myoclonia might predict SE in a subgroup of patients with JME. We may suggest that some patients with JME have a liability to SE, in addition to usual seizure precipitating factors of JME. It seems that SE per se does not affect the outcome of JME and the patients with SE did not have drug-resistant course in the final analysis.

1. Introduction

Juvenile myoclonic epilepsy (JME) is one of the most common types of genetic generalized epilepsies and manifests usually at puberty, as its name implies [1]. Electroencephalogram (EEG) shows typical generalized spike–polyspike and waves and often photosensitivity [2]. There are only a handful of case reports and a few small studies about the subtypes, frequencies, and predictors of status epilepticus (SE) in patients with JME [3,4]. Myoclonic status epilepticus (MSE) and nonconvulsive SE (NCSE) are the most frequent subtypes of SE reported in JME. The most commonly reported precipitating factors of SE are sleep deprivation, fatigue, alcohol, inappropriate antiepileptic drugs (AEDs), and abrupt drug withdrawal, but these risk factors were not compared in other patients with JME without SE episodes [3–6].

Our aim was to analyze the frequency of SE in patients diagnosed with JME and emphasize the risk factors and long-term outcome in a tertiary referral epilepsy center.

2. Material and methods

This retrospective study was performed from the patients' files between the years of 1990 and 2015 in a tertiary epilepsy center. All patients had been thoroughly investigated by detailed medical history and neurological examination. The EEG recordings of our patients were performed according to a protocol described previously and were evaluated by experienced clinical neurophysiologists [7].

2.1. Selection of cases and definitions

We only included those patients with a diagnosis of JME meeting the class II criteria defined in 2011 by an international workshop on JME [8]. Patients without any diagnostic EEG pattern were excluded from the study. Demographic data, including age, sex, age of seizure onset, seizure types, seizure precipitating factors, and duration of epilepsy;

Abbreviations: JME, juvenile myoclonic epilepsy; GTCS, generalized tonic–clonic seizures; SE, status epilepticus; MSE, myoclonic status epilepticus; NCSE, nonconvulsive status epilepticus; AED, antiepileptic drug; ASE, absence status epilepticus.

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Table 1
Characteristics of patients with status epilepticus.

	P1	P2	P3	P4	P5
Sex, current age (years)	M, 32	F, 34	M, 37	F, 36	F, 40
Seizure types/age at onset of seizures (years)	MYO/20	MYO/15	MYO, GTC/13	MYO, GTC/13	MYO, GTC/11
Age at first SE (years)	24	15	20	23	22
Localization of myoclonia	Face, head	Right arm	Left arm	Both arms	Generalized/both arms
Family history of epilepsy	+	+	+	−	−
Type of SE/number of SE episodes	Myoclonic SE/2	Nonconvulsive SE/1	Myoclonic SE/1	Myoclonic SE/2	Myoclonic SE, clonic–tonic–clonic SE/3 (2 MSE)
Ictal EEG available	Yes	No	No	Yes	Yes
Medication during SE	1. SE/CBZ 400 mg/day and VPA 500 mg/day 2. SE/VPA 500 mg/day	None	CBZ 400 mg/day	1. SE/none 2. SE/VPA 500 mg/day	1. SE/VPA 600 mg/day, barbexaclone 300 mg/day 2. SE/VPA 500 mg/day 3. SE/LEV 2000 mg/day
Precipitating factors in SE episodes	1. SE/during drug substitution from CBZ to VPA 2. SE/drug withdrawal	Sleep deprivation	Inappropriate treatment (CBZ)	1. SE/sleep deprivation 2. SE/drug withdrawal	1. SE/AED withdrawal 2. SE/AED withdrawal 3. SE/sleep deprivation
EEG	3.5- to 4-Hz GSW and PSW	4-Hz GSW and PSW right centrotemporal spikes	Asymmetric 3- to 3.5-Hz GSW, photosensitivity	4- to 6-Hz GSW and PSW increase with hyperventilation	3- to 3.5-Hz GSW and PSW generalized paroxysmal fast activity
Comorbid medical disease	Possible drug abuse	None	Alcoholism	None	None
Current medication	LEV 1500 mg/day	None	VPA 500 mg/day	VPA 250 mg/day	LEV 2000 mg/day
Prognosis	Benign	Benign	Benign	Benign	Benign

M: male, F: female, MYO: myoclonus, GTC: generalized tonic–clonic, SE: status epilepticus, CBZ: carbamazepine, LEV: levetiracetam, VPA: valproic acid, P: patient.

information about the disease course; coincidental disorders; and response to treatment were systematically collected from chart reviews. Patients with a follow-up period of at least 1 year after the first SE episode were included in the study.

Benign course was defined as having no GTCS and less than two myoclonic seizures monthly, and accordingly, drug-resistant course was defined as having one or more GTCS per year or more than one myoclonus monthly despite treatment with an appropriate AED with therapeutic drug levels for the last 5 years [9]. Associated psychiatric disorders and other psychosocial issues were also assessed.

2.2. SE classification

Myoclonic status epilepticus was diagnosed if myoclonia continued for at least 30 min [3] but without any interrupting non-ictal periods of >3 min. Nonconvulsive status epilepticus or absence SE (ASE) was diagnosed as a state of impaired consciousness without prominent motor symptoms and convulsions, which lasted at least 15 min according to new SE definitions [10].

Status epilepticus was diagnosed either with EEG monitoring or only with reliable and accurate witnessed history of SE.

2.3. Statistical analysis

Statistical Package for the Social Sciences (IBM SPSS Statistics, New York, USA) 20.0 software was used for statistical evaluations, and $p < 0.05$ was accepted as the limit of significance. The chi-square test was performed for categorical variables. For group comparisons, Mann–Whitney U Test and Fisher's exact tests were used where appropriate.

3. Results

We initially had identified 135 patients satisfying our selection criteria, but two patients with a final diagnosis of progressive myoclonic epilepsy during the follow-up were excluded from the study. Assessments were performed among 133 patients (80 women, mean age: 32.5 ± 9.2 years and 53 men, mean age: 32.2 ± 6.3 years). Mean age of epilepsy onset was 15 ± 4 years. Mean duration of follow-up was 8 ± 7.7 years. Myoclonic seizures were observed in all patients, GTCS occurred in 111 patients (83.4%), and absence seizures were observed in 49 patients (36.8%). Mean duration of epilepsy was 20 ± 5 years (1–51).

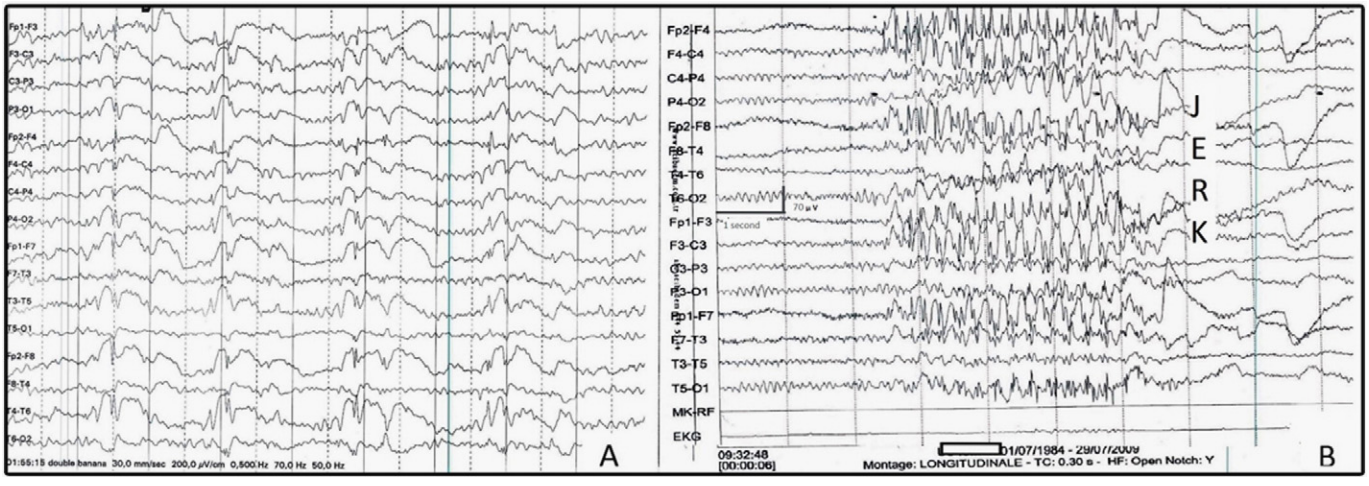

Fig. 1. Myoclonic status epilepticus of P1 (A) and P4 (B) documented by EEG showing generalized spike-and-wave discharges of different morphologies.