PERAMPANEL IN A CASE OF JEAVONS SYNDROME

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Introduction

Epilepsy with Eyelid Myoclonia, also called Jeavons Syndrome, is a rare form of epilepsy characterized by:
- eyelid myoclonia,
- eyelid closure-induced seizures or electro-encephalography paroxysms,
- photosensitivity [1].

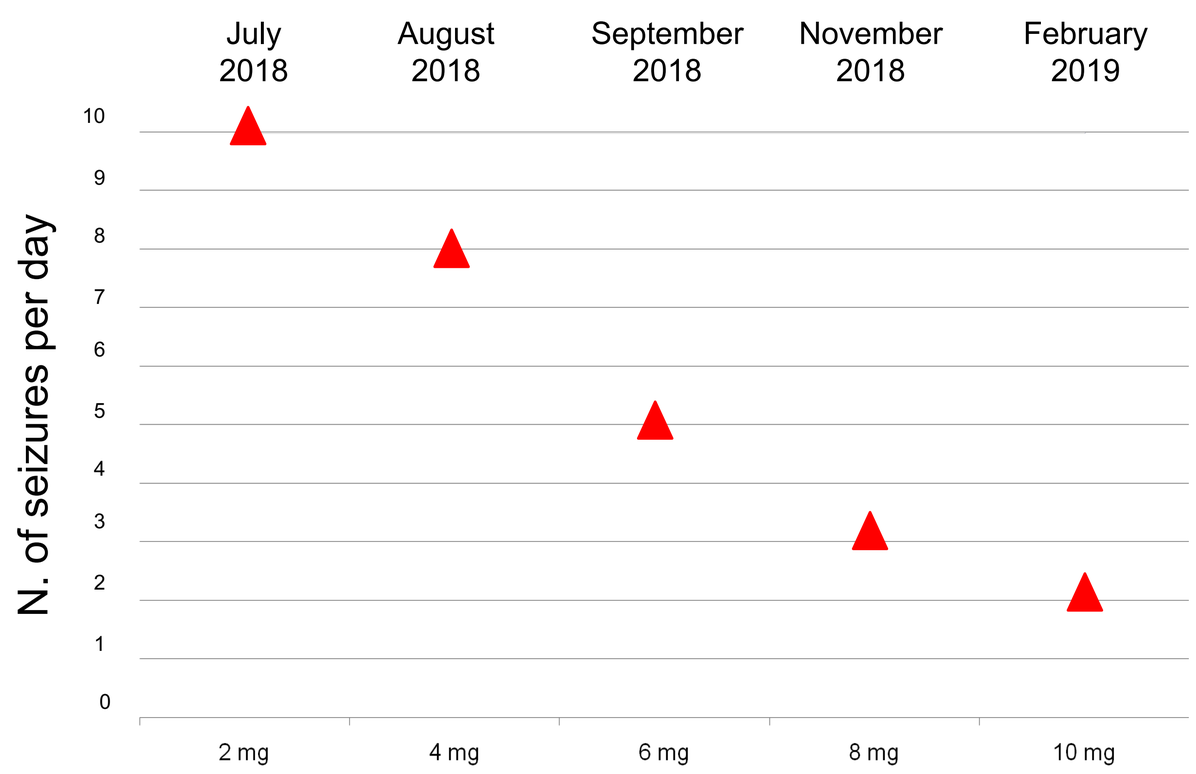
It may be associated with or followed by a brief loss of awareness (eyelid myoclonia with absences, EMA).
EMA is unlikely to be completely controlled with antiepileptic drugs (AED), being sodium valproate (VPA), lamotrigine and ethosuximide the most commonly prescribed drugs.
Perampanel (PER) is a non-competitive antagonist of AMPA receptors, approved as adjunctive therapy for partial-onset seizures with or without secondary generalization, and for generalized-onset seizures [2].

Case report

A 20-year-old Caucasian woman presented at the Epilepsy Clinic of “A. Cardarelli” Hospital with a history of brief absence associated with eyelid myoclonia from the age of 8 years, with a frequency of 10-15 episodes per day. At that age she was diagnosed with EMA and treated with Levetiracetam (LEV) 1000 mg daily, with no clinical benefit.
At the age of 10 years, LEV was changed to topiramate (TPM), but seizure frequency was unmodified.
At the age of 15 years, TPM was changed to VPA 1000 mg daily and, as the clinical response was poor, carbamazepine (CBZ) was added-on at the dose of 800 mg daily.
When the patient was seen for the first time at our Epilepsy Clinic in March 2018, she was taking VPA 1000 mg daily and CBZ 800 mg daily and seizure frequency was 10-12 episodes per day. As it is known that CBZ can make EMA worse, CBZ was reduced and finally stopped in July 2018.
In July 2018 PER was added with a starting dose of 2 mg and increased by 2 mg every 2-4 weeks up to 10 mg.
In September 2018 she was taking PER 6 mg daily and seizure-frequency was reduced of 50% (from 10 to 5 episodes per day).
In November 2018 she was taking PER 8 mg daily and there was a seizure-frequency reduction of 75% (3 episodes per day), with seizures mainly concentrated on awakening and falling asleep. PER was finally increased up to 10 mg daily, with no significant variation in seizure rate. Patient did not report any adverse reaction.

Conclusion

PER has good effectiveness in idiopathic generalized epilepsy [3]. EMA is a rare and drug-resistant syndrome. Here we documented a good efficacy to PER with a dose-dependent response in a case of EMA. PER can be considered a good option in this rare generalized epilepsy.



TIMING	AED
April 2006	Levetiracetam 1000 mg daily
March 2008	Topiramate 150 mg daily
March 2013	Sodium valproate 1000 mg daily
November 2013	Sodium valproate 1000 mg daily Carbamazepine 800 mg daily
March 2018	Sodium valproate 1000 mg daily Carbamazepine 800 mg daily
July 2018	Sodium valproate 1000 mg daily Perampanel 2 mg daily
August 2018	Sodium valproate 1000 mg daily Perampanel 4 mg daily
September 2018	Sodium valproate 1000 mg daily Perampanel 6 mg daily
November 2018	Sodium valproate 500 mg daily Perampanel 8 mg daily
January 2019	Sodium valproate 500 mg daily Perampanel 10 mg daily

References

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3. Villanueva V, Montoya J, Castillo A, et al. Perampanel in routine clinical use in idiopathic generalized epilepsy: The 12-month GENERAL study. Epilepsia. 2018;59(9):1740-52.

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