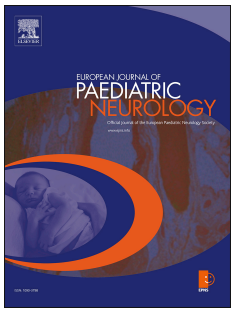
Accepted Manuscript



Febrile infection-related epilepsy syndrome (FIRES) with super-refractory status epilepticus revealing autoimmune encephalitis due to $GABA_AR$ antibodies

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PII: S1090-3798(17)31815-9

DOI: 10.1016/j.ejpn.2017.11.005

Reference: YEJPN 2328

To appear in: *European Journal of Paediatric Neurology*

Received Date: 28 July 2017

Revised Date: 27 October 2017

Accepted Date: 20 November 2017

Please cite this article as: Caputo D, Iorio R, Vigevano F, Fusco L, Febrile infection-related epilepsy syndrome (FIRES) with super-refractory status epilepticus revealing autoimmune encephalitis due to $GABA_AR$ antibodies, *European Journal of Paediatric Neurology* (2017), doi: 10.1016/j.ejpn.2017.11.005.

A CASE OF FEVER INFECTION RELATED EPILEPSY SYNDROME (FIRES) WITH SUPERREFRACTORY STATUS EPILETICUS REVEALING AUTOIMMUNE ENCEPHALITIS DUE TO ANTI-GABAA ANTIBODIES.

ABSTRACT

Background: Fever infection-related epilepsy syndrome (FIRES) has been described as an epileptic encephalopathy of unknown etiology affecting previously healthy children following febrile illness. Despite large investigations on autoimmune pathogenesis no membrane antibodies has been associated since now.

Case Study*:* We report a 13 years-old girl with negative history for neurological or autoimmune disease that developed at the sixth day of high fever a super-refractory status epilepticus. All investigations, including the most common antibodies related to immune-mediated encephalitis were negative. Seizures continued despite several therapeutic trials with anesthetics (midazolam, propofol) and antiepileptic agents as well as i.v. immunoglobulins but responded, at day 10 from the onset, to ketamine and high dose i.v. steroids. Due the high suspicion of autoimmune encephalitis we tested patient's CSF and plasma on mouse brain with positive response. We subsequently detected a high titre of $GABA_AR$ antibodies. After the resolution of the status epilepticus the patient achieved complete recovery of neurological functions.

Conclusion**:** this is the first reported case of a FIRES-like condition due to autoimmune encephalitis mediated by $GABA_AR$ antibodies. Our case suggests that $GABA_AR$ antibodies should be investigated FIRES.