

## **PS1.112**

### **FIRES... a rare epilepsy syndrome**

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**Background & Aim:** Febrile Infection-Related Epilepsy Syndrome (FIRES) describes an explosive-onset, potentially fatal acute epileptic encephalopathy that develops in previously healthy children and adolescents following the onset of a non-specific febrile illness. Overlapping signs and symptoms with other causes of encephalopathy and seizures make diagnosis and treatment challenging.

**Method:** We present a case of a 15 year old girl, previously healthy, with a typical disease course.

**Results:** After a febrile illness, the girl was found comatose with a tongue bite. She was intubated and ventilated, and started on anticonvulsives, antibiotics and antiviral medication. She initially improved and was extubated, but developed more generalised tonic clonic seizures, encephalopathy and in the course of a few days a convulsive state. Seizures were reduced temporarily but never completely inhibited, neither by barbituric coma nor by a ketogenic diet. Dysregulation of the autonomic nerve system including temperature and blood pressure instability were noted.

A full infectious, toxicological, metabolic (including muscle biopsy), autoimmune, cerebrospinal fluid findings and cranial magnetic resonance imaging did not reveal any etiology. Electroencephalography indicated epileptiform discharges in right temporal head region. More serial investigations were performed, and neurologists diagnosed a syndrome compatible with FIRES.

The girl went home subcomatose with a tracheostomy and ventilated, in the care of his parents and nursing service.

**Conclusions:** This case illustrates the diagnostic and therapeutic difficulties in Febrile Infection-Related Epilepsy Syndrome.

Patients with FIRES require immediate hospitalization. Antiepileptic drugs are given to treat seizures but are often ineffective. High-dose phenobarbital and clobazam are most likely to be effective. In severe cases, barbiturates are the only treatment for refractory status epilepticus, however, treatment by inducing a prolonged burst-suppression coma has been associated with a worse cognitive outcome. A ketogenic diet has been beneficial in some cases, especially if initiated early.

FIRES often has a poor prognosis but a few patients fully recover.