



### What is it?

Febrile infection-related epilepsy syndrome (FIRES) is a newly recognized epileptic encephalopathy. FIRES is a rare chronic epilepsy syndrome that suddenly develops after an acute febrile illness. A child will develop frequent and progressively debilitating seizures within a few weeks after a febrile illness such as those from a minor upper respiratory illness or a gastrointestinal bug. Seizures will begin and progress to a continuous state of status epilepticus that is resistant to conventional treatment; called refractory seizures or refractory epilepsy. Diagnosis is often difficult as it is based on clinical symptoms and only made after ruling out other conditions. Unfortunately, the outcome can be poor and surviving children can suffer significant cognitive delay and neurological impairment, along with refractory epilepsy.



### Common terms to know:

According to the *National Organization for Rare Disorder (2016)*, the below list are all synonyms/ names of FIRES that have appeared in literature:

- acute encephalopathy of obscure origin in infants and children
- acute encephalitis with refractory, repetitive partial seizures (AERRPS)
- acute encephalopathy with inflammation-mediated status epilepticus
- devastating epilepsy in school-aged children (DESC)
- idiopathic catastrophic epileptic encephalopathy
- idiopathic hemiconvulsion-hemiplegia and epilepsy syndrome (IHHE)
- fever-induced refractory epileptic encephalopathy in school-age children
- new-onset refractory status epilepticus (NORSE)
- severe refractory status epilepticus owing to presumed encephalitis

### What causes it?

The cause is unknown but researchers believe it may be linked to infection, genetic susceptibility, problems with metabolism or even an autoimmune disorder (GARD, 2017). Children ages 1-17 years are affected, but FIRES is most common in school-age boys.

### What are the signs and symptoms?

The acute phase of FIRES is characterized by sudden onset of nearly continuous seizures that do not respond to traditional treatment; known as refractory seizures or refractory epilepsy. The acute phase is variable and can last from a few days to several months. In the chronic phase, the number of seizures decreases but continue to be difficult to control. Memory and intellectual ability often suffer.

- Seizures can be focal or generalized onset
- Almost all children are diagnosed with chronic epilepsy
- An estimated 2/3 of children suffer mild to severe cognitive impairment
- A child could experience developmental regression
- FIRES can affect a child's intellectual abilities, memory, and motor skills
- Consciousness could be decreased
- A child could suffer migraines
- Temperature instability can occur

## **What is the treatment?**

The initial acute phase uses mainly supportive therapy to treat both fever and seizures. There is currently no specific treatment therapy for FIRES and there has been varying success with traditional anticonvulsants, immunoglobulin, and steroids. Treatment usually consists of benzodiazepines followed by anticonvulsants with a preference for IV medication form. Some studies show that treatment was more successful when the ketogenic diet was started in the acute phase but has yet to be confirmed.

### **SHNIC school nurses information:**

#### **Specific health issues for individual health care plans**

- FIRES diagnosis including onset
- Diagnosis including type of seizure, description of, typical length, characteristics, triggers, warning signs, how often seizures occur, and student's behavior following a seizure
- Note if seizures are controlled
- Current medication list for home and school
- Orders for emergency medications including when to administer, dose, route
- Seizure action plan including when to call 911
- Documentation or seizure log
- Orders for hidden device like a vagus nerve stimulator and how to use/manage
- Orders for special nutrition like ketogenic diet, if applicable
- Safety precautions for ambulating, transitioning in hallways, wearing a helmet, etc.
- Return to class protocol
- Rest period following seizure
- Education of staff for safe environment
- Prepare transportation/bus route accordingly

## Resources & Manuals

### **Genetic and Rare Disease Information Center (GARD)**

<https://rarediseases.info.nih.gov/diseases/11005/febrile-infection-related-epilepsy-syndrome>

### **National Organization for Rare Disorders (NORD)**

<https://rarediseases.org/rare-diseases/febrile-infection-related-epilepsy-syndrome-fires/>