

# FEBRILE INFECTION-RELATED EPILEPSY SYNDROME (FIRES)

## What is it?

Febrile infection-related epilepsy syndrome (FIRES) is a rare, sudden-onset chronic epilepsy syndrome. The progressively debilitating seizures and irreversible neurological damage develop between 24 hours to 2 weeks after an acute febrile infection like an upper respiratory illness or a gastrointestinal bug. Seizures will begin and progress to a continuous state of status epilepticus (SE) resistant to conventional treatment; called refractory seizures or refractory epilepsy. Unfortunately, the outcome can be poor and surviving children can suffer neurological impairment and significant cognitive delay.

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

FIRES is classified as a subcategory of new-onset refractory status epilepticus (NORSE). NORSE is a condition, not a diagnosis, with new onset refractory SE without a clear acute precursor. Several epilepsy syndromes in infants and children share many similarities with NORSE and FIRES. The related disorders include:

- Acute encephalopathy of obscure origin in infants and children
- Acute encephalopathy with inflammation-mediated status epilepticus
- Idiopathic catastrophic epileptic encephalopathy
- Fever-induced refractory epileptic encephalopathy in school-age children
- Severe refractory status epilepticus owing to presumed encephalitis
- Devastating epilepsy in school-aged children (DESC)
- Acute encephalitis with refractory, repetitive partial seizures (AERRPS)
- Presumed encephalitis with refractory status epilepticus

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## 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. 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

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## 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 and inconsistent success with traditional anticonvulsants, immunoglobulin, and steroids. Treatment usually consists of benzodiazepines followed by anticonvulsants with a preference for IV medication form.



Kennedy Krieger Institute

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

## Suggested school accommodations

Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

<b>Communication</b>	Hearing and vision impairments; consider size and font; incorporate their use of gestures, signs, symbols, drawings; assistive technology; repeat signs often in natural communication; encourage sharing for peer interactions; keep daily schedule; slow down, repeat; extra adult support as needed
<b>Teaching</b>	Organizational strategies and aids, communicate clear expectations, use motivating materials, offer choices, incorporate active and passive learning, maintain predictable schedules and routines, pause time for response
<b>Environment</b>	Need sensory breaks, flexible schedule, offer a rest area, know behavior “signs” when break is needed,
<b>Training</b>	Peer support, prepare transportation/bus route accordingly, staff education/training, Emergency Evacuation Plan (EEP)

## Specific health issues for Individualized Healthcare Plan

- 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
- Current medication list including PRN medications
- 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.
- Education of staff for safe environment
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

## Resources & Manuals

### National Institute of Health: Genetic and Rare Diseases Information Center

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

### National Organization of Rare Disorders: New-Onset Refractory Status Epilepticus (NORSE) and Febrile Infection-Related Epilepsy Syndrome (FIRES)

<https://rarediseases.org/rare-diseases/new-onset-refractory-status-epilepticus-norse/>

### Mayo Clinic Children’s Center– What Can We do to Extinguish FIRES? Febrile Infection-Related Epilepsy Syndrome

<https://ce.mayo.edu/sites/ce.mayo.edu/files/FIRESFINALslides03142017.pdf>

Fox, K., Wells, M.E., Tennison, M., Vaughn, B. (2017). Febrile-infection related epilepsy syndrome (FIRES): A literature review and case study. *Neurodiagn J*, 57(3):224-233. doi: 10.1080/21646821.2017.1355181.