

# The NORSE/FIRES Family Registry

Participation in the international NORSE/FIRES Family Registry will help doctors better understand this rare syndrome. Learning who becomes ill, the short and long-term outcomes and the quality of life of survivors can lead to more effective treatments.

Families, patients, and physicians can enter patient data securely online. The experience of every patient counts.

Learn more and gain entry to the Registry at [norseinstitute.org](http://norseinstitute.org) or scan:



**Be part of the answer to  
NORSE**



## **YOU ARE NOT ALONE**

For resources to help understand and manage the NORSE/FIRES experience, go to: [norseinstitute.org](http://norseinstitute.org)

Connect with other NORSE families to learn and share your experience.

**NORSE**  
INSTITUTE

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**NORSE**  
New Onset Refractory  
Status Epilepticus

**FIRES**  
Febrile Infection-  
Related Epilepsy  
Syndrome

FAQ and  
Resources



## What is NORSE ?

NORSE stands for New Onset Refractory Status Epilepticus, a term that describes a condition of prolonged seizures (status epilepticus) that erupt suddenly (new onset) with no apparent cause (no trauma, stroke, toxin, etc) that persist despite administration of at least two standard anti-seizure drugs (refractory).

## What is FIRES and how does it differ from NORSE?

FIRES refers to Febrile Infection-Related Epilepsy Syndrome, a sub-type of NORSE, the distinguishing characteristic being fever between 2 weeks and 24 hours prior to the onset of refractory status epilepticus.

Flu-like symptoms precede the onset of prolonged seizures in two-thirds of NORSE cases.

## What causes NORSE?

Possible causes may ultimately be autoimmune or paraneoplastic encephalitis (an autoimmune reaction caused by cancers) but in over half the cases, no cause is found despite extensive testing. Cases with no identifiable cause are called "cryptogenic NORSE."



## Who gets NORSE?

NORSE and FIRES can affect all ages but is found most often in healthy young adults and children with no history of epilepsy.

Estimating the occurrence is difficult because the medical terms for NORSE and FIRES are relatively new, even though the syndrome is not. Cases may have gone unrecognized, unrecorded or the terms not communicated to families.

## What will happen?

The disease course and patient outcomes are often unpredictable. In the acute phase, which can last from days to several months, the patient remains comatose due to the effect of the seizures and anesthetic treatment. Patients can develop complications associated with prolonged unconsciousness and mechanical ventilation. The mortality rate can reach 30% and is higher in adults than children. Although a significant proportion of cases result in mortality, long-term disability and epilepsy, some patients can regain their former level of functioning.

Due to the unpredictable course of the syndrome, ongoing communication with the patient's medical teams is vital.

## What is the best treatment?

When an underlying cause for the seizures is found, specific therapies can be applied. There is no established treatment protocol for cryptogenic NORSE (cases where no cause is identified). As inflammation is suspected to play a role in these cases, possible treatment options include IV steroids, IV immunoglobulins, plasma exchange therapy (plasmapheresis) and some monoclonal antibodies against inflammatory cells such as rituximab, anakinra, and tocilizumab. Other possible treatments have included cannabinoids and the ketogenic diet.

### Professionals and families can find resources at the NORSE Institute:

- Diagnostic Evaluation Guideline
- Curated reading list
- Grant opportunities
- NORSE Research Bulletins
- Glossary of Terms for Families
- Discharge Planning Guide
- NORSE/FIRES Biorepository

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