

Rasmussen syndrome causes partial seizures, mental deterioration, and loss of movement skills and speech. It is a rare condition with severe and disabling effects.

What are other terms for Rasmussen syndrome?

Other terms for Rasmussen syndrome that you may come across include:

- chronic focal encephalitis of Rasmussen
- chronic progressive epilepsy partialis continua of childhood
- Rasmussen encephalitis
- type 1 epilepsy partialis continua

What causes Rasmussen syndrome?

Rasmussen syndrome is a type of chronic encephalitis (brain inflammation). It usually affects a single hemisphere of the brain. It is progressive, meaning that it gets worse over time.

It is not clear what causes the encephalitis. It may begin when the brain is infected with a virus, such as cytomegalovirus (CMV) or herpes virus. However, there is no evidence for viral infection in many cases, and it is possible that there are several different possible triggers. An autoimmune reaction, in which the body's immune system attacks nerve cells and causes the inflammation to spread, is suspected.

The disease does not have a seasonal pattern and does not seem to cluster in places or run in families. It is not infectious; you cannot catch it from another person.

What are the features of Rasmussen syndrome?

Rasmussen syndrome usually begins when the child is between one and 15 years old, most often when the child is between six and 10 years old. A few cases have been seen in older teenagers and young adults.

The child's first seizure is usually a partial motor, partial sensory, or tonic-clonic seizure. In about 20% of children with this syndrome, the first seizure turns into status epilepticus. The seizures are frequent and may be prolonged, and often do not respond to anti-epileptic drugs.

In most cases the child will have epilepsy partialis continua. This is a form of partial status epilepticus. The child has repeated motor seizures. The seizures happen in the same region and look the same each time.

The child slowly loses movement skills on the same side of the body as the partial seizures. She starts to develop hemiparesis (weakness or partial paralysis on the affected side of the body), usually within a year after the start of seizures, although it can be up to 10 years later in older children and adults. She may also develop problems with her sight on the affected side. Up to 85% of children start to lose mental abilities, intelligence, and speech. On imaging studies, the affected side of the brain atrophies (shrinks). The child's EEG becomes very abnormal.

After two to 10 years, the child's neurological deterioration stops and the child has fewer seizures.

In one study, researchers found that younger children (six years old or younger) developed a more severe form of Rasmussen syndrome more quickly. Older children and adults were slower to develop hemiparesis and brain atrophy. However, this was a very small study and the results are not necessarily true for all people with this syndrome.

How many other children have Rasmussen syndrome?

Rasmussen syndrome is rare. It causes 1% or fewer of all cases of epilepsy.

How do you know that a child has Rasmussen syndrome?

Rasmussen syndrome is difficult to diagnose in the early stages. Once the child develops epilepsy partialis continua, hemiparesis, and mental impairment, the diagnosis is easier to make. MRI studies of the brain, showing atrophy of one hemisphere, can be suggestive of the diagnosis. Imaging studies also help to rule out other possible causes for these symptoms, such as a tumour or other structural problem with the brain. Brain biopsy may be required to make the diagnosis.

How is Rasmussen syndrome treated?

Anti-epileptic drugs usually do not work with Rasmussen syndrome.

The surgical treatment of this rare condition needs to be individually tailored to each child. At the moment, the most effective treatment is

surgery to remove all or part of the affected hemisphere of the brain (hemispherectomy). There are no controlled studies of this procedure, but in most cases it helps to reduce the number of seizures and can prevent or even reverse mental deterioration. It is usually not considered until later stages of the disease, when the child has lost quite a lot of movement on the affected side of her body.

Hemispherectomy is a major surgical procedure that usually results in weakness on one side of the body. Because the child's brain is still developing, the remaining hemisphere may be able to take on many of the tasks that are normally performed by the affected hemisphere. Before and after this surgery is done, the child will need detailed assessments by occupational and physical therapists and neuropsychological assessments to determine the effects of the disease on the body.

A number of medical treatments may also be tried, although in most cases they only suppress the seizures for a short time and there are few studies supporting their use. They include:

- treatment to suppress the immune system
- plasmapheresis, in which the child's blood plasma is removed and replaced with plasma from a donor. This technique may improve the seizures dramatically, at least for a short time, but it can be difficult to perform in children
- high doses of steroids in the early stages of the disease
- ketogenic diet

- intravenous immune globulin (IVIG) to boost the child's immune system
- antiviral drugs

More information

- [Ketogenic Diet for Epilepsy](#)
- [Surgical Treatment of Epilepsy](#)

What is the outlook for a child with Rasmussen syndrome?

Rasmussen syndrome is a progressive disorder that results in weakness or paralysis of one side of the body and cognitive impairment. Treatment may help to halt the progression of the disease.

In some children, surgery reduces the number of seizures. Most people with this syndrome, though, have at least some motor and cognitive problems.

