

Rasmussen's encephalitis (RE)

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What is RE?

Rasmussen encephalitis (RE), also called Rasmussen syndrome, is a rare, progressive, chronic encephalitis (inflammation of the brain) affecting one hemisphere (one side) of the brain. It occurs mainly in children (most cases are seen in six to seven-years-old children). However, around 10% of all cases are adolescents and adults. RE occurs usually in healthy individuals. It is estimated that no more than two new cases per year are identified in large epilepsy centres.

Symptoms of RE

RE is characterised mainly by intractable seizures, progressive hemiparesis, and cognitive loss (learning difficulties).

- Seizures

Seizures may have different forms and characteristics: simple partial seizures, complex partial seizures, generalised tonic-clonic seizures or status epilepticus. But the most notable seizure manifestation of RE is *epilepsia partialis continua* (EPC) which is continuous twitching of the face, arm or leg on one side of the body. About 50% of patients with RE have EPC.

- Hemiparesis

Hemiparesis is a weakness on one side of the body. The wiring of the nervous system determines that a lesion on one side of the brain causes problems on the opposite side of the body. Thus, involvement of one hemisphere causes weakness on the other side of the body.

- Cognitive impairment and other characteristics

Other symptoms depend on which hemisphere is affected. Over time, patients may develop cognitive impairment, aphasia (difficulties with using language correctly), hemianopia (loss of vision in either the right or left sides of both eyes), sensory deficits, dysarthria (difficulty speaking), dysphagia (swallowing difficulties) and psychiatric problems. Language and cognition disturbances are almost always seen in patients with left-side involvement.

The cause of RE

The exact cause of RE is not known. However, there is increasing evidence of an underlying immune disorder. In a majority of cases only one hemisphere is affected (cases when both hemispheres are affected are very rare) and the disease starts focally (one specific area) and spreads across the hemisphere.

Evolution and prognosis

Most children are healthy before the onset of Rasmussen's encephalitis. The progression of the symptoms to significant neurological impairment usually occurs within months to a few years. Some patients have a fast evolution with the rapid development of hemiparesis, while others have a slow evolution with hemiparesis developing more than one year after seizure onset. There are patients who develop only a mild hemiparesis or language disturbance.

In the long-term, the disease is usually expected to 'burn itself out' but not before the patient is left with significant hemiparesis, visual field loss, learning difficulties and, usually, on-going epilepsy. Unfortunately, either quickly or slowly, children begin to show difficulties with learning. At first, their academic performance may stabilise and then they fall behind their peers. Supporting their education is extremely important. The syndrome can spontaneously stabilize at any time. Very rarely, in rapidly progressive cases, RE can lead to death.

Diagnosis

Diagnosis is made based on clinical features (symptoms) and results of radiological investigations. The most useful investigations are:

- Electroencephalography (EEG) which may reveal brainwave patterns characteristic of certain types of epilepsy.
- Serial Magnetic Resonance Imaging (MRI) which shows progressive atrophy (shrinkage) and scarring of the affected side of the brain.

Brain biopsies are not usually needed to make the diagnosis and can often be inconclusive.

Treatment

There are two main forms of treatment:

- an anticonvulsive treatment, with as few side-effects as possible.
- a treatment that stops the destruction of the brain cells and improves the long-term neurological and neuropsychological functional outcomes.

Anticonvulsant treatment (AED)

EPC is usually unresponsive to anticonvulsive treatment. However, the frequency of focal and secondary generalised tonic-clonic seizures in RE may be reduced by anticonvulsive medication. This is the aim of drug treatment, rather than trying to achieve complete seizure resolution.

Immunomodulatory therapy

These treatments target the autoimmune system and include steroids, tacrolimus, azathioprine and intravenous immunoglobulin (IVIg). More recently, treatments with other drugs (monoclonal antibody therapies) such as natalizumab and rituximab are being trialled. These treatments slow down the illness, but they do not stop the disease. Steroids and IVIg may also reduce seizures.

Hemispherectomy (HE)

Surgery, such as HE, remains the only cure for the seizures caused by RE. HE (disconnection of the affected side of the brain from the healthy brain) in one of its modern variants offers a very high chance of seizure resolution. However, HE can have significant consequences:

- irreversible loss of functions located in the affected hemisphere.
- hemiplegia (if there isn't one already from the disease); the patients are expected to walk again but they are not expected to have fine finger function.
- hemianopia (loss of vision for objects coming from one side).
- speech loss if the surgery is on the side of the brain generating language (language dominant side).
- swallowing difficulties.

Despite these concerns, HE has less side-effects in children than in adults. Rehabilitation is very important and should begin early in the postoperative period. The decision to go ahead with surgery and choosing the best time is difficult and individual to each child. Careful decisions should be made by the parents and young person, in conjunction with a specialist epilepsy surgery service. Medical therapy prior to HE may be considered in two scenarios:

- in patients with minimal or no motor deficits.
- in patients with RE involving the dominant hemisphere or bilateral hemisphere.

Further Information on RE

- NORD (The National Organisation for Rare Diseases) (<https://rarediseases.org/rare-diseases/rasmussen-encephalitis/>) provides information, advocacy, research, and patient services to help all patients and families affected by rare diseases.
- The Hemispherectomy Foundation (<http://hemifoundation.homestead.com/welcome.html>) provides emotional, financial, and educational support to individuals and their families who have undergone, or will undergo, hemispherectomy or a similar brain surgery.

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Thank you!

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