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-year-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. Seizures may progress fast within weeks-months, while in some cases of RE seizures may evolve gradually over years.

- Hemiparesis

Hemiparesis is a weakness on one side of the body. Due to how the nervous system is organized, 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 side of the brain is affected. Over time, patients may develop cognitive impairment, aphasia (difficulties with producing or understanding language), hemianopia (loss of vision in either the right or left sides of both eyes), sensory deficits, dysarthria (difficulty speaking), dysphagia (swallowing difficulties) and psychiatric symptoms. Patients with either left or right-sided disease can have language difficulties, but these difficulties are typically more pronounced in those with left-sided RE.

The cause of RE

The exact cause of RE is not known. However, there is increasing evidence of an underlying immune disorder. In most 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:

1. **Anticonvulsant treatment (AED)**

   EPC often does not respond well to anticonvulsant treatment. However, in RE, these medicines can help reduce how often focal and secondary generalised tonic-clonic seizures happen. The aim of treatment is to make seizures less frequent and easier to manage, rather than to stop them completely, using medication that causes as few side-effects as possible

2. **Immunomodulatory therapy**

   These treatments target the autoimmune system preventing further damage to brain cells and improve the long-term neurological and neuropsychological functional outcomes. Treatments 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 fewer side-effects in children than in adults. Rehabilitation is very important and should begin early in the postoperative period. The decision to proceed with surgery—and choosing the right time to do so—is complex and unique to each child. These choices should be made carefully by the parents and the young person, working closely with a specialist epilepsy surgery team to ensure the best possible outcome.

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

- The consequences of RE can affect the whole family and coping with these difficulties can be distressing and challenging for everyone involved. Counselling can help by providing a safe space to talk openly with someone trained to listen and support you. Contact your primary care doctor to find local counselling and support services.
- 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.

Ongoing Research on RE

- The REMEDI Study (Rasmussen's Encephalitis: Mechanisms & Diagnostics for Early Intervention): Funded by Action Medical Research and the British Paediatric Neurology Foundation, a group of clinicians and researchers at UCL Great Ormond Street Institute of Child Health are using cutting edge techniques to understand what causes the disease and find early warning signs.
- The SOLAR Study (Surgical Outcomes, Language and Atrophy in Rasmussen syndrome): Funded by the Child Health Research Charitable Incorporated Organisation, a group of researchers and clinicians at UCL Great Ormond Street Institute of Child Health are characterising disease progression, cognition and how the brain responds to disease in Rasmussen syndrome and are aiming to understand what promotes language recovery after hemispherotomy.

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