

## Clinician's Corner

### What are Epilepsy Syndromes?

Classifying epilepsy by seizure type alone leaves out other important information about the patient and the episodes themselves. Classifying into syndromes takes a number of characteristics into account, including the type of seizure; typical EEG recordings; clinical features such as behavior during the seizure; the expected course of the disorder; precipitating features; expected response to treatment, and genetic factors.

Epileptic syndromes can be either idiopathic (of unknown cause) or symptomatic of underlying brain damage or disease. In general, idiopathic forms have a better prognosis in terms of both seizure control and eventual remission than do symptomatic forms

### Lennox-Gastaut Syndrome

Lennox-Gastaut syndrome (also known as myoclonic-astatic epilepsy) is a combination of seizures usually including atypical absense seizures (starting with automatic behavior without conscious control); tonic seizures (stiffening) atonic or astatic seizures (drop attacks); mental retardation; a distinct slow spike-and-wave EEG; and onset between 1 and 5 years of age.

Some children are developmentally normal when the syndrome begins, but then lose skills, sometimes dramatically, in association with uncontrolled seizures. By age 6 most children with Lennox-Gastaut have some degree of mental retardation.

Children with Lennox-Gastaut syndrome typically have more than one type of seizure. The atonic-astatic (drop attack) seizures are most troubling because of the injuries caused by repeated falls. Many children wear protective helmets. The tonic (stiffening) seizures are most common during sleep, including nap time, whereas generalized tonic-clonic seizures (convulsions) occur most often on awakening.

Some of these children are prone to develop non-convulsive status epilepticus (a continuous seizure state that is associated with a change in the child's level of awareness. This requires medical intervention to bring it to an end).

As children with Lennox-Gastaut syndrome grow older, the types of seizures change. In most cases, the drop seizures abate. They are replaced by partial, complex partial, and secondarily generalized convulsions. Among teenagers with Lennox-Gastaut, complex partial seizures are the most common form.

This seizure syndrome is difficult to treat and often does not respond to the usual seizure medications. Other treatment choices may include the [ketogenic diet](#), [vagus nerve stimulation](#) therapy or occasionally corpus callosotomy surgery.

### Benign Rolandic:

Epilepsy Benign Rolandic epilepsy (also known as benign partial epilepsy of childhood) accounts for more than one-third of all cases of epilepsy that begin in middle childhood,

accounting for 16 percent of those beginning before age 15. There is a family history in 18 percent of cases and the condition is probably genetically determined.

Rolandic epilepsy is the most common type of benign partial epilepsy. Seizures start as simple partial, usually beginning in the face. There may be drooling and temporary inability to speak, although consciousness is preserved. The seizures then generalize to tonic-clonic convulsions.

Most of the seizures are nocturnal and occur during sleep. Neurological and other functioning is usually normal, while the EEG shows a dramatic focal spike most often in the centrotemporal regions of the brain. Most children are seizure free five years after onset; by age 14, 95 percent will have undergone permanent remission.

### **Rasmussen's Syndrome**

Rasmussen's syndrome, also known as Rasmussen's encephalitis, begins in childhood and produces a slow deterioration of one whole side (hemisphere) of the brain with loss of function on the opposite side of the body. An autoimmune response to a viral infection has been suggested as a possible cause. Various types of treatment have been tried, including surgical removal of the affected side of the brain. In children, the remaining hemisphere may compensate for functions lost, but weakness on the affected side will remain.

The condition typically starts with seizures, with weakness appearing later in the course of the disorder. Simple partial seizures affecting movement are the most common form.

**Additional epilepsy syndromes include:** Childhood Absence Epilepsy, Febrile Seizures, Frontal Lobe Epilepsy, Infantile Spasms, Juvenile Myoclonic Epilepsy, Landau-Kleffner Syndrome, Progressive Myoclonic Epilepsy, Reflex Epilepsy, Temporal Lobe Epilepsy.

For more information about epilepsy syndromes or any issues regarding epilepsy, please contact the Epilepsy Foundation of Minnesota Information & Referral Services at 1-800-779-0777 ext. 2303.

Source: Epilepsy Foundation  
<http://www.epilepsyfoundation.org/answerplace/Medical/seizures/syndromes>