

GUIDE TO LENNOX-GASTAUT SYNDROME

What is Lennox-Gastaut Syndrome?

Childhood epileptic encephalopathy (Lennox-Gastaut syndrome [LGS]) is a rare and debilitating form of childhood-onset epilepsy. The syndrome is characterized by frequent seizures and multiple seizure types, a resistance to medications or therapies, cognitive dysfunction, regression, and an abnormal EEG with generalized slow spike-and-wave discharges. LGS constitutes between 1-4% of pediatric epilepsies and typically appears between the second and sixth year of life¹. In 30-35 percent of cases, no cause can be found.²

Most children are typically developing normal when first diagnosed, but then begin to lose skills, sometimes dramatically in association with uncontrolled seizures. As children with Lennox-Gastaut syndrome grow older, the types of seizures change. In most cases, the drop-seizures (atonic) are replaced by partial, complex partial, and secondarily generalized convulsions. The uncontrolled seizures may become less of an issue as children grow, but impaired intellectual functioning and behavioral problems persist.

Complete recovery, with freedom of seizures and normal development, is typically unusual for a child with LGS.

How can LGS affect a child's progress in school?

Children with LGS often have a difficult time in school settings. They may often seem unresponsive or vacant due to the electrical disturbances in their brain and can also exhibit extreme behavior issues in school and at home.

Behavior issues, personality disturbances, mood instability, and slowing of psychomotor development are all consistent with LGS. Behavioral disturbances can include poor social skills and attention seeking behavior³ which can be caused by the effects of the medication, difficulty interpreting information, or electric disturbances in the brain.

Older children with LGS can experience acute psychotic episodes, forms of psychosis with aggressiveness, character problems, and irritability. Cognitive problems include slowed reaction time and information processing. The main characteristics of mental deterioration are reported as apathy, memory disorders, impaired visuomotor speed, and perseverance. School-age children are usually placed into a special education classroom or will receive home tutoring.

Absence seizures are very common in children with LGS and can cause an interruption to consciousness including staring spells. These seizures can interrupt the child's learning and often go undetected by teachers and faculty due to their brief length. Absence seizures can also cause a child to seem unresponsive and vacant.

As a teacher, it's important to understand the severity of LGS and how it affects a child's performance in school. Children with Lennox-Gastaut syndrome can have hundreds of seizures a day with little to no control. Extreme behavior, aggression, and acting out are usually side effects of the seizures and medication. Typically, a child cannot control these behaviors.

¹ Glauser, TA. Lennox Gastaut Syndrome. www.emedicine.com

² National Institute of Health. Lennox-Gastaut Syndrome.

³ http://www.cnsfoundation.org/site/PageServer?pagename=rch_ListofDisordersMain

What are the types of seizures a child with LGS can have?

Generalized Seizures:

Generalized seizures affect both sides of the brain and result in a loss of consciousness.

Absence (Petit Mal): Absence seizures include an interruption to consciousness where the person experiencing the seizure seems to become vacant and unresponsive for a short period of time (usually up to 30 seconds). Slight muscle twitching may occur. Absence seizures include loss of awareness, interruption to consciousness (a person can seem unresponsive for a short period of time), and staring spells. The duration of absence seizures are usually brief, and are sometimes not detected. They are most common in children and can affect the child's learning.

Tonic Clonic (Grand Mal): Tonic-clonic seizures involve an initial stiffening of the muscles (tonic phase) followed by rhythmic jerking (clonic phase). Breathing may decrease or cease during the tonic phase and then resume, sometimes irregularly, during the clonic phase. Incontinence and tongue biting can occur. This seizure type is most often affiliated with epilepsy.

Atonic (Drop Attack): Atonic seizures involve the loss of muscle tone, causing the person to fall to the ground. Most children who have this type of seizure are required to wear a helmet.

Myoclonic: Involve a brief, rapid contraction of muscles and can result in sudden jerks of the muscles.

Clonic Seizures: Clonic seizures are myoclonus which repeat at a rate of 2-3 per second.

Status epilepticus is a continuous seizure state with no recovery between seizures. If a tonic-clonic seizure lasts longer than 5 minutes, it is a life-threatening condition and emergency care should be sought immediately.

Partial Seizures:

Partial seizures cause no interruption of consciousness and may cause a sensory experience (perception of smells, sounds, voices, etc). Partial seizures are limited to a specific area of the brain.

Simple- Partial: Consciousness is retained during this seizure type and patients usually remain awake and can recall what happened. Emotions can be affected.

Complex- Partial: Consciousness may be lost during a complex-partial seizure and often take place in the temporal lobes. A complex partial seizure may involve the unconscious repetition of simple actions or a blank stare. Unawareness of the occurrence and no memory of the seizure is also common.