Lennox-Gastaut Syndrome

What is Lennox-Gastaut Syndrome?

Lennox-Gastaut Syndrome is a rare epilepsy, characterized by the presence of multiple types of intractable seizures (in particular tonic seizures in sleep, but atonic and atypical absence seizures also occur). Children with Lennox-Gastaut Syndrome have cognitive and behavioral abnormalities and diffuse slow spike-and-wave and paroxysms of fast activity on EEG. The diagnosis therefore is made based on the clinical patterns of seizures, development and learning profile and using information from EEGs (especially EEGs done during sleep).

Lennox-Gastaut syndrome has sometimes been used as a label to describe other epilepsies with difficult to treat seizures, however it is important not to do this as other epilepsies are important to recognize in their own right as they may have specific treatment implications and genetic causes. For example, children with Dravet syndrome were previously labelled as having Lennox-Gastaut syndrome until the specific clinical features and genetics of Dravet syndrome were recognized and this syndrome became its own unique entity.

Epilepsy with myoclonic-atonic seizures (Doose syndrome) and atypical epilepsy of childhood with centrotemporal spikes (pseudo-Lennox syndrome) are other epilepsies that should be distinguished from Lennox-Gastaut syndrome because of different treatments or outcomes – children with atypical epilepsy of childhood with centrotemporal spikes might have a period of intractable seizures but seizures stop of their own accord and development and learning are usually not affected.

What types of seizures are seen?

Children with Lennox-Gastaut syndrome begin having seizures between the ages one and seven years of age. Typically multiple types of generalized seizures are seen including atypical absences, tonic and atonic seizures. Seizures are often frequent (multiple every day) and atonic seizures can result in falls and injuries (often children need to wear a helmet for safety). Sometimes seizures can be frequent but may not be obvious (called non-convulsive status) and all that is seen is a child becomes slow and less responsive, sometimes described as ‘like a zombie’. Seizures can be aggravated by sleep deprivation.

Medications typically fail to control seizures fully. Medications such as valproate and lamotrigine (in combination), topiramate, benzodiazepines and levetiracetam are tried in the first instance. Specific medications might be trialled for drop seizures (rufinamide). Benzodiazepines and/or steroids may be trialled for periods of non-convulsive status. Most children are considered for advanced epilepsy management therapies such as the modified Atkins or ketogenic diet or vagal nerve stimulator therapy. Occasionally brain surgery (splitting the fibers that connect both hemispheres of the brain, called a corpus callosotomy) may be considered to treat severe atonic seizures causing falls and injury.

If a child with Lennox-Gastaut syndrome presents with frequent seizures and benzodiazepines (rescue medication such as midazolam) are administered, occasionally this can result in ‘tonic status’ – emergence of frequent tonic seizures. Doctors should be alert to this and if this occurs, they should
change to using other medications to stop the status. Seizures can be aggravated by sleep deprivation and during sleep.

**What effects are there on learning?**
Most children with Lennox Gastaut syndrome experience delays (or regression) in development and learning. This may not be seen in the first years of life but may become apparent after onset of seizures. Early assessment and the support of early intervention services and therapy are important in a child with Lennox Gastaut syndrome because of the high risk of developmental or intellectual difficulties. Most children with Lennox Gastaut syndrome will need a modified curriculum and an individual education plan (IEP).

**What effects are there on behavior?**
Children with Lennox Gastaut syndrome can have behavioral difficulties. Lennox Gastaut syndrome, and the medications used to treat seizures, can be associated with problems with attention and concentration, with processing information and with understanding information. These factors can result in a child who appears not to comply with instruction. Having a good understanding of a child’s developmental and intellectual abilities is therefore important in understanding and managing any apparent behavior. In some situations specific medications can assist with improving behavior, such as the use of medications to improve attention and concentration or to improve sleep.

**What causes Lennox Gastaut syndrome?**
The cause of is not yet known. Genetic causes are suspected, these are likely to be new genetic changes in the child with Lennox Gastaut syndrome (and therefore not inherited from a parent). Further research is underway at this time to identify the possible genetic causes of Lennox Gastaut syndrome.

**How common is it?**
This is a rare childhood epilepsy, it is important that other epilepsy syndromes are carefully excluded before a diagnosis of Lennox Gastaut syndrome is made. It is also important that treatable conditions such as metabolic disorders (that require specific treatment) are excluded. Examples of metabolic disorders that should be excluded include pyridoxine disorders (that may require treatment with pyridoxine or pyridoxal-5-phosphate) or glucose transport disorders (that require treatment with the modified atkins or ketogenic diet).

**What is the long-term outlook?**
As children grow older, the types of seizures change. In most cases, atonic seizures cease with maturation. Usually these are replaced by focal seizures and secondarily generalised seizures. Focal seizures are the most common seizure type among teens who have Lennox-Gastaut syndrome. Adults with Lennox-Gastaut syndrome typically require high levels of support.

Information reviewed by Dr. Kate Riney, Mater Hospital Brisbane.

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