Lennox-Gastaut Syndrome (LGS) is a rare epilepsy syndrome. Nobody is born with LGS. It may develop over time from childhood seizures that remain uncontrolled by treatments.

Children and Adults with LGS share similar features:
- Seizures that start in childhood
- More than one seizure type
- Slow spike-and-wave on EEG
- Developmental delay/cognitive impairment (70% have this at diagnosis)

Any seizure type can be seen in LGS. The most common seizure types are:
- Tonic
- Atonic Drop Seizures
- Generalized Tonic-Clonic
- Atypical Absence
- Non-convulsive status epilepticus
- Myoclonic

While developmental delay/cognitive impairment occurs in most with LGS, it is not always present at the start of LGS and is not required for the diagnosis to be made.

What causes seizures in LGS?

- Some genes and genomic regions associated with LGS:
  - ALG13
  - ARX
  - CACNA2D2
  - CLN1/2/5
  - CDKL5
  - DNA1
  - DOCK7
  - FLNA
  - FON1/Dup
  - GABA1
  - GABRB3
  - GLIO
  - GNAQ1
  - GRIN1
  - GRIN2A
  - SCN1A
  - SCN2A

- Trauma before or during birth
- Abnormal brain formation
- Infections
- Genetic factors
- Metabolic disorders
- Seizures as infant (spasms)
- Head injury
- Autoimmune disorders

Who has LGS?

- LGS occurs secondary to many different causes including injury, brain malformations, infections, and genetic factors.
- Most with LGS may have abnormal brain imaging, but some have normal brain imaging prior to developing LGS.
- LGS can also develop from other epilepsy syndromes such as West, Ohtahara, Hypothalamic Hamartoma, etc.
- Many genes are associated with LGS and each gene is a risk factor for developing LGS if seizures remain uncontrolled.
- Emerging evidence suggests that genetic factors account for most unknown causes of LGS.

How does LGS change over time?

- There is no cure for LGS. Seizures may go into remission, and may also recur.
- 30-50% of children with infantile spasms will develop LGS.
- 80-90% of children with LGS will continue to have seizures into adulthood.
- Up to 70% with LGS will no longer show slow spike-and-wave (<3Hz) on EEG in adulthood.
- Most with LGS show paroxysmal fast rhythms (10-20 Hz) on EEG, mainly during non-REM sleep, at some point in their life.
- 70% with LGS will show cognitive impairment at diagnosis and more than 50% suffer behavioral issues including hyperactivity, sleep disturbances, rage attacks, aggression, and autistic features.
- The mortality rate is 5%. Those with LGS are 24 times more likely to die prematurely.
- Premature death in LGS is often due to SUDEP*, seizures, injury, or the underlying brain disorder.