

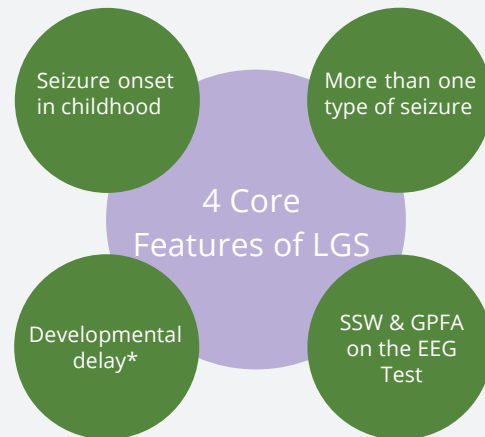


What is LGS?

Lennox-Gastaut Syndrome (LGS) is a severe epilepsy syndrome that develops in young children and often leads to lifelong disability. Nobody is born with LGS. It develops over time.

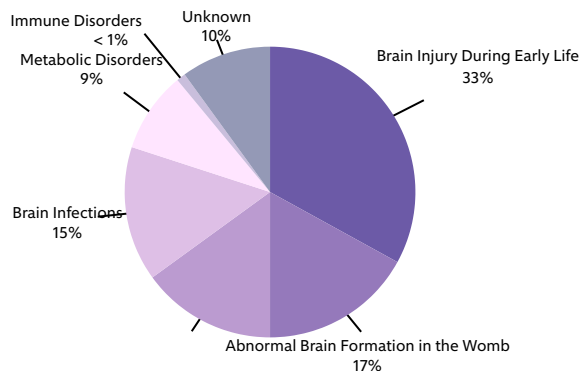
In LGS:

- Seizures usually begin in the pre-school years.
- More than one seizure type is always present.
- Tonic seizures are present in nearly all with LGS at some point.
- Seizures are nearly always treatment-resistant.
- Many LGS Associated Disorders exist including issues with sleep, behavior, movement, feeding, toileting, communication, and others.
- Slow spike and wave (SSW) and Generalized Paroxysmal Fast Activity (GPFA) are seen on the EEG. These are hallmarks of LGS.
- SSW and GPFA usually emerge between ages 3-5 years but can begin later in childhood.
- SSW and GPFA occur between seizures and can worsen seizures, development, and behavior problems.



*Developmental delay is not required to make the LGS diagnosis and 30% of kids are typically developing at diagnosis.

What causes seizures in LGS?



- In LGS, the brain is affected at a critical time in its development. Seizures and developmental problems result.
- There are many causes of seizures that can evolve into LGS. In most but not all cases, a cause can be found. Finding it can take extensive testing. In some cases, more than one cause is found.
- Knowing the cause of seizures can help identify an effective treatment in some cases.
- Most with LGS may have abnormal brain imaging seen on the MRI test, but some have normal brain imaging prior to developing LGS.
- LGS can also develop from other epilepsy syndromes such as West, Ohtahara, EIEE, Hypothalamic Hamartoma, Infantile Spasms, etc.

Some genes and genomic regions that cause seizures that can evolve into LGS:

| | | |
|-----------|--------|---------------------|
| ALG13 | GNAO1 | PCDH19SLC35A2 |
| ARX | GRIN1 | PIGASPTAN1 |
| CACNA2D2 | GRIN2A | PLCB1ST3GAL3 |
| CLN1/2/5 | GRINBB | PPP3CASTXBP1 |
| CDKL5 | HNRNPU | PTENTBD1D24 |
| DNM1 | KCNT1 | SCA2TCF4 |
| DOCK7 | KCNQ2 | SCN1ATSC1/2 |
| FLNA | MAGI2 | SCN2AWWOX |
| FOXP1 Dup | MEF2C | SCN8ADup 15q |
| GABRA1 | NEDDL4 | SETBP122q Del |
| GABRB3 | NDP | SIK1Trisomy 21 |
| GLI3 | NRXN1 | SLC25A22 |
| | | and many more . . . |

Who has LGS?

1-2%
of people with epilepsy

3-4%
of children with epilepsy

48,000
children and adults in the U.S.

1,000,000
children and adults have LGS worldwide

How does LGS change over time?

- There is no cure for LGS.
- Seizures in LGS 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.
- Over 95% with LGS are intellectually disabled.
- Up to 70% with LGS will no longer show slow spike-and-wave (<3Hz) on EEG in adulthood.
- Over 50% suffer behavioral issues including hyperactivity, sleep disturbances, rage attacks, aggression, and autistic features.
- Those with LGS are 14 times more likely to die prematurely. Premature death in LGS is often due to SUDEP*, aspiration pneumonia, seizures, injury, or the underlying brain disorder.

The LGS Foundation is committed to finding a cure for this devastating disorder!

