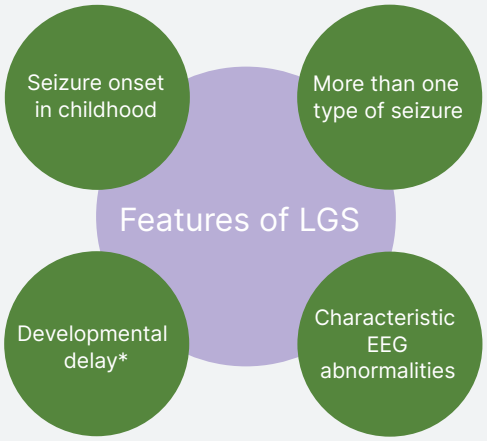
LGS FOUNDATION
LENNOX GASTAUT SYNDROME

CELEBRATING 13 YEARS
A decade of service improving the lives of individuals affected by LGS through research, family support programs, and education.

www.LGSfoundation.org

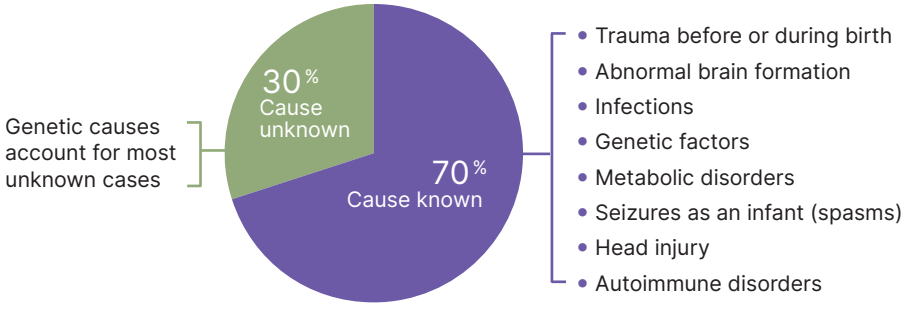
What is LGS?

- Lennox-Gastaut Syndrome (LGS) is a rare epilepsy syndrome. It is one of the Developmental and Epileptic Encephalopathies (DEEs).
- 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.



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

What causes seizures in LGS?



Some genes and genomic regions associated with LGS:

ALG13	GNAO1	PCDH19	SLC35A2
ARX	GRIN1	PIGA	SPTAN1
CACNA2D2	GRIN2A	PLCB1	ST3GAL3
CLN1/2/5	GRINBB	PPP3CA	STXBP1
CDKL5	HNRNPU	PTEN	TBD1D24
DNM1	KCNT1	SCA2	TCF4
DOCK7	KCNQ2	SCN1A	TSC1/2
FLNA	MAGI2	SCN2A	WWOX
FOXG1 Dup	MEF2C	SCN8A	Dup 15q
GABRA1	NEDDL4	SETBP1	22q Del
GABRB3	NDP	SIK1	Trisomy 21
GLI3	NRXN1	SLC25A22	

- 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.

Who has LGS?

1-2%
of people with epilepsy

3-4%
of children with epilepsy

48,000
childen 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 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.

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*SUDEP – Sudden Unexpected Death in Epilepsy.

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