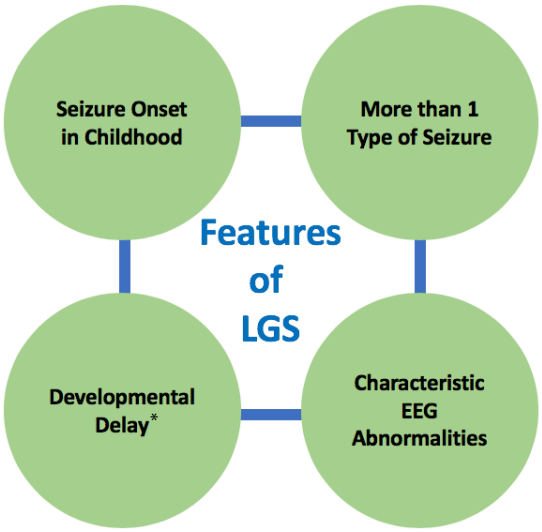
LGS FOUNDATION
LENNOX-GASTAUT SYNDROME

CELEBRATING 10 YEARS

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

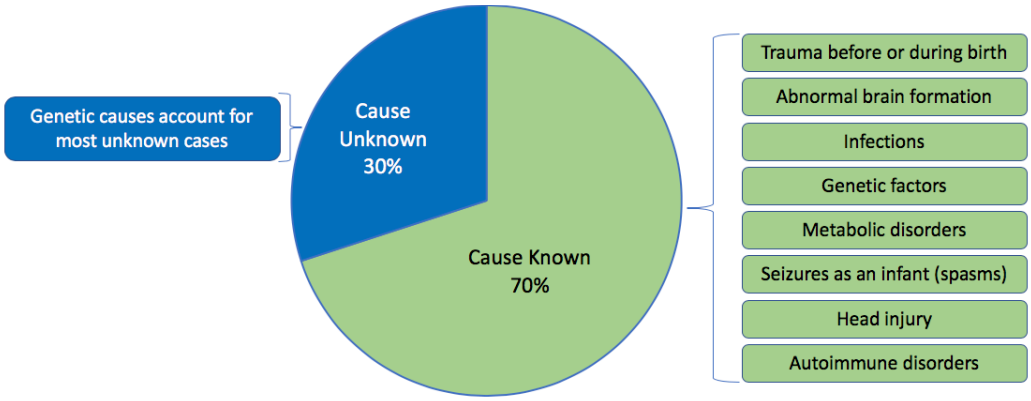
What is LGS?

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

- 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
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 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.

www.LGSfoundation.org

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*SUDEP – Sudden Unexpected Death in Epilepsy.