Lennox-Gastaut syndrome (LGS) key facts

Inspired by **patients.** Driven by **science.**

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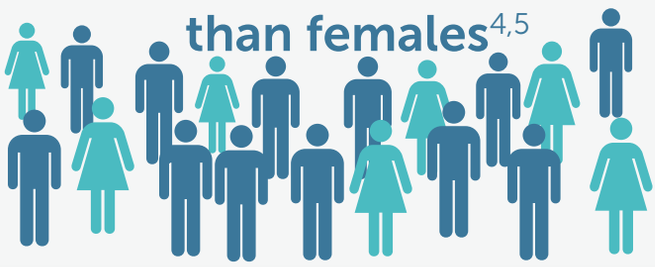
LGS: a rare and severe form of epilepsy that usually starts in childhood[1]

Nobody is born with LGS. It may develop over time from childhood seizures that remain uncontrolled by treatments[2]

LGS affects an estimated **2** in **10,000** people in the European Union[3]

LGS is more common in males than females[4,5]



Known causes (in approximately 65% to 75% of patients with LGS):[4,6,7]

- Underlying structural brain abnormality (from head trauma, birth injury from childbirth complications, tuberous sclerosis, infection such as encephalitis and meningitis, brain malformations, or tumours)
- Genetic disorders
- Metabolic causes

Diagnostics:[8-10]

- Clinical history (seizure types and intellectual impairment)
- Clinical evaluation (e.g., EEG and neuroimaging studies such as MRI, CT or SPECT)
- Genetic testing
- Lab testing

Key characteristics:[11]

- Seizure onset in childhood
- More than one type of seizure
- Abnormal brain waves on the electroencephalogram (EEG)
- Developmental delay*[12]

*Developmental delay is not required to make the LGS diagnosis and 30% of children are typically developing at diagnosis[12]

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Estimated incidence in children

0.1 to 0.28 per 100,000 population[7,13]

75%

of patients may experience daily seizures[14]

80-90%

of children with LGS will continue to have seizures into adulthood[15]

Children and adults with LGS

share similar features, primarily suffering from multiple types of seizures that do not respond well to treatments (drug-resistant seizures)[10,11]

The most common seizure types are:[2,11,16–18]

Tonic	stiffness in the arms and legs
Atonic seizures	sudden relaxing of muscles, usually causing the person to fall
Generalised tonic-clonic	begins with stiffness in the arms and legs, followed by jerky movements in the arms, legs, and head with a loss of consciousness
Atypical absence	brief altered consciousness with prolonged staring and subtle movements
Non-convulsive status epilepticus	prolonged seizure activity without convulsions
Myoclonic	involuntary, brief, jerk-like movements that cause a sudden muscle contraction
Focal impaired awareness	affects a limited area of the brain and the affected person remains conscious. These may remain focal or evolve to bilateral tonic-clonic seizures
Epileptic spasm	brief events of arm, leg, head flexion or extension

Other features in LGS include:[11]

- Impact on sleep
- Cognitive delay and behavioural issues
- Motor and mobility difficulties

LGS

- Has a significant impact on quality of life (QoL) for both patients and their families[19]
- Patients often suffer from lifelong motor, cognitive, and behavioural abnormalities[9,19]
- Poor long-term outcomes for patients[9]
- Complete seizure freedom is unusual[9]
- Has an increased risk of sudden unexpected death in epilepsy (SUDEP) due to uncontrolled seizures[20-22]

Key challenges for caregivers:[23,24]

- Logistics around **providing care**
- **Emotional burden** and **isolation**
- Family **dynamics**
- Financial **strain**
- Transitioning from **paediatric** to **adult** care
- **Lack** of understanding of **LGS**
- **Fear, anxiety, stress** and **depression**

LGS and other epilepsy syndromes share clinical and imaging features[11,25]

Epilepsy syndrome	Distinguishing features compared to LGS
Epilepsy with myoclonic-atonic seizures	• Normal development prior to seizure onset • Myoclonic-atonic seizures • Faster generalised spike-and-wave pattern, typically >3 Hz
Prolonged, hemiclonic seizures in infancy (typically seen in Dravet syndrome)	• Prolonged, hemiclonic seizures • Tonic seizures occur later
DEE spike-and-wave activation in sleep (SWAS) and EE-SWAS	• Regression and marked activation of epileptiform abnormalities in sleep • SWAS – 'nearly continuous diffuse SW complexes in slow-wave sleep'
Ring chromosome 20 syndrome	• Refractory epilepsy • Intellectual disability • Behavioural abnormalities • Tonic seizures usually appear during sleep

Abbreviations: LGS, Lennox-Gastaut syndrome; EEG, Electroencephalogram; MRI, Magnetic resonance imaging; CT, Computed tomography; SPECT, Single-photon emission computed tomography; QoL, Quality of life; SUDEP, Sudden unexpected death in epilepsy; DEE, Developmental and epileptic encephalopathy; SWAS, Spike-wave activation in sleep; EE-SWAS, Epileptic encephalopathy with spike-wave activation in sleep; SW, Spike-wave.

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