Living with Lennox-Gastaut Syndrome

A severe childhood-onset developmental and epileptic encephalopathy,[1,2] characterized by different types of seizures as well as serious impairment of neurodevelopmental, cognitive, and motor functions[3]

Safety concerns for patients with LGS

LGS is characterized by many different seizure types. These seizure types—tonic, atonic, generalized tonic-clonic, atypical absence and myoclonic—may result in frequent falls, injuries and "drop attacks" that can involve the head or the whole body.[1,3]

Moreover, the cognitive impairment of patients with LGS may impact their speech. Some patients with LGS can only use non-verbal communication[10] and this may put patients in a position of an increased risk of entering into potentially dangerous situations, if not properly supervised.[11] Importantly, families and caregivers may need to take appropriate precautions, to ensure their loved one diagnosed with LGS is safe.[11]

What is LGS?

LGS is a rare, severe form of epilepsy that typically starts during childhood and that persists into adulthood.[4] Approximately 50% of infants with a severe developmental and epileptic encephalopathy (DEE) evolve over time to LGS.[5]

LGS has far-reaching effects beyond seizures; issues with communication, psychiatric symptoms, sleep, behavioral challenges, and mobility are common.[6] Patients with LGS also experience developmental slowing, plateauing or regression, culminating in cognitive impairment.[7]

People with LGS are typically on multiple therapies and are often on three or more antiepileptic therapies at one time. However, seizures largely remain uncontrolled on currently available medications.[8]

Sudden unexpected death in epilepsy (SUDEP) is a major concern for patients with LGS.[9]

Burden of disease

- **Multiple, uncontrolled seizure types**[3]
- **Moderate to severe intellectual disability**[12]
- **Behavioral disorders (aggression, hyperactivity, or autistic spectrum)**[1]
- **High rate of seizure-related injuries**[10]
- **Long-term disability, and early mortality**[13]
- **Persists through childhood and adolescence to adult years**[14]

Reduced quality of life and impact of LGS on parents/caregivers and families

The associated intellectual and behavioral problems, as well as around-the-clock care requirements, add to the complexity of life with LGS.[8] LGS has a broad and dramatic impact[1] on the lives of patients and their caregivers due to the severity and frequency of treatment-resistant seizures, as well as co-morbidities and symptoms experienced by patients, including cognitive impairment, developmental delays, and behavioral problems. Nearly all adults with LGS are unable to live independently,[1] with many unable to walk, even with support.[15]

LGS impacts the entire family—parents, caregivers, and siblings—including:[16]

- **Stress and anxiety (especially if/when seizures will occur, the future prospects of the individual with LGS, and the social stigma associated with the condition)**
- **Feelings of isolation, which can lead to depression**
- **Restriction of social life and relationship problems between partners and other family members**
- **Physical exhaustion, chronic fatigue, and sleep deprivation**
- **Sibling isolation and neglect**
- **Financial concerns[13]**

LGS by the numbers:

30K-50K
is the estimated number of **patients with LGS** in the U.S.[8]

3-4%
of all **childhood epilepsies are diagnosed as LGS**[17]

95% of patients experience **cognitive impairment** within 5 years from the onset of epilepsy[3]

25.4%
of patients with LGS **cannot walk, even with support**[15]

How is LGS treated?

- Treatment options vary for patients with LGS, and none have been shown to eliminate seizures in all patients. Several ways to treat LGS include a combination of anti-seizure medicines, dietary therapy, surgery, neuromodulation, and other management issues.[18]
- Patients are often uncontrolled after multiple therapies.[8]

1. Strzelczyk A, Schubert-Bast S. Expanding the Treatment Landscape for LGS: Current and Future Strategies. CNS Drugs. 2021;35(1):61-83.
2. Scheffer IE, Liao J. Deciphering the concepts behind "Epileptic encephalopathy" and "Developmental and epileptic encephalopathy". Eur J Paediatr Neurol. 2020;24:11-14.
3. Mastrangelo M. LGS: A State of the Art Review. Neuropediatrics. 2017;48(3):143-151.
4. NORD. LGS. https://rarediseases.org/rare-diseases/lennox-gastaut-syndrome. Accessed October 7, 2021.
5. International League Against Epilepsy. Specchio, N, Wirrell, EC, Scheffer, IE, et al. Draft guideline: ILAE Classification and Definition of Epilepsy Syndromes with Onset in Childhood: Position Paper by the ILAE Task Force on Nosology and Definitions. https://www.ilae.org/files/dmfile/CHILDApril6withfigs.pdf. Accessed October 19, 2021.
6. LGS Foundation. LGS Characteristics and Major Concerns Survey. www.lgsfoundation.org/wp-content/uploads/2021/08/2019-PFDD-Caregiver-Survey-1.pdf, Accessed October 17, 2021.
7. LGS. Child Neurology Foundation. https://www.childneurologyfoundation.org/disorder/lennox-gastaut-syndrome. Accessed Nov 2, 2021.
8. Zogenix data on File. 2021.
9. Harden C, Tomson T, Gloss D, et al. Practice guideline summary: Sudden unexpected death in epilepsy incidence rates and risk factors: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society [published correction appears in Neurology. 2019 Nov 26;93(22):982] [published correction appears in Neurology. 2020 Mar 3;94(9):414]. Neurology. 2017;88(17):1674-1680.
10. Gallop K, Wild D, Verdian L, et al. LGS: development of conceptual models of health-related quality of life (HRQL) for caregivers and children. Seizure. 2010;19(1):23-30.
11. LGS Foundation. Seizures and Safety. www.lgsfoundation.org/seizure-safety. Accessed October 17, 2021.
12. NIH. LGS. https://medlineplus.gov/genetics/condition/lennox-gastaut-syndrome. Accessed October 17, 2021.
13. Reaven NL, Funk SE, Montouris GD, Saurer TB, Story TJ. Burden of illness in patients with possible Lennox-Gastaut syndrome: A retrospective claims-based study. Epilepsy Behav. 2018;88:66-73.
14. Kerr M, Kluger G, Philip S. Evolution and management of LGS through adolescence and into adulthood: are seizures always the primary issue?. Epileptic Disord. 2011;13 Suppl 1:S15-S26.
15. Kim HJ, Kim HD, Lee JS, Heo K, Kim DS, Kang HC. Long-term prognosis of patients with LGS in recent decades. Epilepsy Res. 2015;110:10-19.
16. Cross JH, Auvin S, Falip M, Striano P, Arzimanoglou A. Expert Opinion on the Management of Lennox-Gastaut Syndrome: Treatment Algorithms and Practical Considerations. Front Neurol. 2017;8:505. Published 2017 Sep 29.
17. Literature review of LGS epidemiology by bioStrategies Group, January 2021.
18. LGS Foundation. How is LGS treated? www.lgsfoundation.org/about-lgs-2/how-is-lgs-treated/. Accessed November 2, 2021.

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