

LGS CHARACTERISTICS AND MAJOR CONCERNS SURVEY

In 2018, the LGS Foundation conducted an in-depth survey of its 4,500 member patient and family community to better understand the frequency of symptoms seen in LGS, aspects of seizure treatment, and patient priorities. The survey was designed, with permission, from a survey of a similar Developmental and Epileptic Encephalopathy (DEE, Villas et al., 2017). 56 questions were asked in varying formats and it took an average of 20 minutes to complete.

Demographics

A total of 413 individuals from 17 counties (89% U.S.) responded to the survey. Only one respondent had LGS. The remaining were family members answering on behalf of a loved one. Ages for the individual with LGS ranged from 7 months to 60 years.

(N=413)

RESPONDENTS

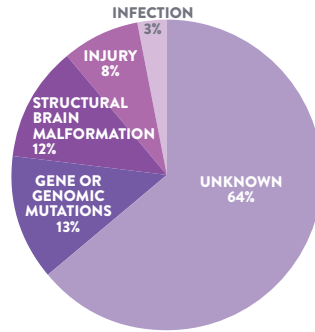
Individual with LGS	0.2%
Caregivers/family members	99.8%

INDIVIDUALS WITH LGS

Female	41%
Male	59%
Current age (3 years or younger)	4%
Current age (4-15 years)	49%
Current age (16-25 years)	29%
Current age (26-60 years)	18%

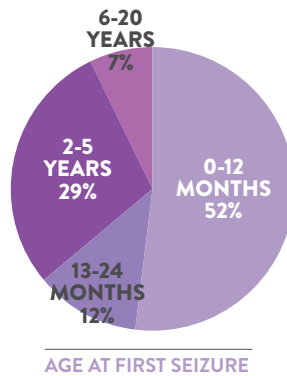
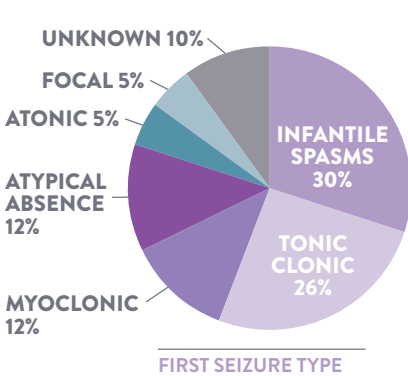
Cause/Etiology

A diverse number of causes of the seizures were reported. The majority (64%) did not know the cause. For genomic causes, 41 different genes/gene regions were reported. 94% of respondents said an EEG showing slow-spike and wave was present at some point in the disease.



Seizure Onset

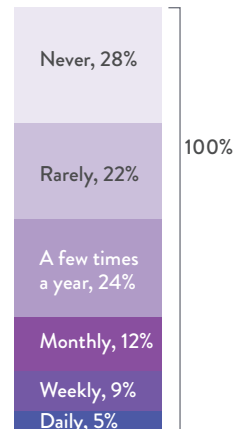
45% reported previous infantile spasms even if it was not the first seizure type. 72% said it took more than 1 year to get a diagnosis of LGS (range 1-20 years).



Seizure Injuries

81% report ever having a seizure-related head injury. Frequency of injuries in the last 12 months are listed below.

HEAD INJURIES IN THE LAST 12 MONTHS



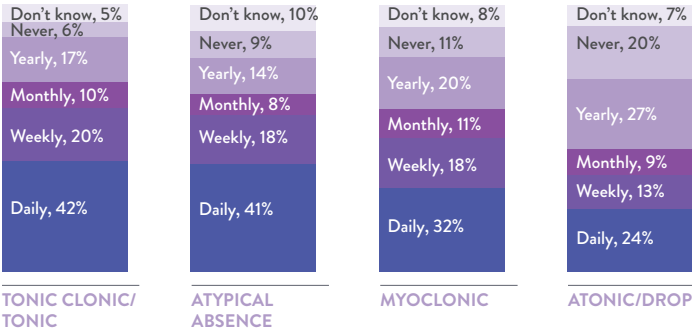
LGS and the many DEEs it is associated with represent a lifelong disability for most patients. Patients experience seizures throughout their lifetime, frequent injuries and hospitalizations, and many comorbid conditions. This survey highlights major unmet needs for better treatments for LGS and offers insight into caregiver priorities on the backdrop of the multiple chronic conditions seen in LGS. This work identifies potential areas where therapies and supportive services would be meaningful.

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Current Seizure Frequency

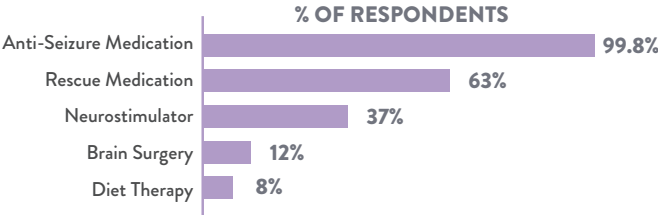
55% of respondents said the person with LGS was currently having 4 or more types of seizures. 42% said the person with LGS was having 1-3 types of seizures. Only 3% reported seizure freedom. The most problematic seizure types currently present were 1) Tonic Clonic, 2) Tonic, 3) Atonic, 4) Myoclonic, 5) Atypical Absence.

TYPE AND FREQUENCY OF CURRENT, MOST PROBLEMATIC SEIZURES



Current Treatments

Treatments for LGS include medications, neurostimulation, brain surgery, diets, and alternative therapies. Respondents report that on average they have tried 10-12 seizure drugs to date. The frequency of those currently on a given treatment are reported below.



Current Number of Anti-Seizure Drugs Used

- 99.8% currently use anti-seizure drugs
- 75% report the person with LGS is taking 3 or more drugs (range 3-10)
- 25% report the person with LGS is taking 1 or 2 drugs
- One respondent reported zero medication use
- The top 5 medications reported were Clobazam, Valproate, Rufinamide, Levetiracetam, and Cannabidiol.

Most common respondent concerns in the open response section:

1. Seizures & safety
2. Developmental delay, especially communication
3. Behaviors, especially aggression
4. Mobility & physical care issues
5. Sleep disturbances
6. Medical costs & access to care
7. Social isolation

Seizure Clusters and Status Epilepticus

- 94% have experienced seizure clusters
- 65% have had seizures lasting longer than 5 minutes
- 63% report using at home rescue medicines in the last 12 months

Symptoms Other than Seizures

LGS has far reaching effects beyond just seizures. Issues with development, communication, psychiatric symptoms, sleep, behavior, and mobility are common. The frequency of the most commonly reported issues in these domains are shown below.

ISSUE	%
DEVELOPMENTAL	
Severe Development Delay	80%
Moderate Delay	18%
Mild Delay	2%
COMMUNICATION	
Unable to read	72%
Unable to write	69%
Non-verbal	60%
PSYCHIATRIC	
Aggression	59%
Irritability/Agitation	41%
Impulsiveness	33%
SLEEP	
Sleep Disturbances	87%
Nocturnal Seizures	81%
Insomnia	58%
AUTISTIC FEATURES	
Diagnosed with Autism	26%
Repetitive body movements	39%
Tactile sensitivity or sensory issues	38%
MOBILITY	
Uses a wheelchair or adaptive stroller	73%
FEEDING	
Tube fed	27%