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%

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.

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.

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 | %
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**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%

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.

| % OF RESPONDENTS | Anti-Seizure Medication | Rescue Medication | Neurostimulator | Brain Surgery | Diet Therapy |
--- | --- | --- | --- | --- | ---
99.8% |  |  |  |  | 8%
63% |  |  | 37% |  |  |
12% |  |  |  | 12% |  |
8% |  |  |  |  | 8%

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