2.106 REST-LGS Tool: Real-World Use to Screen for LGS and Improve Access to Care

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Background

- Lennox-Gastaut syndrome (LGS) is frequently undiagnosed in the adult population (Figure 1).
- Identifying LGS in adult patients can be difficult because the disease evolves over time, resulting in changes in the characteristic seizure type(s) and patient's electroencephalogram (EEG) status[1–3]; additionally, details of patients' medical histories may be lost during transfer of pediatric testing results to adult health care providers.[4,5] Without appropriate diagnosis, patients do not receive optimal care.
- The *Refractory Epilepsy Screening Tool for LGS* (REST-LGS) was created to improve the identification of patients with LGS (Figure 1).[6]
- The tool was previously validated in two medical centers and can be used by both epilepsy and non-epilepsy experts to identify patients with LGS.[6]

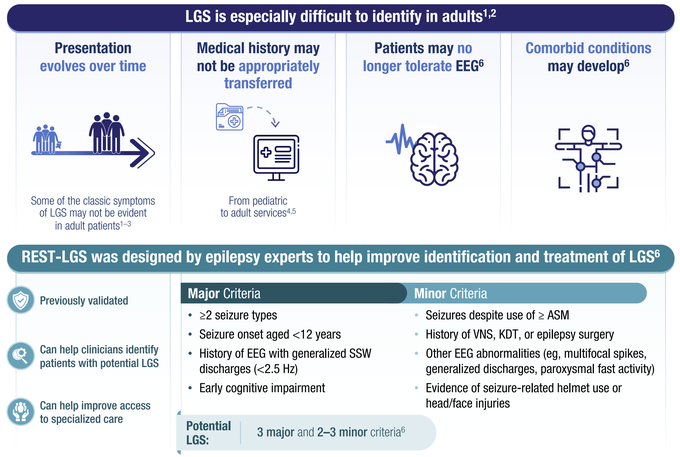
Objective

- To further validate the REST-LGS in a real-world setting for adults with drug-resistant epilepsy (DRE) and intellectual and developmental disabilities (IDD).
- To develop a scoring system that will simplify use of the tool.

Methods

- This was a retrospective chart review of 100 patients (aged ≥18) with DRE and IDD who lived in a residential care facility for patients with IDDs.
- Reviews were performed by two primary care providers who were blinded to the prior diagnoses.
- The primary care providers reviewed patients' chart notes from the last 3 visits and used the previously validated REST-LGS[6] to identify which, if any, of the criteria the patients met.
- Study data were collected on paper case report forms, which were entered directly into a Research Electronic Data Capture (REDCap) database and each independently completed CRFs for each patient with DRE was analyzed to determine the validity of the REST-LGS. The expert team met to discuss the outcomes of the analysis and to refine the tool.
- The criteria in REST-LGS were weighted to obtain a total score, which could then be used to identify "likely," "possible," or "unlikely" LGS.

Figure 1. Identifying potential LGS in adult patients with DRE

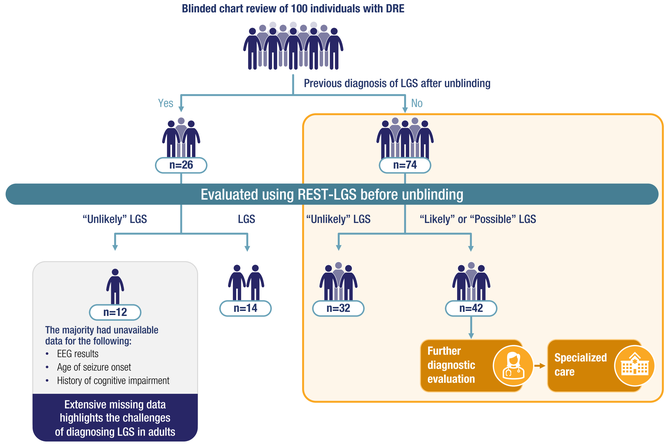


ASM, antiseizure medication; DRE, drug-resistant epilepsy; EEG, electroencephalogram; KDT, ketogenic diet therapy; LGS, Lennox-Gastaut syndrome; REST-LGS, Refractory Epilepsy Screening Tool for LGS; SSW, slow spike-and-wave; VNS, vagus nerve stimulation.

Table 2. Summary of the reviewers' responses to REST-LGS criteria

	n (%)		
Gender			
Male	54 (54)		
Female	46 (46)		
Criteria	**Yes**	**No**	**Unavailable**
1. Persistent seizures despite trial of 2 or more antiseizure medications	65 (65)	32 (32)	3 (3)
2. Two or more seizure types	56 (56)	36 (36)	8 (8)
3. Seizure onset before the age of 12 years	57 (57)	1 (1)	42 (42)
4. Evidence of seizure-related helmet use or head/face injuries	15 (15)	46 (46)	39 (39)
5. Cognitive impairment since childhood (may include past or current learning difficulties, a history of special education, autism, intellectual disabilities, or developmental delays)	71 (71)	1 (1)	28 (28)
6. History of vagal nerve stimulator, ketogenic diet, or epilepsy surgery	10 (10)	59 (59)	31 (31)
7. History of EEG with generalized slow spike-and-wave discharges (<2.5 Hz)	36 (36)	18 (18)	46 (46)
8. One of the following electroencephalogram abnormalities: multifocal spikes, symptomatic generalized discharges, generalized polyspikes, generalized periods of attenuation of background/electrodecrement, or paroxysmal fast activity	36 (36)	15 (15)	49 (49)

Figure 3. Evaluation of patients using REST-LGS



EEG, electroencephalogram; DRE, drug-resistant epilepsy; LGS, Lennox-Gastaut syndrome; REST-LGS, Refractory Epilepsy Screening Tool for LGS.

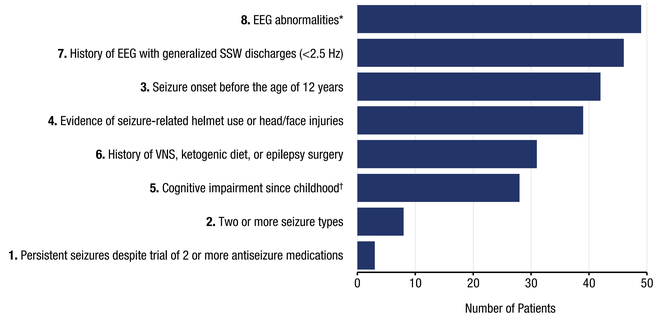
Table 1. REST-LGS with weighted criteria to identify patients with potential LGS

Complete demographic information and determine whether the patient meets the criteria:			
Patient name:	__Male __Female __Nonbinary		
Patient ID:	**Date of birth:**		
Date of evaluation:	**Name of evaluator:**	**Check appropriate response and add assigned values:**	
1. Persistent seizures despite trial of 2 or more antiseizure medications	❑ YES – 1	❑ NO – 0	❑ Unavailable – 0
2. Two or more seizure types	❑ YES – 3	❑ NO – 0	❑ Unavailable – 0
3. Seizure onset before the age of 12 years	❑ YES – 3	❑ NO – 0	❑ Unavailable – 0
4. Evidence of seizure-related helmet use or head/face injuries	❑ YES – 1	❑ NO – 0	❑ Unavailable – 0
5. Cognitive impairment since childhood (may include past or current learning difficulties, a history of special education, autism, intellectual disabilities, or developmental delays)	❑ YES – 3	❑ NO – 0	❑ Unavailable – 0
6. History of vagal nerve stimulator, ketogenic diet, or epilepsy surgery	❑ YES – 1	❑ NO – 0	❑ Unavailable – 0
7. History of EEG with generalized slow spike-and-wave discharges (<2.5 Hz)	❑ YES – 3	❑ NO – 0	❑ Unavailable – 0
8. One of the following EEG abnormalities: multifocal spikes, symptomatic generalized discharges, generalized periods of attenuation of background/electrodecrement, or paroxysmal fast activity	❑ YES – 1	❑ NO – 0	❑ Unavailable – 1
Likely LGS: >11 **Possible LGS: 8–11** **Unlikely LGS: <8**	**Total:**		

EEG, electroencephalogram; LGS, Lennox-Gastaut syndrome.

- Based on the total score, patients could be identified as likely, possible, or unlikely to have LGS.

Figure 2. Summary of criteria for which the response was 'unavailable'



*EEG abnormalities include multifocal spikes, symptomatic generalized discharges, generalized polyspikes, generalized periods of attenuation of background/electrodecrement, or paroxysmal fast activity; †Cognitive impairment may include past or current learning difficulties, a history of special education, autism, intellectual disabilities, or developmental delays. SSW, slow spike-and-wave; VNS, vagal nerve stimulator.

- No information was available on the history of SSW in EEG in 45% of patients and the age at seizure onset in 40% of patients; both are considered major criteria for the identification of patients with potential LGS.

Conclusions

- In a retrospective chart review study of 100 patients to further validate the previously developed REST-LGS for the identification of potential LGS in patients with DRE:
  - The charts reviewed had extensive data missing, which highlights the challenges associated with diagnosing LGS in adults with IDD
  - Of the 74 patients who had not been previously diagnosed with LGS, 42 (57%) were identified as 'likely' or 'possible' LGS using REST-LGS
  - All individuals identified as 'likely' or 'possible' to have LGS should be referred for further diagnostic evaluation at a specialized epilepsy center
  - The study was limited by a small sample size and the inclusion of only the last 3 entries from patients' charts in the evaluation; additionally, the analysis relied on patient records only, no consultation with patients was conducted during the screening phase
- REST-LGS will enable the user to identify potential LGS in previously undiagnosed patients with DRE, thus opening the door to specialized care.
- Further education is needed to promote use of screening tools among physicians to identify patients with LGS to ensure proper care and treatment is provided to these patients.

References: 1. Ferlazzo E et al. *Epilepsy Res.* 2010;89:271-277. **2.** Oguni H et al. *Epilepsia.* 1996;37(Suppl. 3):44-47. **3.** Yagi K. *Epilepsia.* 1996;37(Suppl. 3):48-51. **4.** Arzimanoglou A et al. *Lancet Neurol.* 2009;8:82-93. **5.** Kerr M et al. *Epileptic Disord.* 2011;13:S15-S26; **6.** Piña-Garza JE. *Epilepsy Behav.* 2019;90:148-153.

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