

The refractory epilepsy screening tool for Lennox–Gastaut syndrome (REST-LGS)

Piña-Garza JE, Boyce D, Tworek DM, Davis KA, Gatens H, Lai G, McGoldrick PE, Thomas B, Wolf SM. *Epilepsy Behav.* 2019;90:148–153.

Introduction

Lennox–Gastaut syndrome (LGS) is a severe childhood-onset epileptic encephalopathy typically characterized by multiple intractable seizure types, cognitive impairment, and an abnormal electroencephalogram (EEG) with generalized slow spike-and-wave (SSW) discharges (1.5–2.5 Hz). The syndrome continues to persist into adulthood, at which point the typical symptoms become harder to identify.

Early diagnosis of LGS can help patients and caregivers set informed expectations related to prognosis and long-term management in addition to providing services specific to LGS. To improve both the identification and treatment of patients with LGS, a group of experts who care for patients with refractory epilepsy and intellectual/developmental disabilities created a tool that works to distinguish between LGS and refractory epilepsy—the Refractory Epilepsy Screening Tool for LGS (REST-LGS).

This study is limited by its retrospective design, which could result in incomplete patient records (i.e., a designation of helmet use not available on all charts) and nonresponder bias. Additionally, rather than specifying the Cronbach's α analysis a priori, it was undertaken after unblinding to determine the tool's internal consistency.

The REST-LGS was developed through a rigorous 10-step process

Collective judgment from a group of experts was combined with available scientific evidence

- Clinical panelists were required to be:
- Long-tenure experts in the field of epilepsy
 - Affiliated with clinical centers with high-volume management of epilepsy including LGS

Clinical criteria were identified and evaluated based on importance

Major criteria	Minor criteria
≥2 seizure types	Persistent seizures despite trial of ≥2 antiepileptic drugs (AEDs)
Seizure onset at <12 years of age	History of vagal nerve stimulator (VNS), ketogenic diet, or epilepsy surgery
History of EEG with generalized SSW discharges <2.5 Hz	Evidence of seizure-related helmet use/head or face injuries
Cognitive impairment since childhood	History of other EEG abnormalities

The validity and reliability of the REST-LGS were evaluated using medical records from 200 patients

- Diagnosis-blind raters at 2 large epilepsy centers completed a case report form for 100 patients at each site (50 patients with LGS diagnosis and 50 with drug-resistant epilepsy)

Preliminary validation suggests REST-LGS is beneficial to distinguish between LGS and other refractory epilepsies

- In a diagnosis-blinded chart review, a majority of patients with confirmed LGS diagnosis met 3 major criteria and 2 to 3 minor criteria
- Nonexpert and expert raters scored similarly

Refractory epilepsy screening tool case report form

Please fill out demographic information and determine whether the patient meets the following criteria:

Patient ID #		Gender: <input type="checkbox"/> Male <input type="checkbox"/> Female	
Patient's age at last visit:		Date of chart review:	
1. Persistent seizures despite trial of 2 or more antiseizure medications			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
2. Two or more seizure types			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
3. Seizure onset before age of 12 years			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
4. Evidence of seizure-related helmet use, or head or face injuries			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
5. Cognitive impairment since childhood (may include past or current learning difficulties, history of special education, autism, intellectual disabilities or developmental delays)			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
6. History of vagal nerve stimulator (VNS), ketogenic diet, or epilepsy surgery			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
7. History of EEG with generalized slow spike-and-wave (SSW) discharges (<2.5Hz)			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
8. One of the following EEG abnormalities: multifocal spikes, symptomatic generalized discharges, generalized polyspikes, generalized periods of attenuation of background/ electrodecement, or paroxysmal fast activity			<input type="checkbox"/> YES <input type="checkbox"/> NO <input type="checkbox"/> Unavailable
Rater feedback, comments, and/or questions:			

Use of the REST-LGS in clinical practice may lead to more timely diagnosis of LGS.

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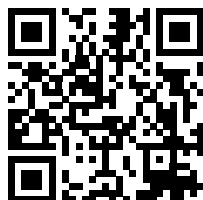
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