■ Multimedia teaching material

Epileptic Disord 2022; 24 (4): 729-732

Epileptic Disorders

Diagnosing Lennox-Gastaut syndrome in an adult and its direct impact in epilepsy care

Fábio A. Nascimento[1], Elizabeth A. Thiele[1], Daniel J. Weber[2]

[1] Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA
[2] Department of Neurology, Saint Louis University, Saint Louis, MO, USA

Received February 3, 2022;
Accepted April 2, 2022

• **Correspondence:**
Fábio Augusto Nascimento e Silva
55 Fruit St., Wang 7th floor, Boston, MA 02114, USA
<nascimento.fabio.a@gmail.com>
<fnascimento@mgh.harvard.edu>

doi:10.1684/epd.2022.1445

We report a 34-year-old male with intellectual disability and refractory, childhood-onset, non-lesional epilepsy featuring absence, generalized tonic-clonic, and tonic seizures leading to falls. Seizure onset occurred at the age of 11 years, and tonic seizures did not appear until he was an adult. Notably, tonic seizures were mostly during sleep. Physical examination did not show dermatologic or ophthalmologic abnormalities. Current antiseizure drugs include zonisamide and cenobamate, and he underwent vagus nerve stimulator implantation and corpus callosotomy as an adult. EEG showed slow (≤2.5 Hz) generalized spike-and-wave discharges, and generalized paroxysmal fast activity (*figures 1, 2*). Brain MRI did not show any epileptogenic lesion; genetic testing was not pursued. The triad of intellectual disability, multiple seizure types including tonic seizures, and slow generalized spike-and-wave discharges is consistent with Lennox-Gastaut syndrome (LGS). Diagnosing this syndrome has direct implications in care including several new medication options [1, 2]. Notably, although LGS can typically be differentiated from other epilepsy syndromes (such as Doose, Dravet, West syndromes and atypical benign partial epilepsy) based on electroclinical features, achieving an accurate diagnosis may be challenging [3]. ■

Supplementary material.
Summary slides accompanying the manuscript are available at www.epilepticdisorders.com.

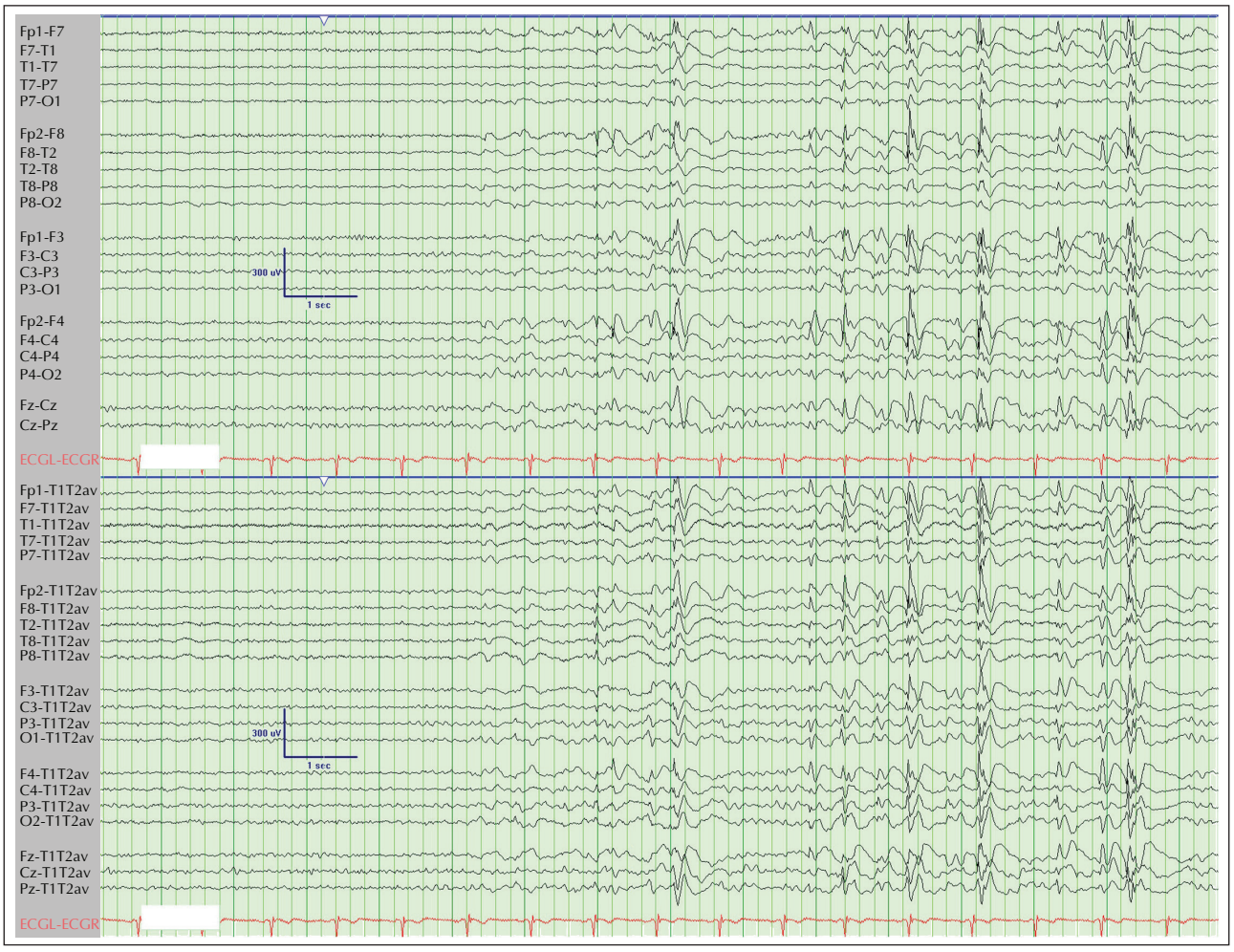
Disclosures.
F. Nascimento is an Associate Editor of Epileptic Disorders. D. Weber is on the Speaker's Bureau for SK Life Science. E. Thiele reports no disclosures relevant to the manuscript.

References

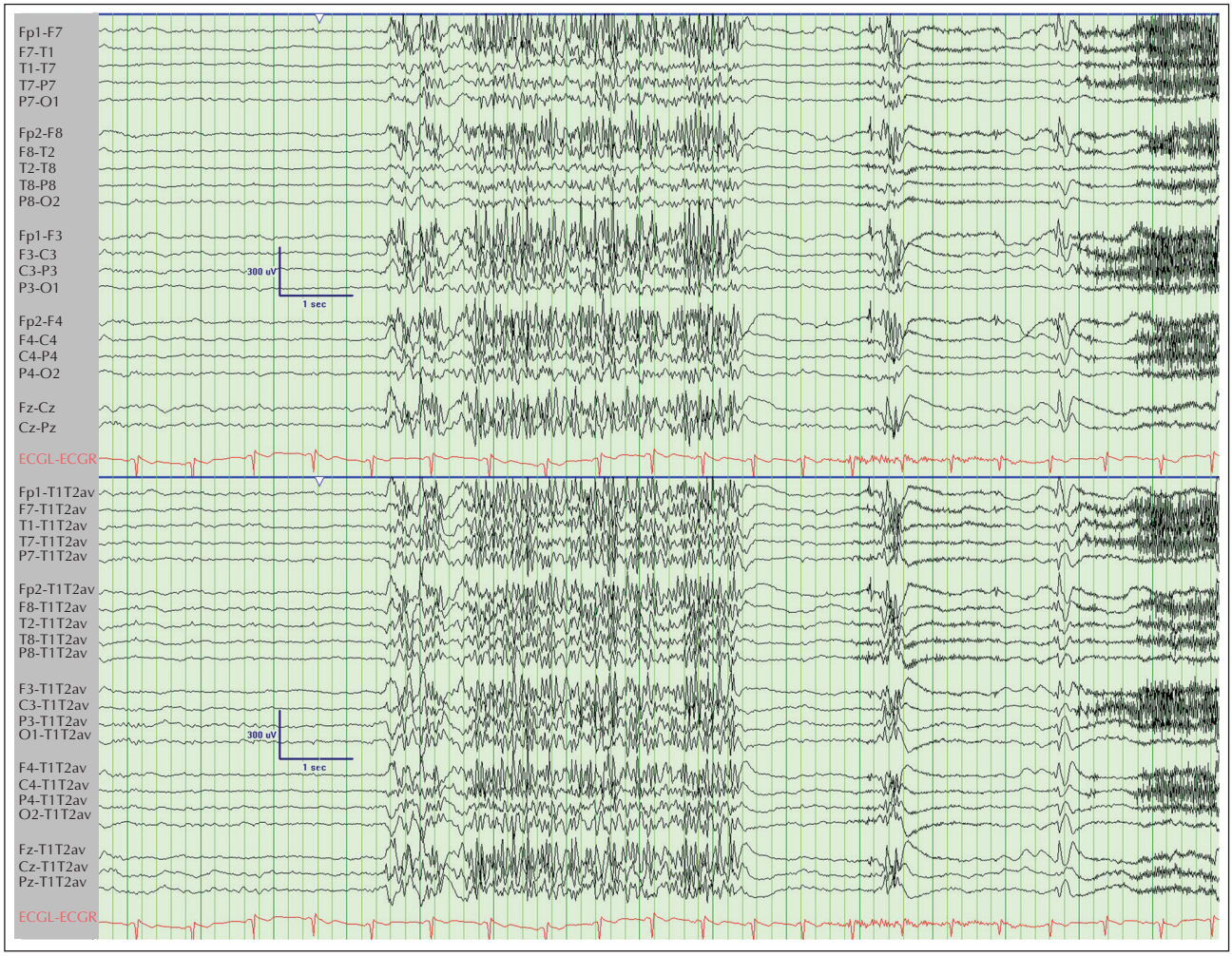
1. Brigo F, Jones K, Eltze C, Matricardi S. Antiseizure medications for Lennox-Gastaut syndrome. *Cochrane Database Syst Rev* 2021; 4(4): CD003277.

2. Thiele EA, Marsh ED, French JA, Mazurkiewicz-Beldzinska M, Benbadis SR, Joshi C, *et al*. Cannabidiol in patients with seizures associated with Lennox-Gastaut syndrome (GWPCARE4): a randomized, double-blind, placebo-controlled phase 3 trial. *Lancet* 2018; 391(10125): 1085-96.

3. Bourgeois B, Douglass LM, Sankar R. Lennox-Gastaut syndrome: a consensus approach to differential diagnosis. *Epilepsia* 2014; 55(4): 4-9.



■ **Figure 1.** Routine EEG shows slow generalized spike-and-wave discharges: bipolar (upper panel) and referential (lower panel) montages show 100-300 µV, 2-2.5 Hz generalized (poly)spike-and-wave discharges (sensitivity: 15 µV/mm, LF: 1 Hz, HFF: 70 Hz, notch off).



■ **Figure 2.** Routine EEG shows generalized paroxysmal fast activity: bipolar (upper panel) and referential (lower panel) montages show a 6-to-7-second run of 100-200 µV, 15-25 Hz generalized, sharply contoured activity without clinical manifestations, consistent with generalized paroxysmal fast activity (GPFA) (sensitivity: 15 µV/mm, LF: 1 Hz, HFF: 70 Hz, notch off).

TEST YOURSELF

(1) The electroclinical triad characteristic of Lennox-Gastaut syndrome includes (please select all that apply):

A. Intellectual disability
B. Multiple seizure types
C. 3-4 Hz generalized spike-and-wave-discharges
D. ≤2.5 Hz generalized spike-and-wave discharges

(2) Lennox-Gastaut syndrome should be differentiated from other epilepsy syndromes including (please select all that apply):

A. Dravet syndrome
B. Doose syndrome
C. West syndrome
D. Atypical benign partial epilepsy

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com.