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

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Vignette

 We report a 34-year-old male with intellectual disability and refractory, childhoodonset, non-lesional epilepsy featuring absence, generalized tonic-clonic, and tonic seizures, leading to falls. Seizure onset was 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.



Vignette

- 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].



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).

Disorders

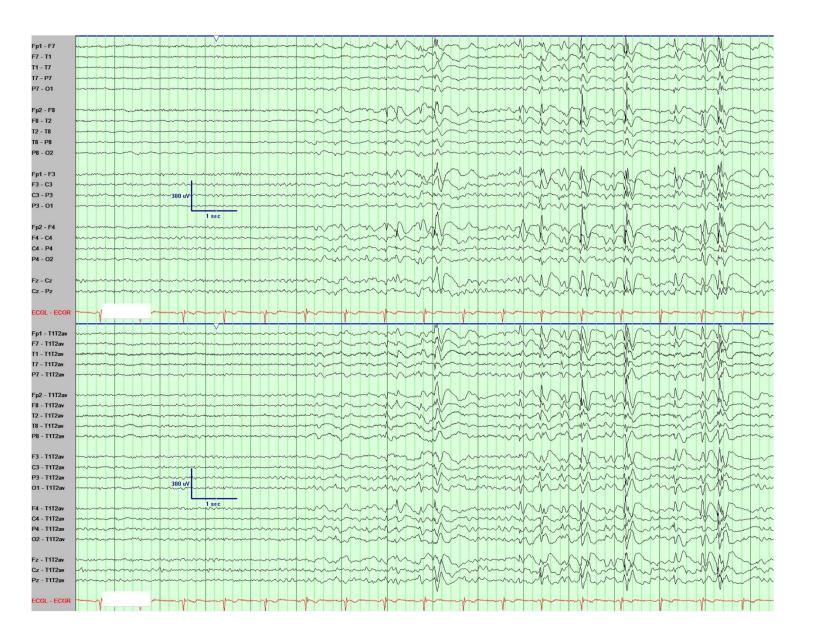


Figure 2

Epile

Routine EEG shows generalized paroxysmal fast activity: bipolar (upper panel) and referential (lower panel) montages show a 6-to-7second 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).

