

Teaching NeuroImage: Fasting EEG in Glucose Transporter-1 Deficiency Syndrome

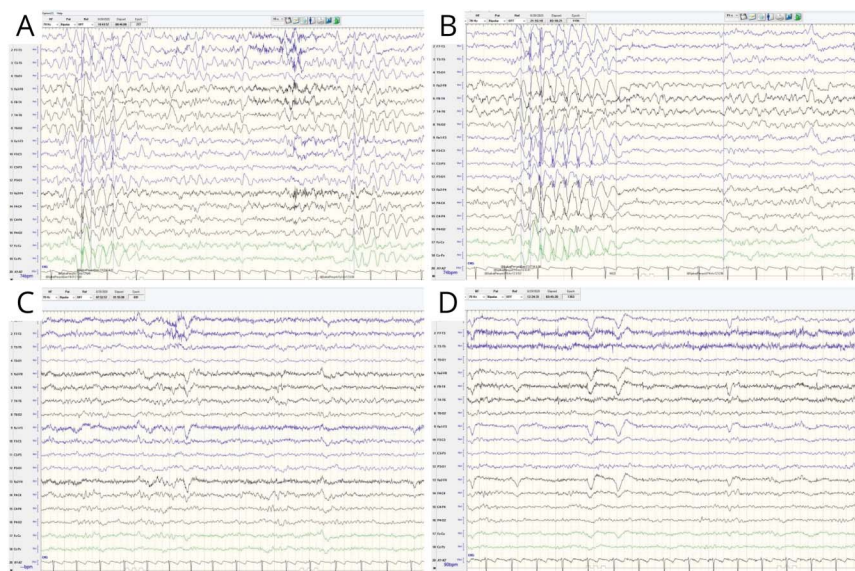
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Figure EEG During Fasting (A and B) Showing Diffuse, Poorly Formed 3–4 Hz Spike-Wave Discharges



EEG after meal (C and D) showing background normalization. This EEG is shown on a 15-second epoch with a sensitivity of 7 μ V in a longitudinal bipolar montage, left over right.

A 12-year-old boy presented for the evaluation of chorea induced by fasting and exercise. EEG, brain MRI, spectroscopy, and metabolic and microarray evaluations were unremarkable. Fasting EEG showed diffuse spike-wave discharges with postprandial normalization (Figure); chorea was not captured. Genetic testing confirmed a pathogenic *SLC2A1* variation consistent with glucose transporter-1 (GLUT-1) deficiency syndrome, which is uniquely responsive to ketogenic diet. EEG findings in this syndrome may include multifocal or generalized spikes, with postprandial improvement.^{1,2} This case suggests a supportive role for fasting EEG in the diagnosis of glucose transporter-1 deficiency syndrome, but the sensitivity and specificity remain unclear.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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Appendix Authors

Name	Location	Contribution
Hassan Imtiaz, MD	Wayne State University, Detroit, MI	Drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data; and analysis or interpretation of data
Afra Can, MD	Wayne State University, Detroit, MI	Drafting/revision of the article for content, including medical writing for content, and major role in the acquisition of data
Daniela Tapos, MD	Wayne State University, Detroit, MI	Drafting/revision of the article for content, including medical writing for content, and major role in the acquisition of data

Appendix *(continued)*

Name	Location	Contribution
Amanda Weber, DO	Wayne State University, Detroit, MI	Drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data; and analysis or interpretation of data

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