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

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A 12 year old male presented for evaluation of chorea induced by fasting and exercise. EEG, Brain MRI, spectroscopy, metabolic and microarray evaluations were unremarkable. Fasting EEG showed diffuse spike-wave discharges with postprandial normalization (Figure 1); chorea was not captured. Genetic testing confirmed a pathogenic SLC2A1 mutation 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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Ethical Compliance Statement:

The authors confirm that the approval of an institutional review board was not required for this work. We confirm that the informed consent was obtained from the parents. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Figure 1: EEG during fasting (A, B) showing diffuse, poorly-formed 3-4 Hz spike-wave discharges. EEG after meal (C, 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.

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