A 7-month-old boy with glutaric aciduria type 1 (GA1) presented with 1 week of clustered flexor spasms. Examination revealed mild axial hypotonia without encephalopathy. Video-EEG monitoring revealed hypsarrhythmia and infantile spasms (figure, A). MRI showed acute basal ganglia injury (figure, B). After 3 weeks of prednisolone treatment, 5-month follow-up showed continued resolution of hypsarrhythmia and spasms. GA1 is an autosomal recessive condition due to deficiency of the enzyme necessary for degradation of lysine, tryptophan, and hydroxylysine. Infantile spasms have only been reported once before in GA1. New-onset spasms may be associated with...
suboptimal metabolic control, even in the absence of encephalopathy.2

AUTHOR CONTRIBUTIONS
Nichole Young-Lin: participated in conceptualization of the manuscript, drafted the manuscript and figure legend, and revised the manuscript for intellectual content. Sarah Shalev: selected appropriate EEG images (i.e., interpretation of data) and revised the manuscript for intellectual content. Orit A. Glenn: selected appropriate brain MRI images (i.e., interpretation of data) and revised the manuscript for intellectual content. Marisa Gardner: participated in analysis of results and revised the manuscript for intellectual content. Chung Lee: revised the manuscript for intellectual content. Anthony Wynshaw-Boris: participated in conceptualization of the manuscript and revised the manuscript for intellectual content. Amy A. Gelfand: participated in conceptualization of the manuscript and revised the manuscript for intellectual content.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
N. Young-Lin reports no disclosures. S. Shalev reports no disclosures. O. Glenn reports no disclosures. M. Gardner reports no disclosures. C. Lee reports no disclosures. A. Wynshaw-Boris received grants from NIH/NINDS and NIH/NIGMS. A. Gelfand received research grant support from NIH/NINDS (K12NS001692) and the UCSF Clinical and Translational Science Institute; she has received personal compensation for legal consulting as well as honoraria from Journal Watch Neurology. Go to Neurology.org for full disclosures.

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Teaching NeuroImages: Infant with glutaric aciduria type 1 presenting with infantile spasms and hypsarrhythmia
Nichole Young-Lin, Sarah Shalev, Orit A. Glenn, et al.

Neurology 2013;81:e182-e183
DOI 10.1212/01.wnl.0000437291.75075.53

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