A 3-year-old girl presented with episodic ataxia for the past month. It lasted for 1–2 days continuously after a febrile illness. MRI was suggestive of a neurometabolic disorder (figure, A–L). Plasma valine and leucine and urinary branched-chain aminoacids were...

Patients with intermittent maple syrup urine disease (MSUD) may develop episodic decompensation during periods of stress secondary to acute leucinuria and abnormal neurotransmitter activity.\textsuperscript{1} Bilateral, symmetrical diffusion restriction in myelinated areas of the brain is seen in aminoacidopathies, including MSUD, nonketotic hyperglycinemia, and Canavan disease.\textsuperscript{2}

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

\section*{Appendix Authors}

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<tr>
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\section*{References}

Teaching NeuroImages: When MRI is a clue in episodic ataxia
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