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 hyperintense signal in (A) cerebellar deep white matter including nuclei (short white arrow) and dorsal pons (long white arrow), (B) globus pallidi (black arrow) and thalami, and (C) bilateral cerebral white matter (white arrow) in axial T2 scan. These changes are hypointense on T1 scan (D–F). Diffusion-weighted images (G–I, b = 1,000) and apparent diffusion coefficient (J–L) show acute diffusion restriction.
Clinical exome revealed a homozygous, missense, pathogenic variation in BCKDHB gene (exon 5, chr6: 80878686A>C; p.His191Pro).

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

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

**Appendix**

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**References**

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