

Teaching Video NeuroImage: Oculogyric Crises in a 12-Year-Old Girl With Rapid-Onset Dystonia Parkinsonism

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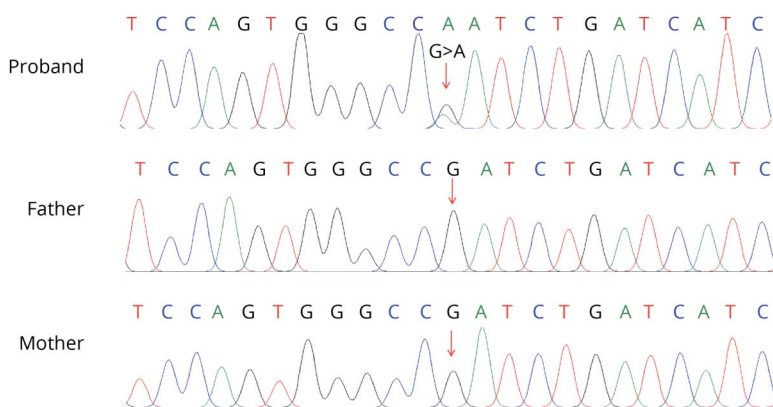
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Figure Electropherogram of the Patient and Her Parents



Electropherogram showed a heterozygous variant (c. 2767 G > A, p. Asp923Asn) in *ATP1A3* of the proband. Her parents did not carry this variant in *ATP1A3*.

A 12-year-old girl presented to the emergency department for forced upward eye deviation for 2 hours with craniocervical dystonia and intact consciousness. During the half-year, she experienced several emotionally triggered attacks (Video 1). Sudden-onset dystonic gait occurred 1 month before the first attack and progressively deteriorated with subsequent attacks. All limbs showed bradykinesia and rigidity without ataxia. Normal EEG and consciousness suggested oculogyric crises rather than epilepsy. A de novo pathogenic variant in *ATP1A3* (c. 2767 G > A) confirmed rapid-onset dystonia parkinsonism (Figure), which is not generally dopa-responsive.¹ However, since her oculogyric crises might correlate with a hypodopaminergic state,² levodopa was beneficial. Adding flunarizine improved her condition.

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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
Junyu Lin, MD	West China Hospital, Sichuan University	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; and study concept or design
Chunyu Li, MD	West China Hospital, Sichuan University	Drafting/revision of the manuscript for content, including medical writing for content

Appendix (continued)

Name	Location	Contribution
Huifang Shang, MD	West China Hospital, Sichuan University	Drafting/revision of the manuscript for content, including medical writing for content, and analysis or interpretation of data

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