A 6-year-old boy from Maharashtra, India, presented with subacute-onset generalized dystonia after a febrile illness; his sister manifested insidious-onset multifocal dystonia (Video 1). T2-weighted MRI showed striatal hyperintensities (Figure 1). Clinical exome sequencing detected a novel homozygous variant (c.127 G > T; G43C) in the \textit{PRKRA} gene which encodes PACT, a stress-response protein. The variant was confirmed by Sanger sequencing (eFigure 1, links.lww.com/WNL/C111). DYT-PRKRA is a childhood-onset progressive dystonia.\textsuperscript{1} Some patients have exacerbation with fever and evidence of striatal degeneration in neuroimaging.\textsuperscript{2} PACT-dependent activation of protein kinase R (PKR) leads to apoptosis.\textsuperscript{2} Perhaps, the G43C PACT triggered abnormal activation of PKR and intensified neuronal apoptosis during febrile illness.

**Study Funding**

No targeted funding reported.

**Disclosure**

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

**Publication History**

Received by \textit{Neurology} January 17, 2022. Accepted in final form May 2, 2022. Submitted and externally peer reviewed. The handling editor was José Merino, MD, MPhil, FAAN.
Appendix Authors

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suverit Subhas Bhowmick, MD</td>
<td>Movement Disorders Clinic, Vadodara Institute of Neurological Sciences, India</td>
<td>Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data</td>
</tr>
<tr>
<td>Sarbani Raha, MD</td>
<td>Child Neurology and Epilepsy Clinic, Vadodara, India</td>
<td>Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data</td>
</tr>
</tbody>
</table>

References


Call for Voices: Lived Experiences

The Editor of the *Neurology* specialty section Inclusion, Diversity, Equity, Anti-racism, & Social Justice (IDEAS) encourages you to submit short first-person accounts (1,000 words or less) of experiences lived within the realm of IDEAS with the goal of informing and enlightening our community on these critical issues. Some topics to consider include, but are not limited to:

- Descriptions of personal experiences that shaped your views of IDEAS.
- Reflections on the intersection between personal identity and career.
- Discussions at the intersection of IDEAS and neurology patient care, research, education, advocacy, or policy.

Submit your contributions to journal@neurology.org and include “Voices Submission” in the subject line.

Visit the *Neurology*® Resident & Fellow Website

Click on Residents & Fellows tab at Neurology.org.

Now offering:

- *Neurology* Resident & Fellow Editorial team information
- “Search by subcategory” option
- E-Pearl of the Week
- Direct links to Career Planning and AAN Resident & Fellow Pages
- Recently published Resident & Fellow articles
- Commentaries by Editors and Resident & Fellow team members

Find *Neurology* Residents & Fellows Section on Facebook: facebook.com/AANResidentsAndFellows

Follow *Neurology* on Twitter: @GreenJournal #NeurologyRF

Find *Neurology* Residents & Fellows Section on Instagram: @aanbrain #NeurologyRF
Early-Onset Dystonia, Exacerbation With Fever, and Striatal Signal Changes: Emerging Phenotype of DYT-PRKRA

Suvorit Subhas Bhowmick, Sarbani Raha and Amita Bohora

Neurology 2022;99:206-207 Published Online before print June 3, 2022
DOI 10.1212/WNL.0000000000200858

This information is current as of June 3, 2022

Updated Information & Services
including high resolution figures, can be found at:
http://n.neurology.org/content/99/5/206.full

References
This article cites 2 articles, 0 of which you can access for free at:
http://n.neurology.org/content/99/5/206.full#ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
All Movement Disorders
http://n.neurology.org/cgi/collection/all_movement_disorders
Basal ganglia
http://n.neurology.org/cgi/collection/basal_ganglia
Dystonia
http://n.neurology.org/cgi/collection/dystonia
MRI
http://n.neurology.org/cgi/collection/mri

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints
Information about ordering reprints can be found online:
http://n.neurology.org/subscribers/advertise