

In Response To:

Vynogradova I, Savitski V, Heckmann JG. Hemichorea associated with CASPR2 antibody. Tremor Other Hyperkinet Mov. 2014; 4. doi: 10.7916/D8VM49C5

Original Article:

Ramdhani RA, Frucht SJ. Isolated Chorea Associated with LGI1 Antibody. Tremor Other Hyperkinet Mov. 2014; 4. doi: 10.7916/D8MG7MFC

Letter to the Editor

Reply to: Hemichorea Associated with CASPR2 Antibody**Ritesh A. Ramdhani^{1*}, Steven J. Frucht¹**¹ Movement Disorders Division, Icahn School of Medicine at Mount Sinai, New York, New York, United States of America**Keywords:** Choreatic syndrome, chorea, hemichorea, paraneoplastic syndrome, CASPR2 antibody**Citation:** Ramdhani RA, Frucht SJ. Reply to: hemichorea associated with CASPR2 antibody. Tremor Other Hyperkinet Mov. 2014; 4. doi: 10.7916/D8QV3JM0

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Editor: Elan D. Louis, Columbia University, United States of America**Received:** April 17, 2014 **Accepted:** May 01, 2014 **Published:** May 19, 2014**Copyright:** © 2014 Ramdhani et al. This is an open-access article distributed under the terms of the Creative Commons Attribution–Noncommercial–No Derivatives License, which permits the user to copy, distribute, and transmit the work provided that the original author(s) and source are credited; that no commercial use is made of the work; and that the work is not altered or transformed.**Funding:** None.**Financial Disclosures:** None.**Conflict of Interest:** The authors report no conflict of interest.

To the editor:

The case presented by Vynogradova and colleagues adds to the ever-expanding etiological spectrum of autoimmune chorea. This is the first reported case of isolated paraneoplastic chorea associated with anti-CASPR2, paralleling our report of isolated idiopathic chorea associated with anti-LGI1. Both cases share an asymmetric predominance of the chorea as well as the absence of limbic encephalitis. LGI1 and CASPR2 antibodies are specific to the VGKC-protein complex and each are associated with a number of distinct clinical phenotypes: Faciobrachial dystonic seizures (FBDS), hyponatremia and limbic encephalitis (LE) in the former; and Morvan's syndrome¹, peripheral nerve excitability, and Isaac's syndrome² in the latter. Despite their phenotypic differences, these two cases highlight that phenotypically-similar late-onset isolated chorea can be a result of distinct pathophysiological mechanisms which will result in a different clinical course. This is illustrated by the fact that, five months after having a complete remission following pulse steroid therapy, our patient's chorea relapsed. A second course of steroids

did not engender as robust a response as the initial treatment. Seizure, metabolic disturbances, and LE remain absent and a steroid-sparing therapy (i.e., rituximab, IVIG) will be considered next. These cases emphasize the importance of antibody screening in late-onset chorea due to the diagnostic and treatment implications.

References

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2. Panzer J, Dalmau J. Movement disorders in paraneoplastic and autoimmune disease. *Curr Opin Neurol* 2011;24:346-353.