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LETTER TO THE EDITOR

Response to comment to "Magnetic resonance imaging findings of bilateral thalamic involvement in severe paroxysmal sympathetic hyperactivity: a pediatric case series"

Serge Mrkobrada[1]

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Dear Editor:

Dr. Shukla's comment points out limitations of our study [1], including the small sample size of three patients. We recognize this limitation within our manuscript and agree that larger cohorts are needed to prove our hypothesis of bilateral thalamic involvement in severe paroxysmal hyperactivity (PSH) in patients with hypoxemic brain injury.

We also agree that the pathophysiology of PSH is very complex. We do not argue that the *only* neuroanatomical basis for developing severe PSH is bilateral thalamic involvement, and there may be others, as pointed out by Dr. Shukla. However, in severe hypoxemic brain injury, our case series suggests that bilateral thalamic involvement may be an important marker of severe PSH. To support our hypothesis, we outlined both studies on animal models as well as fatal familial insomnia that have implicated the role of the thalamus in autonomic control [2–4]. Interestingly, the clinical manifestations of fatal familial insomnia are similar to PSH, a disease that is characterized by severe degeneration of bilateral mediodorsal and anterior thalamic nuclei with sparing of other autonomic centers [2, 3]. In publishing this hypothesis, it was our hope to stimulate further research and discussion of PSH, which we are pleased to see, has already happened.

Compliance with ethical standards

Conflict of interest The author declares that there are no conflicts of interest.

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✉ Serge Mrkobrada
mrkobrada@gmail.com

[1] Calgary, AB, Canada