Teaching Video NeurolImages: Needle-like central spikes evoked by hand tapping in Rett syndrome

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Figure EEG of the reported case

(A) There were runs of needle-like right and left central spikes independently or synchronously (initially lower amplitude and then higher amplitude) with bilateral hand tapping (note the bilateral deltoid EMG channels). (B) There was marked activation of these spikes during non-REM sleep without clinical signs. One such spike (in red box) is shown in referential montage (C) and on increased time base in (D), to demonstrate the relation between EMG and EEG spikes. The cursor (D) shows the onset of bilateral central spikes at C3 and C4. The latency between the left EMG spike (arrow) produced by hand tapping and the onset of C3 and C4 spikes was around 30 ms, which may be consistent with somatosensory cortical evoked response. (Referential montage [system reference O₂], sensitivity 10 μ V/mm, low-frequency filter 1 Hz, high-frequency filter 70 Hz, sampling rate 256 Hz).

A 4-year-old-girl with Rett syndrome (RS) presented with drug-resistant epilepsy since 4 years of age. Her video-EEG captured rhythmic hand tapping with bilateral central spike-and-waves (figure and video 1). Characteristic EEG features in RS include prominent central rhythmic theta activity during wakefulness and needle-like central spikes evoked by contralateral passive finger-tapping. These central spikes may represent exaggerated somatosensory evoked potential secondary to the presence of hyperexcitable motor and sensory cortices. Reflex seizures (focal and atonic) provoked by rhythmic hand pressure have been described in 2 children with

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RS.¹ In contrast, hand clapping has also been reported to suppress continuous centrottemporal spikes.² Thus, the clinical significance of these provoked central spikes is unclear.

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Author	Location	Role	Contribution
Pratima Gulati	The Hospital for Sick Children, Toronto, Canada	Author	Provided clinical care to the patient, analyzed the data, drafted the manuscript for intellectual content
Puneet Jain	The Hospital for Sick Children, Toronto, Canada	Author	Provided clinical care to the patient, analyzed the data, drafted the manuscript for intellectual content

Appendix (continued)

Author	Location	Role	Contribution
Felippe Borlot	The Hospital for Sick Children, Toronto, Canada	Author	Provided clinical care to the patient, analyzed the data
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Ayako Ochi	The Hospital for Sick Children, Toronto, Canada	Author	Provided clinical care to the patient, analyzed the data, interpreted the data, revised the manuscript for intellectual content

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