Teaching NeuroImages:

A central theta EEG rhythm in Rett syndrome can masquerade as seizure

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Figure Rhythmic frontocentral theta in a patient with Rett syndrome

(A) Electroencephalography demonstrated a prominent 50–100 μ V 4–5 Hz rhythmic, fluctuating central theta rhythm (*) during wakefulness. (B) After tactile stimulation (line), a blocking phenomenon occurred (arrow), during which the central theta rhythm abated. All data are presented in longitudinal bipolar montage, sensitivity 7 μ V/mm.

A 12-year-old girl with Rett syndrome presented for characterization of staring spells. Paroxysmal runs of fluctuating 4- to 5-Hz rhythmic theta activity maximal in the frontocentral electrodes occurred during periods of relative inactivity and abated with movement or tactile stimulation (figure). This patient's

electrographic phenomenon had been previously interpreted as potential seizure. A prominent central theta rhythm in individuals with Rett syndrome was previously described as a slow mu rhythm equivalent, which is supported by the blocking phenomenon.^{1,2} Recognition of this phenomenon may forestall

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misdiagnosis of this fluctuating rhythmic pattern during clinical inactivity as an ictal pattern.

AUTHOR CONTRIBUTIONS

Dr. Amanda Weber was involved with the conceptualization of the study and in drafting and revising the manuscript. Dr. Adam Ostendorf was involved with the conceptualization of the study and in revising the manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

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

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Neurology 2016;87:e29-e30

DOI 10.1212/WNL.0000000000002866

This information is current as of July 18, 2016

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