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Teaching Neurolmages: Notched Delta and Angelman Syndrome

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Case Description

We report a 9-year-old boy with a history of Angelman syndrome (AS) with associated epilepsy, behavioral issues, intellectual disability, and sleep disturbance. Genetic testing revealed a methylation imprinting defect, thereby confirming the diagnosis of AS. His seizures were well controlled on monotherapy with clobazam and he underwent a routine EEG (**figure**), which showed a notched delta pattern.

Notched delta pattern, one of the characteristic EEG findings in AS, is characterized by delta waves intermixed with spikes or sharp waves giving it a “notched” appearance. In the context of a suggestive phenotype, this EEG pattern should raise suspicion for AS. Clinicians should be reminded, nonetheless, that notched delta can be seen in other conditions such as Rett syndrome and different chromosomal abnormalities including 4p(-) syndrome. Lastly, patients with AS may have other patterns on EEG such as persistent rhythmic high-voltage 4-6 Hz activity. [1,2,3]

Appendix 1. Authors

Name	Location	Contribution
Fábio A. Nascimento, MD	Massachusetts General Hospital, Boston, MA	Conceptualized and designed study, analyzed and interpreted data, drafted manuscript.
Elizabeth A. Thiele, MD, PhD	Massachusetts General Hospital, Boston, MA	Conceptualized and designed study, analyzed and interpreted data, reviewed manuscript, supervised study.

Ron L. Thibert, DO, MsPH	Massachusetts General Hospital, Boston, MA	Conceptualized and designed study, analyzed and interpreted data, reviewed manuscript, supervised study.
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Teaching Slides -- <http://links.lww.com/WNL/B425>

References

- [1] Korff CM, Kelley KR, Nordli Jr DR. Notched delta, phenotype, and Angelman syndrome. J Clin Neurophysiol. 2005;22(4):238-243.
- [2] Thibert RL, Larson AM, Hsieh DT, Raby AR, Thiele EA. Neurologic manifestations of Angelman syndrome. Pediatr Neurol. 2013;48(4):271-279.
- [3] Vendrame M, Loddenkemper T, Zarowski M, et al. Analysis of EEG patterns and genotypes in patients with Angelman syndrome. Epilepsy Behav. 2012 Mar;23(3):261-265.

Figure

Title: Routine EEG shows notched delta pattern [sensitivity 20 $\mu\text{V}/\text{mm}$, LF 1 Hz, HFF 70 Hz, notch on/60 Hz, timebase 30 mm/sec].

Legend: EEG: bipolar (A) and referential/average (B) show bilateral occipital, right more than left, delta waves (200-250 μV , 4 Hz) superimposed with spikes (arrows) consistent with notched delta.

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Teaching NeuroImages: Notched Delta and Angelman Syndrome

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