

E-Pearl of the Week

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Paraspinal myotonic discharges in acid maltase deficiency

Acid maltase deficiency is an autosomal recessive glycogen storage disorder that can present in adults with slowly progressive limb-girdle weakness. These patients often have chronic respiratory insufficiency as well. On needle electromyography a majority of these patients have myopathic changes in the limbs-- increased insertional activity, irritability, and fibrillations. Some will have myotonic discharges found exclusively in the paraspinal musculature in the absence of clinical myotonia. The savvy electrodiagnostician will not omit needle examination of the paraspinal muscles in adult patients with slowly progressive limb-girdle weakness and myopathic EMG abnormalities in the limbs.

Barohn RJ, McVey AL, Dimauro S. Adult acid maltase deficiency. *Muscle & Nerve* 1993;16:672-676.

Submitted by Ryan Overman, MD and Robert Pascuzzi, MD.

Disclosure: Dr. Overman serves on the Neurology Resident and Fellow Section editorial team and the Neurology Podcast Committee.

Dr. Pascuzzi served on the Board of Directors of the ABPN for the past eight years and has been involved with the ABPN decisions to move the two hours of exam vignettes into an enhanced single certifying examination and move the assessment of basic clinical skills into the residency period. Also served on the Neurology RRC the past six years and been involved with the development of new program requirements for neurology residency. I am the immediate past Program Director for Neurology Residency Training at Indiana University. I volunteered to be one of the evaluators in the current study and thus participated as an ABPN examiner (although ironically as a Director of the ABPN I have not directly examined any candidate for the past 8 years). I did not request nor accept the honorarium offered for participation in this study. I participate in multicenter clinical trials for a variety of neuromuscular disorders but receive no direct compensation. I lecture for the AAN and at multiple academic medical centers but take no personal compensation (any honoraria go directly to my institution).

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