Teaching Video NeuroImages: Trapezius myotonia percussion sign in myotonic dystrophy type 2

Myotonic dystrophy type 2 (DM2) is an autosomal dominant disorder with proximal weakness, muscle pain, and early-onset cataracts. In comparison with myotonic dystrophy type 1 (DM1), myotonia is less symptomatic, more proximal, and harder to detect during clinical and electrodiagnostic testing. Here we document the presence of trapezius myotonia in patients with DM2 (video on the Neurology® Web site at www.neurology.org). In our experience, similar proximal percussion does not produce as marked a response in DM1 or nondystrophic myotonic disorders. This sign demonstrates a mechanism to test for proximal myotonia, and in at-risk patients may be suggestive of an underlying diagnosis of DM2.

AUTHOR CONTRIBUTIONS
Nicholas E. Johnson: drafting manuscript, data collection. Chad R. Heatwole: revision of manuscript, design of project.

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REFERENCES
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