Teaching Video NeuroImages:

Lid lag sign and diplopia in paramyotonia congenita

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Figure Lid lag sign

Lid lag sign as a result of prolonged muscle relaxation (myotonia) after voluntary contraction of the upper eyelids (sustained upward gaze for a few seconds) photographed directly after immediate downgaze.

Clinical bedside tests to provoke myotonia, such as eyelid closure, handgrip, and percussion myotonia, help to diagnose and differentiate myotonic disorders and prevent delayed genetic confirmation. Sodium channelopathies present with predominantly myotonia of the ocular muscles.¹ In these cases, testing for myotonia of the upper eyelid and extraocular muscles could be of decisive diagnostic value. We present additional clinical bedside tests: the lid lag sign² and provocation of short-term diplopia (video on the *Neurology*[®] Web site at Neurology.org and the figure). In our experience, these symptoms are especially present in paramyotonia congenita. Therefore, we advise to test for lid lag sign and short-term diplopia every time a myotonic disorder is suspected.

AUTHOR CONTRIBUTIONS

Dr. Stunnenberg: manuscript writing, clinical history taking, neurologic examination, and collection of videoimages, all under the supervision of Dr. Drost.

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DISCLOSURE

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

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