

Teaching Video NeuroImages: Lid lag sign and diplopia in paramyotonia congenita

Bas C. Stunnenberg, MD
Gea Drost, MD, PhDCorrespondence to
Dr. Drost:
G.Drost@umcg.nl**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.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

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

REFERENCES

1. Trip J, Drost G, Ginjaar HB, et al. Redefining the clinical phenotypes of non-dystrophic myotonic syndromes. *J Neurol Neurosurg Psychiatry* 2009;80:647–652.
2. Harvey JT, Anderson RL. Lid lag and lagophthalmos: a clarification of terminology. *Ophthalmic Surg* 1981;12:338–340.

Supplemental data
at Neurology.orgDownload teaching slides:
Neurology.org

From Donders Institute for Brain, Cognition and Behaviour (B.C.S.), Department of Neurology, Radboud University Nijmegen Medical Centre; and the Department of Neurology and the Department of Neurosurgery (G.D.), University Medical Center Groningen, University of Groningen, the Netherlands.

Neurology®

Teaching Video *NeuroImages*: Lid lag sign and diplopia in paramyotonia congenita

Bas C. Stunnenberg and Gea Drost

Neurology 2014;83:e68

DOI 10.1212/WNL.0000000000000649

This information is current as of July 28, 2014

| | |
|---|--|
| Updated Information & Services | including high resolution figures, can be found at: http://n.neurology.org/content/83/5/e68.full |
| Supplementary Material | Supplementary material can be found at: http://n.neurology.org/content/suppl/2014/07/26/83.5.e68.DC1 http://n.neurology.org/content/suppl/2014/07/26/83.5.e68.DC2 |
| References | This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/83/5/e68.full#ref-list-1 |
| Subspecialty Collections | This article, along with others on similar topics, appears in the following collection(s): Clinical neurology examination http://n.neurology.org/cgi/collection/clinical_neurology_examination Diplopia (double vision) http://n.neurology.org/cgi/collection/diplopia_double_vision Ion channel gene defects http://n.neurology.org/cgi/collection/ion_channel_gene_defects |
| Permissions & Licensing | Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions |
| Reprints | Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise |

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2014 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

