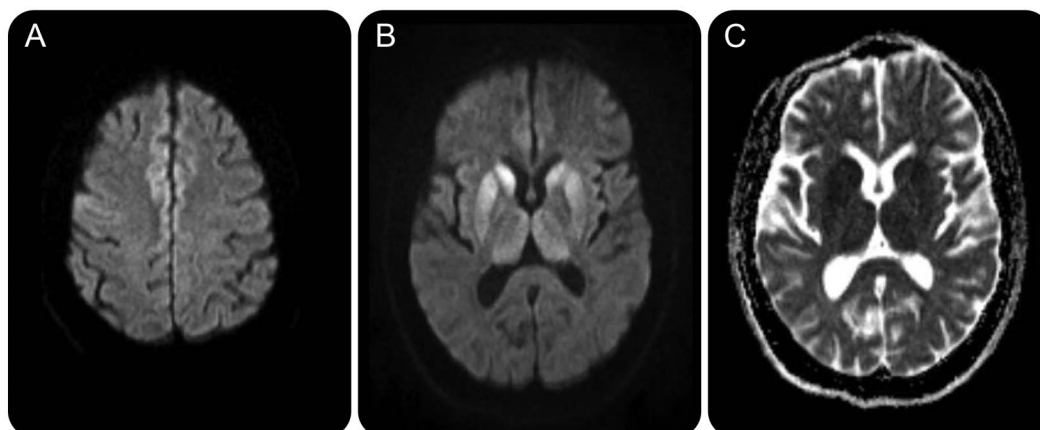


# Supranuclear gaze palsy and horizontal ocular oscillations in Creutzfeldt-Jakob disease



**Figure** MRI diagnostic of Creutzfeldt-Jakob disease



MRI brain diffusion-weighted imaging with hyperintense cortical gyri (A), basal ganglia and pulvinar diffusion restriction (B), with apparent diffusion coefficient dropout (C).

A 59-year-old patient presented with marked cognitive impairment. Examination revealed supranuclear gaze palsy and ocular oscillations (video at [Neurology.org](http://Neurology.org)). Extremity and axial muscle tone was increased, accompanied by full body tremulousness suggestive of polymyoclonus. Motor abnormalities had progressed over 3 months but cognitive problems developed in 1–2 weeks. MRI (figure) and CSF (positive real-time quaking-induced conversion) were diagnostic of Creutzfeldt-Jacob disease (CJD).

Supranuclear ophthalmoplegia is a known feature of CJD but when combined with parkinsonism can lead to a misdiagnosis of progressive supranuclear palsy.<sup>1</sup> Pendular oscillations, not previously described in CJD, are typically due to failure of brainstem eye movement neural integrators.<sup>2</sup>

*Asya I. Wallach, MD, Hannah Park, MD, Janet C. Rucker, MD, Horacio Kaufmann, MD*

From NYU School of Medicine and Bellevue Hospital, New York.

*Author contributions:* Asya Wallach: clinical care of the patient, study concept, acquisition of data, writing the manuscript. Hannah Park: clinical care of the patient, acquisition of data. Janet Rucker: acquisition of data, analysis and interpretation of data, study supervision, critical revision of the manuscript for intellectual content. Horacio Kaufmann: study concept and supervision, critical revision of the manuscript for intellectual content.

*Study funding:* Supported in part by funding from the NIH (U54NS065736 and 1U01NS078025-01).

*Disclosure:* The authors report no disclosures relevant to the manuscript. Go to [Neurology.org](http://Neurology.org) for full disclosures.

*Correspondence to Dr. Wallach:* [Asya.Wallach@nyumc.org](mailto:Asya.Wallach@nyumc.org)

1. Petrovic IN, Martin-Bastida A, Massey L, et al. MM2 subtype of sporadic Creutzfeldt-Jacob disease may underlie the clinical presentation of progressive supranuclear palsy. *J Neurol* 2013;260:1031–1036.
2. Gresty MA, Ell JJ, Findley LJ. Acquired pendular nystagmus: its characteristics, localising value and pathophysiology. *J Neurol Neurosurg Psychiatry* 1982;45:431–439.

Supplemental data  
at [Neurology.org](http://Neurology.org)

# Neurology®

## Supranuclear gaze palsy and horizontal ocular oscillations in Creutzfeldt-Jakob disease

Asya I. Wallach, Hannah Park, Janet C. Rucker, et al.

*Neurology* 2017;89;749

DOI 10.1212/WNL.0000000000004242

**This information is current as of August 14, 2017**

<b>Updated Information &amp; Services</b>	including high resolution figures, can be found at: <a href="http://n.neurology.org/content/89/7/749.full">http://n.neurology.org/content/89/7/749.full</a>
<b>Supplementary Material</b>	Supplementary material can be found at: <a href="http://n.neurology.org/content/suppl/2017/08/14/WNL.0000000000004242.DC1">http://n.neurology.org/content/suppl/2017/08/14/WNL.0000000000004242.DC1</a>
<b>References</b>	This article cites 2 articles, 1 of which you can access for free at: <a href="http://n.neurology.org/content/89/7/749.full#ref-list-1">http://n.neurology.org/content/89/7/749.full#ref-list-1</a>
<b>Subspecialty Collections</b>	This article, along with others on similar topics, appears in the following collection(s): <b>All Neuro-ophthalmology</b> <a href="http://n.neurology.org/cgi/collection/all_neuroophthalmology">http://n.neurology.org/cgi/collection/all_neuroophthalmology</a> <b>Assessment of cognitive disorders/dementia</b> <a href="http://n.neurology.org/cgi/collection/assessment_of_cognitive_disorders_dementia">http://n.neurology.org/cgi/collection/assessment_of_cognitive_disorders_dementia</a> <b>Nystagmus</b> <a href="http://n.neurology.org/cgi/collection/nystagmus">http://n.neurology.org/cgi/collection/nystagmus</a> <b>Prion disease; see Infections/prion</b> <a href="http://n.neurology.org/cgi/collection/prion_disease">http://n.neurology.org/cgi/collection/prion_disease</a> <b>Prognosis</b> <a href="http://n.neurology.org/cgi/collection/prognosis">http://n.neurology.org/cgi/collection/prognosis</a>
<b>Permissions &amp; Licensing</b>	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: <a href="http://www.neurology.org/about/about_the_journal#permissions">http://www.neurology.org/about/about_the_journal#permissions</a>
<b>Reprints</b>	Information about ordering reprints can be found online: <a href="http://n.neurology.org/subscribers/advertise">http://n.neurology.org/subscribers/advertise</a>

*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 © 2017 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

