The pivotal sign of CANVAS

A 75-year-old woman complained about insecure gait since age 55. Clinical examination revealed signs of cerebellar ataxia, bilateral vestibulopathy, and peripheral sensory impairment. Sensory nerve action potentials were absent. The visually enhanced vestibulo-ocular reflex (VVOR) was impaired (video on the Neurology® Web site at www.neurology.org, figure 1) and the diagnosis of cerebellar ataxia (figure 2) with neuropathy and bilateral vestibular areflexia syndrome (CANVAS) was made. CANVAS is considered to be a recessive disorder with a mean age at onset of 60 years. VVOR impairment is its characteristic clinical sign. It can only be elicited if both smooth-pursuit eye movements and the vestibulo-ocular reflex are deficient. Normally, both are redundant at low head velocities.

Jens Alexander Petersen, MD, Werner W. Wichmann, MD, Konrad Peter Weber, MD

From the Departments of Neurology (J.A.P., K.P.W.), Neuroradiology (W.W.W.), and Ophthalmology (K.P.W.), University Hospital Zurich, Switzerland.

Author contributions: J.A. Petersen recorded the video and wrote the manuscript. W. Wichmann acquired the MRI and revised the manuscript. K.P. Weber designed the study, acquired the eye movement data, and revised the manuscript.

Study funding: Supported by the Betty and David Koetser Foundation for Brain Research.
Figure 2  T2-weighted sagittal MRI illustrates the cerebellar atrophy in CANVAS

Marked cerebellar atrophy involves the vermal lobules VI, VIIa, and VIIb. Note also the spinal atrophy. CANVAS = cerebellar ataxia with neuropathy and bilateral vestibular areflexia syndrome.

Disclosures: J. Petersen and W. Wichmann report no disclosures. K. Weber acts as an unpaid consultant and has received funding for travel from GN Orometrics. Go to Neurology.org for full disclosures.

Correspondence to Dr. Weber: konrad.weber@usz.ch


NeuroImages Are Free at www.neurology.org!

All Neurology® NeuroImages can now be freely accessed on the Neurology Web site. See them at www.neurology.org, where you can also sign up for journal email alerts and check out other online features, including the Resident & Fellow section, Neurology: Clinical Practice, and the weekly Neurology Podcasts.
The pivotal sign of CANVAS
Jens Alexander Petersen, Werner W. Wichmann and Konrad Peter Weber
Neurology 2013;81;1642-1643
DOI 10.1212/WNL.0b013e3182a9f435

This information is current as of October 28, 2013

Updated Information & Services
including high resolution figures, can be found at:
http://www.neurology.org/content/81/18/1642.full.html

Supplementary Material
Supplementary material can be found at:
http://www.neurology.org/content/suppl/2013/10/26/81.18.1642.DC1

References
This article cites 2 articles, 1 of which you can access for free at:
http://www.neurology.org/content/81/18/1642.full.html##ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Gait disorders/ataxia
http://www.neurology.org/cgi/collection/gait_disorders_ataxia

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/misc/about.xhtml#permissions

Reprints
Information about ordering reprints can be found online:
http://www.neurology.org/misc/addir.xhtml#reprintsus