

Teaching Video NeuroImages: The Beevor sign in late-onset Pompe disease



Matteo Garibaldi, MD
Jordi Diaz-Manera, MD,
PhD
Eduard Gallardo, PhD
Giovanni Antonini, MD

Correspondence to
Dr. Garibaldi:
matteo.garibaldi@uniroma1.it

Figure 1 The Beevor sign in late-onset Pompe disease



Still image from the video shows a Beevor sign in a 45-year-old man with late-onset Pompe disease. The Beevor sign is due to a weakness of the caudal part of the rectus abdominis muscle with relative sparing of the cranial part.

The Beevor sign, an upward deflection of the umbilicus on flexion of the neck, is a characteristic finding in facioscapulohumeral muscular dystrophy.¹ Many other neuromuscular disorders involving axial muscles can present a Beevor sign.² We report a 45-year-old man with late-onset Pompe disease showing a major Beevor sign (figure 1 and video on the *Neurology*[®] Web site at Neurology.org). He had progressive limb-girdle weakness that started in his 20s and severe axial weakness. Whole-body muscle MRI showed a complete fatty replacement and atrophy of the lower part of rectus abdominis (figure 2, arrowheads) and a milder involvement of the upper part (figure 2, arrows).

AUTHOR CONTRIBUTIONS

Dr. Garibaldi: design of the study, analysis and interpretation of the data, drafting manuscript, multimedia files creation. Dr. Diaz-Manera: analysis

and interpretation of the data, revising the manuscript. Dr. Gallardo: analysis and interpretation of the data, revising the manuscript. Dr. Antonini: analysis and interpretation of the data, revising the manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

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

REFERENCES

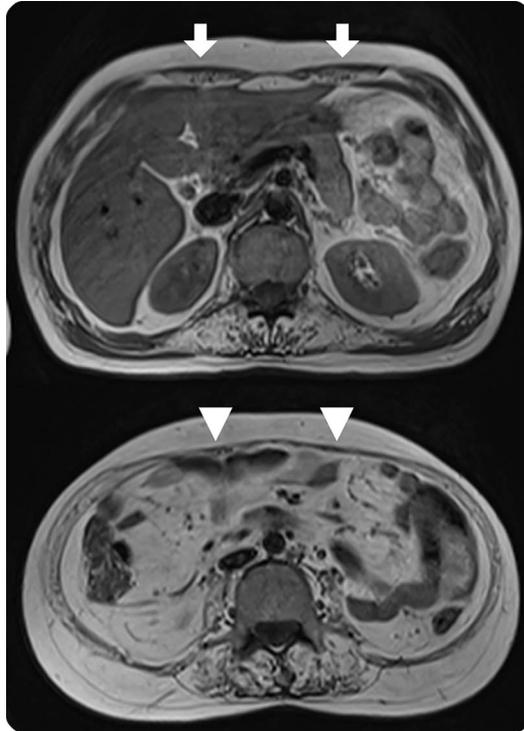
1. Awerbuch GI, Nigro MA, Wishnow R. Beevor's sign and facioscapulohumeral dystrophy. *Arch Neurol* 1990;47:1208–1209.
2. Shahrizaila N, Wills AJ. Significance of Beevor's sign in facioscapulohumeral dystrophy and other neuromuscular diseases. *J Neurol Neurosurg Psychiatry* 2005;76:869–870.

Supplemental data
at Neurology.org

Download teaching slides:
Neurology.org

From the Unit of Neuromuscular Diseases (M.G., G.A.), Department of Neurology, Mental Health and Sensory Organs (NESMOS), Faculty of Medicine and Psychology, "Sapienza" University of Rome, Italy; Laboratori de Malalties Neuromusculars (J.D.-M., E.G.), Institut de Recerca de HSCSP, Universitat Autònoma de Barcelona (UAB), Barcelona; and Centro de Investigación Biomédica en Red de Enfermedades Neurodegenerativas (CIBERNED) (J.D.-M., E.G.), Madrid, Spain.

Figure 2 Muscle MRI shows a gradient involvement of rectus abdominis



Two different slides from whole-body muscle MRI (T1-weighted) show the cranio-caudal gradient of fatty replacement in the rectus abdominis muscle, which is more mildly involved above the umbilicus (arrows). Rectus abdominis is completely replaced and atrophic below the umbilicus (arrowheads).

Neurology[®]

Teaching Video NeuroImages: The Beevor sign in late-onset Pompe disease

Matteo Garibaldi, Jordi Diaz-Manera, Eduard Gallardo, et al.

Neurology 2016;86:e250-e251

DOI 10.1212/WNL.0000000000002772

This information is current as of June 13, 2016

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 © 2016 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.



Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/86/24/e250.full
Supplementary Material	Supplementary material can be found at: http://n.neurology.org/content/suppl/2016/06/11/WNL.0000000000002772.DC1 http://n.neurology.org/content/suppl/2016/06/11/WNL.0000000000002772.DC2
References	This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/86/24/e250.full#ref-list-1
Citations	This article has been cited by 1 HighWire-hosted articles: http://n.neurology.org/content/86/24/e250.full##otherarticles
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Clinical Neurology http://n.neurology.org/cgi/collection/all_clinical_neurology All Neuromuscular Disease http://n.neurology.org/cgi/collection/all_neuromuscular_disease Clinical neurology examination http://n.neurology.org/cgi/collection/clinical_neurology_examination Muscle disease http://n.neurology.org/cgi/collection/muscle_disease
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 © 2016 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

