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

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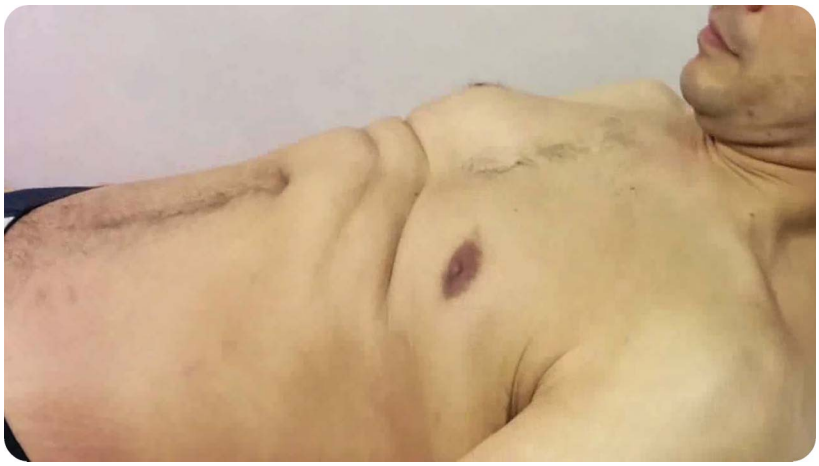
Figure 1 The Beevor sign in late-onset Pompe disease



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.

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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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Supplemental data at Neurology.org

Download teaching slides: Neurology.org

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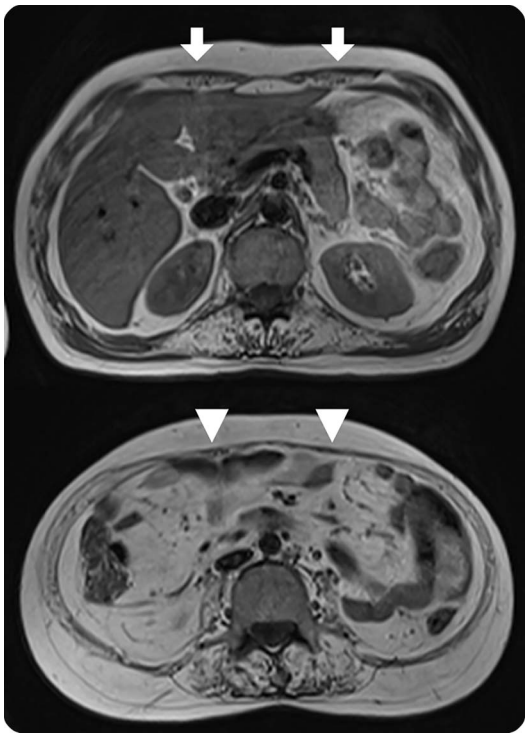
Figure 2 Muscle MRI shows a gradient involvement of rectus abdominis



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).

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