

Images in...

# Muscle weakness and a sustained handgrip (a video demonstration)

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## DESCRIPTION

A 52-year-old male farmer presented to us with a history of gradual weakness of his lower limb with initial difficulty in getting up from sitting that progressed to difficulty in combing his hair.

On examination he was found to have frontal baldness, with mild temporal wasting and MRC grade III/V power in his shoulders and hips with inability to raise the hands against resistance and inability to get up from squatting.

His younger brother came to the hospital to pay him a visit and we noticed he had similar features although with more prominent temporal wasting and frontal baldness. There are four brothers; the elder two did not have any hair loss or wasting or weakness.

The diagnosis was further established clinically on testing their handgrip (videos 1A and B).

**Video 1 (A, B)** Shaking hands - note patient's difficulty in release 10.1136/bcr.04.2009.1819v1a  
10.1136/bcr.04.2009.1819v1b

This was a classical presentation and the diagnosis was made clinically. The key clinical diagnostic feature (as also shown in the videos) is a delayed relaxation of muscle after sudden forceful contraction.<sup>1</sup> The differential

diagnosis of myotonic dystrophy includes other causes of myotonia: paramyotonia, congenital myotonia, mild tetanus and the rare stiff man syndrome. At later stages, the myotonic dystrophy may resemble limb-girdle atrophy, polymyositis or dermatomyositis.<sup>2</sup> DNA testing is definitive for the diagnosis and before it became available the electromyography was used and often showed a typical myotonic discharge with a waxing and waning quality giving rise to the descriptive term, the 'divebomber' sound. In myotonia, abnormalities in specific sarcolemmal ion conductances can lead to a reduced electrical threshold for firing action potentials and in conduction of repetitive impulses that result in sustained muscle fibre contraction.<sup>3</sup>

**Competing interests** None.

**Patient consent** Obtained.

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