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Unbalance in Myotonic Dystrophy-1 may follow cervical ataxia and respond to exercise

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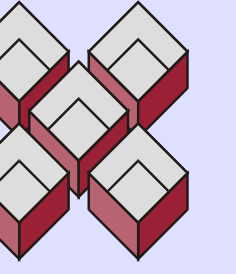
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Patients with myotonic dystrophy-type 1 (DM-1) often fall, due to “intrinsic” mechanisms (eg legs “giving out”) for unknown reasons (Wiles CM.JNNP 2006;77:393-396). A case is presented, in which neck position sense was impaired. DA was a woman, 57 yrs old, employed. DM-1 was diagnosed at age 35. She had fallen 12 times in 2 months. She felt unsteady (DHI_{sf} inventory 7/13, normal=13/13). Strength was 4/5 at the lower limbs and on the neck extensors, 2 to 4/5 at the upper limbs, 0 to 2/5 on the neck flexors. Neck position sense was tested through a goniometric helmet. The head was placed at target angles of 20% or 70% of the full range of motion, in opposite directions of yaw, pitch or roll, and then replaced to neutral. Repositioning was asked for 6 times for each target, in a balanced design. Overall error (% of target) was +29±45 (normal <5±5). On force platforms, stability in standing was low (EquiTest[®] SOT score 33%, normal >70); the capacity to lean around towards the limits of stability, LOS, was also low (Balance Master[®] LOS 57%, normal >85). Eleven 1-hr exercise sessions were held in 6 days, based on hands-on strenghtening of the neck and the shoulder girdle. The DHI_{sf} score rose to 11/13. Head repositioning errors declined to +17±24. The SOT rose to 56 and the LOS rose to 85. During the following 3 months the patient reported no falls. Neck position sense is crucial to balance (Brandt T. JNNP 2001 ;71:8-22) and it relies upon muscle spindles. In DM-1 severe alterations of neck spindles have been demonstrated (Swash M. Clin Neuropathol 1983;2,2:75-783). The case suggests that this may lead to unbalance in DM-1, and that a short program of neck strenghtening may be beneficial.

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Patients with myotonic dystrophy-type 1 often **fall** for unknown reasons.
(Wiles CM. *J Neurol Neurosurg Psychiatry* 2006;77(3):393-396)

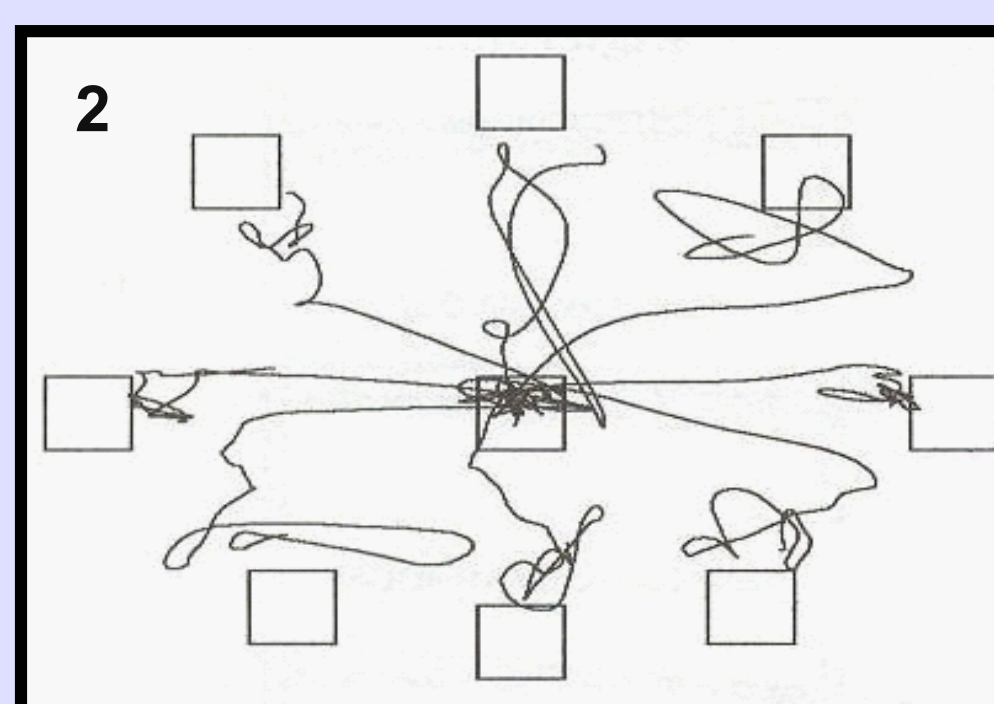
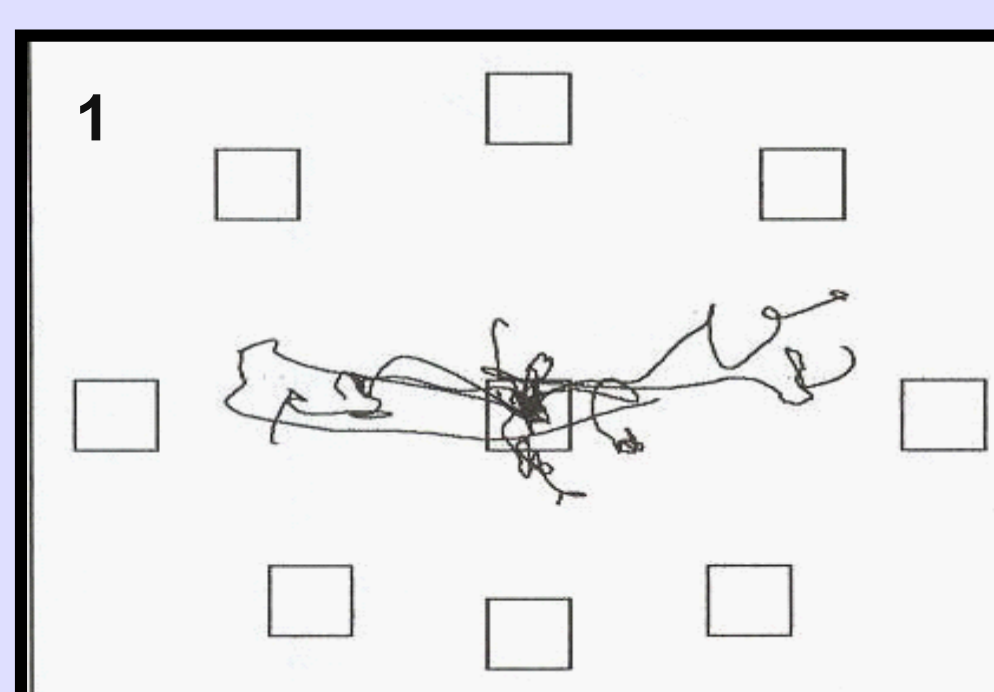
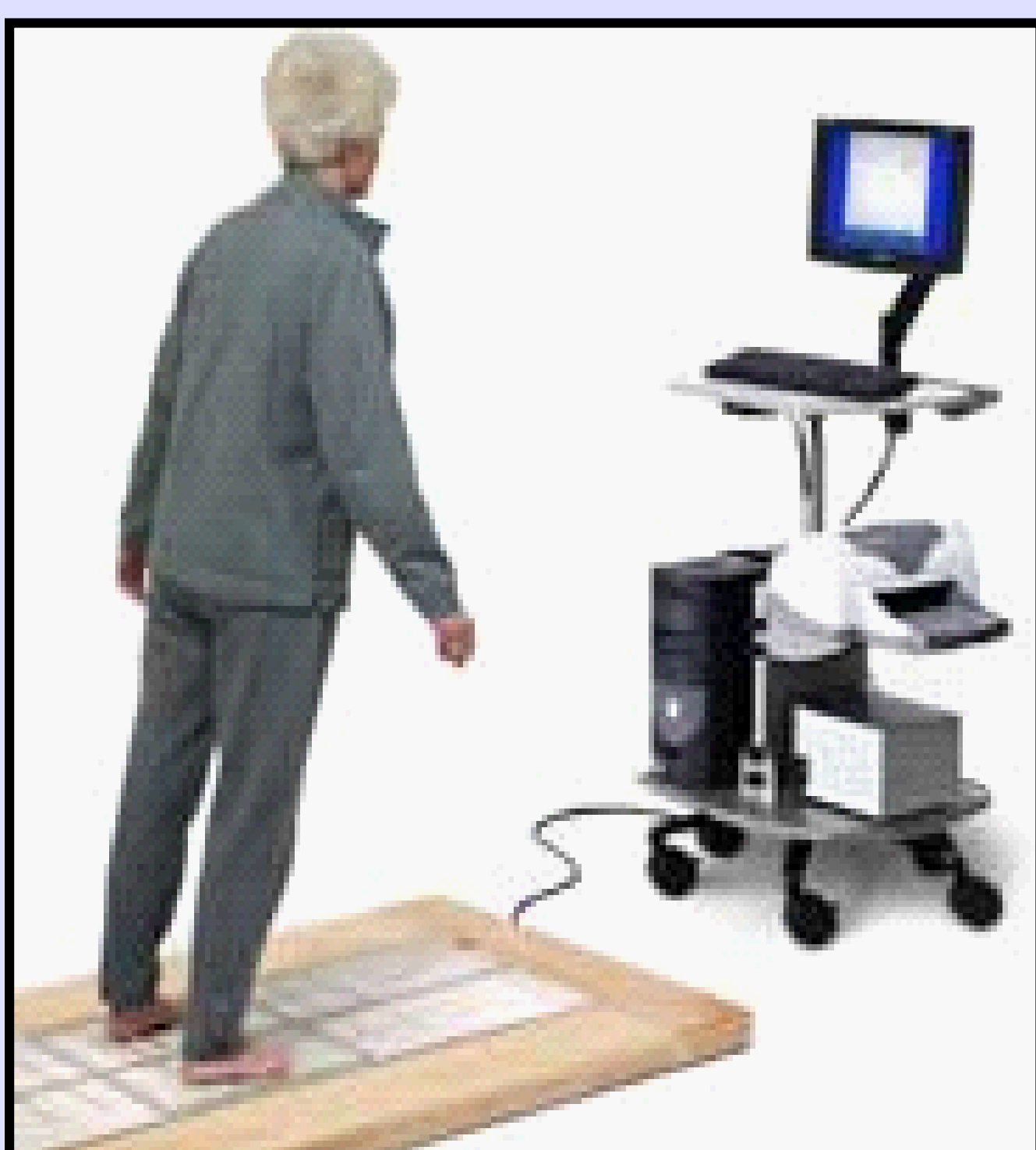


- woman, 57 yrs old
- onset: age 35
- falls: 12 in 2 months



- Neck position sense was tested through a goniometric helmet.
- Repositioning (yaw-pitch-roll target angles) was highly impaired.

11, 1-hr exercise sessions in 6 days.
Strengthening: neck and shoulder girdle.



Neck repositioning and balance greatly improved.
No more falls over the next 3 months.

Neck position sense is crucial to balance.

Brandt T. *J Neurol Neurosurg Psychiatry* 2001;71(1):8-12

In DM-1 severe alterations of neck spindles have been demonstrated.

Swash M. *Clin Neuropathol* 1983;2(2):75-78

A short program of neck strengthening may be beneficial.

