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Knee bobbing in Charcot–Marie–Tooth disease

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Abstract

Patients with Charcot-Marie-Tooth (CMT) disease most commonly have a slowly progressive neuropathy where ankle dorsiflexion weakness is much more prominent than ankle plantar flexion weakness. The eventual involvement of the ankle plantar flexors has major functional consequences for patients as they lose the ability to stand still. We have found the knee bob sign whereby both knees bob up and down when standing still to be a reliable marker of ankle plantar flexion weakness in CMT.

Patients with Charcot–Marie–Tooth (CMT) disease most commonly have a slowly progressive neuropathy where ankle dorsiflexion weakness is much more prominent than ankle plantar flexion weakness. However, the eventual development of ankle plantar flexor weakness has major functional consequences for patients, as they lose the ability to stand still. Patients typically report finding it easier to walk than to stand still and find situations where standing still is important (eg, queues, cocktail parties) very difficult. Patients often view the inability to stand still as a major deterioration in their condition; as such it is important this is diagnosed correctly, especially as appropriate orthotics can help.

We have found the ‘knee bob sign’ – where both knees bob up and down when attempting to stand still – to be a reliable marker of ankle plantar flexion weakness. Knee bobbing describes the flexed knee posture and alternating dorsi and plantar flexion of the ankle that gives the appearance of a bobbing knee (figure 1).

The knee bob sign develops in patients with CMT who have significant distal weakness. This is almost always due to weakness of ankle plantar flexion with coexisting weakness in ankle dorsiflexion, which can be difficult to detect on conventional examination. This difficulty in detecting ankle plantar flexion weakness is partly due to the way in which we traditionally examine strength at the ankle. When lying supine on the bed, a patient can appear to have full strength by pushing the entire leg down from the hip. Also, the gastrocnemius and soleus muscles are normally so strong that they need to be significantly weak to detect weakness by this usual method, even if they are sufficiently weakened so that the patients cannot stand on their toes.

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Contributors

AR wrote the original draft in conjunction with SM. MMR came up with the idea of the article and corrected the original drafts.

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Patients with a positive knee bob sign often adopt a standing posture with the knees flexed (figure 1). If asked to stand with the legs straight, the patients become unsteady as the plantar flexion weakness impairs their ability to stand straight. The patient then leans forward, shifting the centre of gravity forward and compensating by bending at the knee.

Having noted this sign in several patients with CMT, we prospectively examined 13 patients with CMT in the neuropathy clinic for the knee bob sign. Of six patients with a positive knee bob, none could stand on their toes; their mean Medical Research Council (MRC) score for strength of ankle plantar flexion was 2.2 out of 5. Of the seven patients without a knee bob sign, all but two could stand on their toes and their mean MRC score for ankle plantar flexion strength was 4.5 ($p=0.016$). While the association of ankle plantar flexion weakness with knee bobbing is most obvious in CMT due to its slow and predictive progression, we also find it a reliable indicator in acquired neuropathy.

The term ‘knee bobbing’ has also been used to describe the appearance of the patella in patients with primary orthostatic tremor, although in orthostatic tremor the movement is rhythmic and has a higher frequency and smaller amplitude.¹ Patients with peripheral neuropathies with significant proprioceptive loss also have difficulties standing still but this sensory ataxia looks different from knee bobbing. Furthermore, significant proprioceptive loss is not common in a typical patient with CMT.

In conclusion, if a patient with CMT reports not being able to stand still, the clinician should examine the ability to stand on the toes and look for the knee bob sign to identify plantar flexion weakness. The presence of plantar flexion weakness is an important sign indicating progression of typical CMT and the needs for an appropriate foot drop splint, and appropriate and timely orthotic intervention.

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Figure 1.
Note the flexed knee posture which is characteristic of the knee bob sign seen in Charcot–Marie–Tooth patients with ankle plantar flexion weakness.