

# Hung-up knee jerk in stiff-person syndrome

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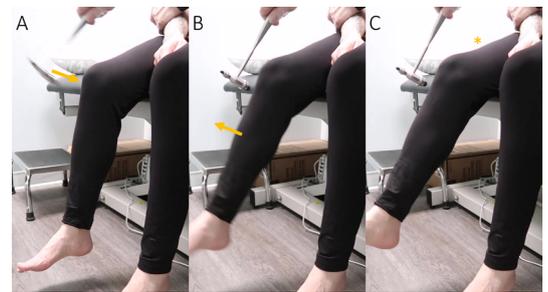
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## DESCRIPTION

A woman in her 30s presented with progressive muscle rigidity, spasms, low back pain, gait difficulty and falls, over 5 years. Symptoms were initially attributed to functional neurological disorder in the context of depression. Examination revealed rigidity in the axial and lower limb muscles, involving the abdominal and thoracolumbar paraspinal muscles causing hyperlordosis, as well as stiff, slow movements of both legs with limited hip and knee flexion. Deep tendon reflexes demonstrated ‘hung-up’ knee jerks (video 1, figure 1). Neuroimaging with MRI of the brain and spinal cord with and without contrast were unremarkable. Electromyography showed continuous motor unit activity and co-contraction of agonist and antagonist muscles, and glutamate decarboxylase-65 (GAD65) levels were elevated in serum (115.68 IU/mL; ref < 1.0) and cerebrospinal fluid (high positive, titre not reported) establishing a diagnosis of stiff-person syndrome. Serum thyroid function testing and haemoglobin A1c were normal, and systemic autoimmune and malignancy testing were negative. Symptoms responded well to regular diazepam alone (10 mg three times daily) without immunotherapy, leading to significantly improved gait and no further falls.

The hung-up knee jerk is a tonic reflex response demonstrated when there is delayed relaxation of the quadriceps femoris after eliciting the patellar tendon reflex (video 1, figure 1).<sup>1 2</sup> Normally, passive stretch of the patellar tendon elicits a muscle response through the mono-synaptic reflex arc between the afferent Golgi tendon organ (dorsal root ganglion) and alpha motoneuron (anterior horn cell), causing a brief muscle contraction which extinguishes the passive stretch and ends the reflex—relaxation may be delayed with muscle hyperexcitability. Also known as Gordon’s phenomenon,<sup>3</sup> it was classically described as a finding in Huntington’s



**Figure 1** Hung-up knee jerk: (A) leg at rest before tendon hammer strike (arrow indicating direction of force); (B) eliciting the patellar tendon reflex (arrow indicating knee extension); (C) there is delayed relaxation of the tonic quadriceps femoris (\*), causing a ‘hung-up’ appearance as the lower leg slowly returns to neutral position (note diffuse muscle contraction seen throughout the leg).

disease by Bing,<sup>2</sup> who observed that in some patients, the elevated foot may remain elevated for a few seconds because of repeated quadriceps contractions.<sup>2 3</sup> This sign has also been reported in Sydenham’s chorea<sup>4</sup> and more recently stiff-limb variant of stiff-person syndrome.<sup>5</sup> A similar phenomenon of delayed relaxation of reflexes occurs in hypothyroidism (myxedema reflex or Woltman’s sign), best demonstrated in the Achilles tendon.<sup>6</sup> In fact, it was Woltman—along with Moersch—who in 1956 published their seminal paper defining stiff-person syndrome and the characteristic ‘wooden-man’ appearance in 14 cases, the first of which was observed in 1928.<sup>7</sup> While neither diagnostic nor pathognomonic, the hung-up knee jerk is a simple clinical sign that may provide an important clue towards a diagnosis of stiff-person syndrome, avoiding misdiagnosis of functional neurological disorder.<sup>5</sup>

Stiff-person syndrome is rare, with an estimated prevalence of 1–2 per million.<sup>8</sup> Most cases are thought to be autoimmune, associated with high-titre GAD65 antibodies, often with concurrent thyroid disease or type 1 diabetes.<sup>8 9</sup> However, rare paraneoplastic cases have been described, often without GAD65 antibodies.<sup>8</sup> Inhibition of gamma-aminobutyric acid-mediated pathways leads to hyperexcitability manifesting as muscle spasms and rigidity.<sup>8 9</sup> Electromyography may characteristically show co-contraction of antagonistic muscles.<sup>7 8</sup> Symptoms are generally treated with benzodiazepines and in some cases immunotherapy<sup>8 9</sup>; response to diazepam is often included in diagnostic criteria.<sup>8</sup>



**Video 1** Hung-up knee jerk: there is delayed relaxation of the quadriceps femoris after eliciting the patellar tendon reflex, causing a ‘hung-up’ appearance as the lower leg slowly returns to neutral position.



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## Learning points

- ▶ Stiff-person syndrome is characterised by progressive muscle rigidity and spasms, often affecting the axial and lower limb muscles along with abdominal and thoracolumbar paraspinal muscles causing hyperlordosis.
- ▶ The hung-up knee jerk is demonstrated when there is delayed relaxation of the quadriceps femoris after eliciting the patellar tendon reflex and can be seen in stiff-person syndrome as well as Huntington's disease and Sydenham's chorea.
- ▶ While neither diagnostic nor pathognomonic, this simple clinical sign may be an important clue towards a diagnosis of stiff-person syndrome and avoiding misdiagnosis of functional neurological disorder.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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