

Schiff-Sherrington syndrome

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Buddy, a two-year-old fit and athletic female boxer, developed acute signs of paralysis while fetching a ball in the garden. The very worried owners reported Buddy had given a sudden loud yelp and when they rushed to her aid they had found her lying distressed on her side and unable to stand. Without delay, they rushed her to your surgery in search of help.

Considering the history and clinical presentation, you feel pretty certain your patient is suffering from an acute spinal cord injury, and you think Buddy's posture could be suggestive of Schiff-Sherrington syndrome.

Question

Somewhere in the back of your mind you remember Schiff-Sherrington syndrome might be associated with a poor prognosis – but is this correct?

Answer

Absolutely not. The presence of a Schiff-Sherrington syndrome is not a prognostic factor, but simply an indicator for a specific (and, unfortunately, often misleading) clinical presentation in patients with acute thoracolumbar injuries.

With Schiff-Sherrington syndrome, the patient is usually unable to stand unaided on all four limbs and is presented in lateral recumbency with severe extension of the forelimbs and hindlimb paralysis ([Figure 1](#)).

This can mimic a cervical spinal cord lesion, but when the patient is held up, the forelimbs are normal apart from stiffness, and the hindlimbs are paralysed. There are very good voluntary movements in the forelimbs, whereas voluntary movements are usually absent in the hindlimbs

(paraplegia). The proprioception and the flexor withdrawal reflexes are normal in the forelimbs, but proprioception is absent in the hindlimbs. Quite commonly, these patients will also have a thoracolumbar cut-off of their cutaneous trunci reflex.

The sign of increased extensor tone in the forelimbs that results in the clinical picture in Schiff-Sherrington is the result of the disinhibition of the extensor motor neuron in the caudal cervical spinal cord segments. This occurs due to the injury of a specific layer of neurons in the cranial lumbar spinal cord segments called the border cells. These cells, located in the border of the grey column of the cranial lumbar spinal cord, will normally inhibit the extensor tone of the forelimbs via their axons sent cranially to the cervical intumescence (from L1-L5 spinal cord segments to C6-T2 spinal cord segments).

In cases of Schiff-Sherrington syndrome, the acute thoracolumbar myelopathy leads to an injury of these border cells and a lack of inhibition of the cervical spinal cord motor neurons, hence the hyperextension of the forelimbs associated with the hindlimb paralysis.

The Schiff-Sherrington posture only occurs in association with acute and severe thoracolumbar spinal cord injury, but is not a prognostic indicator. The prognosis in patients with Schiff-Sherrington instead depends – as is usually the case in patients with paraplegia – on the presence or absence of nociception in the hindlimbs, perineum and tail. This is independent from the disinhibition of the forelimb extensor muscles, which will normally spontaneously resolve in the first days after the onset of the neurological signs.

Take home message

Having correctly identified Schiff-Sherrington syndrome, you can now carry out a detailed neurological examination to confirm your suspicion that Buddy's spinal injury is located in the thoracolumbar area, and consider diagnostic investigations such as radiography, a CT scan or an MRI scan.

In Buddy's case, an MRI scan ([Figure 2](#)) of the thoracolumbar spine revealed a fibro-cartilaginous embolus (FCE), which was managed conservatively. Buddy made a quick recovery with subsequent resolution of both her forelimb extension and paraplegia.

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