

Hoyland House, Painswick GL6 6RD, UK k.barraclough@btinternet.com

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EASILY MISSED?

Cauda equina syndrome

Kevin Barraclough *general practitioner*

What you need to know

- Sciatica is pain or numbness that is usually referred below the knee (in contrast to non-radicular pain referred to the upper posterior thigh). Although bilateral sciatica is the classic "red flag" symptom for cauda equina syndrome (CES), it is present in only about 50% of cases
- It is critical to diagnose CES before the patient becomes incontinent. Advice to return if the patient becomes incontinent is too little too late
- Pain inhibition may cause difficulty passing urine, but patients with pain inhibition alone do not have loss or reduction in bladder or urethral sensation or perineal sensory disturbances
- Assessment of anal tone is a poor predictor of cauda equina function, while subjective disturbance of saddle sensation is an unusual symptom that needs to be considered carefully. The accuracy of perianal sensory testing is unknown, and normal results should not be over-interpreted

A 42 year old woman presented to an out-of-hours general practitioner with a five day history of low back pain with burning pain radiating into her right foot.

She had a body mass index of 39 and a 12 year history of chronic low back pain with intermittent left sided "sciatica" pain in her thigh. She had noted "tingling" in her genital area. There was no incontinence. The general practitioner considered, on examination, that anal tone and perianal sensation were normal, as were power, tone, reflexes, and sensation in the legs. The patient was treated with analgesia and given advice to seek review if she developed bilateral sciatica, became incontinent, or developed leg weakness. Three days later, she was admitted with cauda equina syndrome and underwent surgical decompression. She was left with lower limb weakness, numbness of the genitalia, loss of sexual function, and urinary and faecal incontinence.

What is cauda equina syndrome?

Cauda equina syndrome (CES) is a rare condition in which the lumbosacral nerve roots that extend below the spinal cord itself are compressed within the lumbosacral spinal canal (fig 1). Usually the cause is a central disc prolapse at the L4/5 or L5/S1 level. More rarely, compression can occur due to pathology in a higher disc or to infection or a tumour.

This is one of a series of occasional articles highlighting conditions that may be more common than many doctors realise or may be missed at first presentation. The series advisers are Anthony Harnden, professor of primary care, Department of Primary Care Health Sciences, University of Oxford, and Dr Kevin Barraclough, School of Social and Community Medicine, University of Bristol. You can read more about how to prepare and submit an Education article on our Instructions for Authors pages: https://www.bmj.com/about-bmj/resources-authors/article-types.

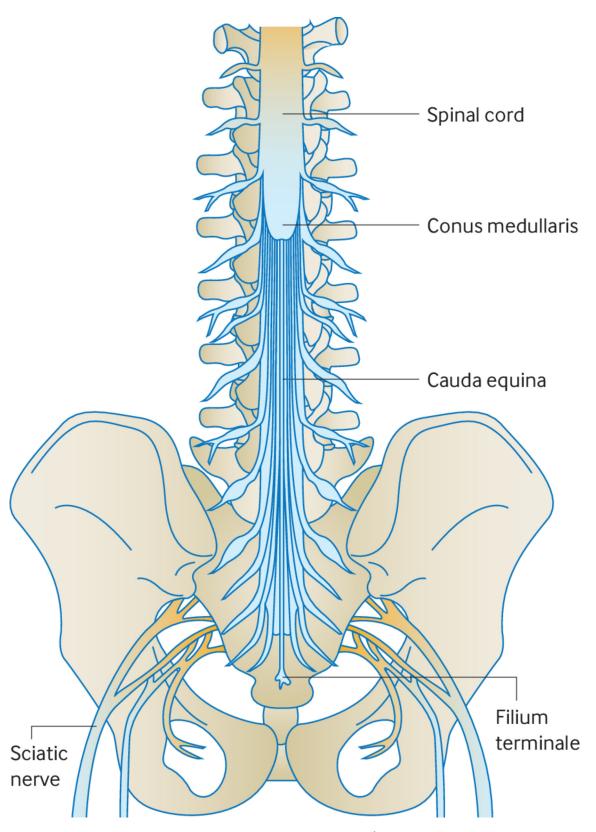


Fig 1 | Anatomy of the lower lumbar and sacral spine showing the cauda equina (redrawn from Lavy et al¹)

The nerve fibres in the cauda equina supply saddle, bladder, and rectal sensation; sensation and motor control of the external urethral and anal sphincters; and the fine calibre parasympathetic fibres of

the pelvic viscera. There are also lower motor neurones, so any weakness in CES will be flaccid, not spastic. The autonomic fibres,

in particular, are fine in calibre and are rapidly and irreversibly damaged by pressure.

The clinical features of CES are those of loss of bladder and urethral sensation, and alteration of saddle sensation in someone who usually also has unilateral or bilateral sciatica. CES can present in one of three ways. It can present acutely as one of the first features of lumbar disc herniation (type I), as an endpoint in someone with chronic low back pain (type II), or develop slowly over days (or even weeks) with gradually progressive saddle numbness and urinary symptoms (type III).

CES is diagnosed when there are both clinical features of CES and evidence of compression of the cauda equina on magnetic resonance imaging (MRI). This poses a challenge since: (a) some of the features of CES occur frequently in patients without radiological evidence of CES,³ 4 and (b) radiological evidence of central disc prolapse causing compression of the cauda equina occurs in patients without clinical features of CES.⁵

Why is it missed?

CES is missed in primary care because it is rare, the clinical features (particularly disturbance of urinary function and saddle sensation) may initially be subtle, and many patients with uncomplicated acute back pain have some type of urinary symptoms.

CES is rare in primary care, whereas acute back pain, sciatica, and lumbosacral disc prolapse are common. The incidence of CES is estimated to be between 1 in 33 000 and 1 in 100 000 per year. ⁶ Thus, a full-time general practitioner may never encounter a case in a clinical career.

A second difficulty is that many patients with severe, acute low back pain have urinary symptoms that are not due to CES. Given the high prevalence of back pain and low incidence of CES, it would not be practical to refer all patients with low back pain and any urinary symptoms to the local emergency department or for an MRI scan. It is a subset of urinary symptoms (those that are not due to pain inhibition) that are suggestive of CES.

Yet another difficulty is that evidence is lacking on whether a normal clinical examination carried out by a non-specialist rules out CES. The performance characteristics and inter-observer agreement of, for example, perianal sensory testing carried out by a non-specialist are unknown. The performance characteristics of assessment of anal tone are poor. Yet a referring general practitioner will usually be asked if anal tone and perianal sensation are normal.

Finally, guidance to primary care clinicians on when to suspect CES is somewhat vague. Several guidelines list "red flag" features but do not give adequate lead time for intervention and do not advise clinicians how to weight the features. Urinary incontinence is a well recognised "red flag" for CES, for example, but, by the time a patient with CES has urinary incontinence, it is too late: the fine parasympathetic nerves and the sacral sensory fibres in the cauda equina that maintain continence and sexual function will not recover, and the patient is likely to remain incontinent for life. As is often the case in clinical medicine, it is unclear which features might reasonably "rule out" CES (for example, normal examination findings) or "rule in" the condition (for example, any perianal symptoms in someone with back pain). This leaves the assessing generalist clinician, who is unlikely to have ever encountered the condition before, torn between not wanting to miss the diagnosis or to needlessly alarm patients with unnecessary testing.

Why does it matter?

If a patient who is developing CES but is not yet incontinent undergoes surgery, there is a reasonable chance of avoiding the potentially catastrophic consequences of urinary incontinence, faecal incontinence, loss of sexual function, saddle anaesthesia, neuropathic pain, and sometimes paralysis of the legs. This early stage is often referred to as cauda equina syndrome incomplete (CESI). The patient may have reduced bladder or urethral sensation and/or saddle sensory disturbance, but they retain bladder control. Once the patient has developed urinary retention and overflow incontinence (known as CES with retention or CESR), the outcome of surgery is much worse. ^{8 9} CES is one of the leading causes of medical litigation in the UK, and the average compensation, even by 2004, was £336 000. ¹⁰

How is it diagnosed?

The initial crucial step is that the clinician must take a careful history with the possibility of CES and its multiple presentations in mind. The clinician also needs to determine how much weight to put on the presence or absence of clinical symptoms and (importantly) clinical signs.

The clinical features of CES are usually considered to be a combination of the following occurring in someone with acute or chronic low back pain: disturbance of urinary function, disturbance of saddle sensation, reduced anal tone, and possibly bilateral sciatica. Many patients with severe low back pain have some difficulties with passing urine. They may find it difficult to maintain the necessary position (sitting or standing) over the lavatory, for example, and relaxing the muscles of the pelvic floor can exacerbate already severe low back pain. Pain may also cause an involuntary inhibition of micturition. Drugs such as gabapentin, anticholinergics, and opiates also affect bladder function. Crucially, in patients with pain inhibition alone, there is no loss of bladder sensation and no disturbance of saddle sensation.

A patient with neurological bladder dysfunction due to CES will have some or all of the following symptoms:

- Reduced awareness of bladder filling
- · Loss of the urge to void
- Reduced awareness that micturition is occurring
- Inability to voluntarily interrupt the stream of urine in mid-flow
- Recent onset or progressively worsening weak urinary stream with terminal dribbling (this is obviously also common in men with bladder outflow obstruction and can occur with anticholinergic drugs)
- Loss of urethral sensation or altered sensation over the genitalia
- Some alteration of peri-anal sensation (a "numb bum" on wiping). There may also be perianal paraesthesia or pain.

Of note, patients with loss of bladder muscle tone can sometimes "compensate" by contracting abdominal muscles or pressing on the lower abdomen to force micturition.

Bilateral sciatica

Around half of patients with CES have bilateral sciatica. ^{47 11} Sciatica is pain or altered sensation in a nerve root distribution that generally goes below the knee to the foot. Many people with severe low back pain get pain referred into the posterior thighs. That is typically not radicular nerve pain (sciatica). In CES, the affected dermatomes are almost always L4, L5, or S1, ²⁸ and these cause pain or altered

sensation that extends below the knee into the foot. However, half of patients have only unilateral sciatica, and, somewhat paradoxically, as a disc fragment migrates centrally to cause CES, the sciatica may improve.

Three useful UK studies retrospectively examined the clinical features of sequential patients referred to tertiary units with features of suspected CES who underwent MRI scans.^{3 47} Of the total of 413 patients in the three studies, 106 (26%) had CES. Of the 106 patients with CES, only 43 (41%) had bilateral sciatica. Of the 307 patients without CES, 78 (25%) had bilateral sciatica. Thus, the absence of bilateral sciatica does not exclude CES, nor does its presence rule it in; the appropriate safety-netting advice for someone with sciatica but no other features to suggest CES is also unclear.

Perianal sensation, saddle anaesthesia, and reduced anal tone

If a patient has acute loss or reduction of perianal sensation, they will usually be quite aware of it, just as a patient who has had dental anaesthesia will be aware that the skin over the jaw is numb. No studies have examined the internal agreement of clinicians when assessing perianal sensation. It is not an examination that general practitioners do often, so the reliability of findings (normal or abnormal) should not be over-estimated.

Table 1 shows the frequency with which perianal sensory loss and reduced anal tone have been found in patients with and without CFS.

Study	No of patients	Anal tone (recorded)		Saddle sensation (recorded)	
		Reduced	Normal	Numbness	Normal
Hoeritzauer et al ⁴ :	276				
With CES	78 (28% of total)	14 (18% of CES cases)	17 (22% of CES cases)	50 (64% of CES cases)	18 (23% of CES cases)
No CES	191	37 (19% of no CES cases)	83 (43% of no CES cases)	101 (53% of no CES cases)	77 (40% of no CES cases)
Balasubramanian et al ³ :	80				
With CES	15 (19% of total)	3 (20% of CES cases)	10 (66% of CES cases)	8 (53% of CES cases)	5 (33% of CES cases)
No CES	65	4 (6% of no CES cases)	58 (89% of no CES cases)	12 (18% of no CES cases)	52 (80% of no CES cases)
Gooding et al ⁷ :	57				
With CES	13 (23% of total)	5* (38% of CES cases)	8* (62% of CES cases)	5* (38% of CES cases)	8* (62% of CES cases)
No CES	44	11* (25% of no CES cases)	33* (75% of no CES cases)	22* (50% of no CES cases)	22* (50% of no CES cases)

- Reduced anal tone was recorded in 23 (22%) of the 106 patients with CES in the three studies, ^{3 4 7} and normal anal tone was recorded in 35 (33%) of those with CES. Other studies of anal tone assessments in clinical circumstances and using model simulations have shown poor levels of sensitivity, specificity, and internal agreement. ^{12 13}
- Objective saddle numbness was recorded in 63 (59%) of those
 with CES, and normal saddle sensation was recorded in 31 (29%).
 Thus, around 40% of those with CES were not recorded as having
 objective signs of perianal sensory loss. However, one potential
 limitation of the data is that the experience level of the examining
 clinician is not recorded.

It could be argued that, if a patient has symptoms of disturbed saddle sensation, then specialist opinion should be sought irrespective of signs, while if there are no sensory symptoms it is most unlikely there will be signs. It also seems likely that symptoms which are volunteered (such as, "When I wipe myself it seems numb") are likely to have a higher positive predictive value than elicited symptoms (such as "Have you noticed any numbness when wiping your bottom?" "Well, yes. A bit."). However, this has not been studied, and it is not what current guidance states.

Thus, the perceived absence of clinical signs (particularly with non-specialist assessment) does not necessarily rule out CES in patients in whom the clinical suspicion is high.

Guidance on suspicion and referral for assessment of possible CES

Most international guidance on the assessment of low back pain and sciatica advises physicians to consider CES when there are "red flags," as does the UK's NHS Clinical Knowledge System (https://cks.nice.org.uk/topics/sciatica-lumbar-radiculopathy/). However, the "red flags" do not always coincide, and they list features without giving any sense of the clinical weight that can be given to the presence or absence of these features. ¹⁴ ¹⁵ Ideally, prescriptive advice would describe early features of CES, not late or irreversible features such as urinary and faecal incontinence. A general practitioner could consider advising a patient with sciatica to return or seek urgent help if they develop the clinical features listed in box 1.

Box 1: "Red flags" for cauda equina syndrome in patients with sciatica

- Onset of bilateral numbness or weakness in the legs
- Onset of any sense of numbness or pins and needles around the anus (a "numb bum") or genitals
- Any alteration in the sensation of a full bladder, desire to pass urine, or awareness of passing urine

In the case described above, the general practitioner followed current guidance on the assessment and management of possible

CES. However, safety netting advice to seek help if incontinence develops was probably ill advised and illogical, since bladder function will be lost by that stage. Perianal sensory disturbance is an unusual symptom and should possibly be given more weight than current guidance suggests, even in the absence of clear signs of perianal sensory loss. It is also necessary to be clear that a significant proportion of patients with CES never develop bilateral sciatica or limb weakness.

How is cauda equina syndrome managed?

Cauda equina syndrome incomplete (CESI) or CES with retention is diagnosed if the patient has clinical features of CES and radiological evidence of compression of the cauda equina on MRI. CES is a neurosurgical emergency and patients undergo surgical decompression.

How patients were involved in the creation of this article

No patients were involved in the creation of this article.

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Patient consent not required (patient anonymised, dead, or hypothetical).

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I (KB) have seen only one case of CES, and that was in hospital practice. I have been involved in 74 medicolegal cases of CES between 2010 and 2019. I learnt a great deal about CES from Mr Robert Macfarlane over the years, but the opinions expressed here are my own.

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