

Tethered Cord Syndrome

A patient guide

In brief: Tethered cord syndrome is a clinical condition in which the lower end of the spinal cord is abnormally anchored within the spinal canal, so that normal movement and growth place it under sustained stretch and progressively impair its function. It is defined by a pattern of slowly worsening neurological, urological and orthopaedic problems attributable to that traction — not by any single appearance on a scan. This distinction matters, because a low-lying spinal cord, a spina bifida occulta, or a thickened filament seen on imaging are anatomical findings or underlying causes, and are not the same thing as the syndrome itself.

What tethered cord syndrome is (a clinical syndrome, not just a low conus)

Tethered cord syndrome (TCS) is best understood as a **functional** diagnosis. The defining feature is progressive dysfunction of the lower spinal cord — affecting the legs, the bladder and bowel, sensation in the saddle region, and sometimes the spine itself — arising because the cord is held under abnormal, sustained tension at its lower end.

The crucial point, and one that is frequently misunderstood, is that the radiological appearances commonly associated with tethering are not, by themselves, the diagnosis. The conus medullaris (the tapered lower tip of the spinal cord) sitting lower than usual is an anatomical observation. Many people have a low-lying conus and never develop any symptoms at all. Equally, a thickened filum terminale, a small spinal lipoma, or a split-cord septum may be present on a scan in a person who is entirely well. These are **tethering lesions** — structures capable of anchoring the cord — but they are not the syndrome.

The syndrome is the clinical consequence that arises when traction on the cord produces a documented, progressive injury. In practice this means that the diagnosis rests on three pillars considered together: a plausible anatomical cause of tethering, a corresponding pattern of symptoms and signs, and — critically — evidence of **progression** over time, often supported by functional tests of bladder and nerve function. A static, lifelong, non-worsening finding is a different matter from a syndrome that is actively evolving.

This framing — syndrome versus finding — runs through everything that follows. It explains why two people with apparently similar scans may be managed very differently, and why a careful clinical history matters as much as the imaging.

The anatomy of tethering (conus, filum terminale)

The spinal cord does not run the full length of the spinal canal. In adults it ends in a tapered cone — the conus medullaris — typically at around the level of the first lumbar vertebra (the T12–L1 region). Below this, the canal contains a fan of nerve roots known as the cauda equina, which descend to exit at their respective levels.

From the tip of the conus, a fine fibrous thread called the filum terminale runs downward to anchor the cord to the base of the sacrum. In health this filament is thin and elastic. Its role is to fix the cord gently in position while still allowing it to accommodate the small movements that occur with bending, growth and changes in posture. It acts, in effect, as a shock-absorber against cranio-caudal (up-and-down) traction.

In the developing child, the spinal cord normally appears to "ascend" relative to the bony spine, because the vertebral column grows faster than the cord. The conus reaches its adult position within the first months of life. A conus that fails to ascend, or that is held down, ends up lower than expected.

Tethering occurs when the elastic, buffering filum is replaced by an inelastic anchor, or when some other structure fixes the cord. A filum that has become thickened or fat-infiltrated loses its elasticity, and instead of buffering traction it transmits it directly to the conus. The same effect can be produced by a fatty mass continuous with the cord, by a bony or fibrous septum splitting the cord, or by scar tissue from previous surgery. Whatever the mechanism, the result is that the lower cord can no longer glide freely, and is held under sustained stretch.

The prevailing explanation for how this stretch causes harm derives from the work of Yamada and colleagues, who proposed that traction on the lower cord impairs its oxidative metabolism — in effect, a stretch-induced reduction in the cord's ability to use oxygen. In experimental terms this manifests as measurable changes in the cord's energy chemistry and electrical activity. Importantly, these changes appear to be reversible in their early stages and to become fixed only later, which is the conceptual basis for the observation that earlier release of tethering tends to give better results.

What causes it

Tethered cord syndrome is the common endpoint of several distinct underlying lesions. Identifying which one is present is central to assessment, because it shapes the anatomy and the surgical considerations.

- **Tight or fatty filum terminale.** The classic cause of so-called "primary" tethering. The filament is thickened (a diameter greater than about 2 mm at the L5–S1 level is the conventional threshold) and/or infiltrated with fat, rendering it inelastic.
- **Spinal lipoma and lipomyelomeningocele.** A fatty mass continuous with the cord, often passing through a defect in the posterior elements of the spine. This is one of the commonest causes of tethering identified in children, where both the anchoring effect and the bulk of the lesion contribute to dysfunction.
- **Diastematomyelia (split cord malformation).** Here the cord is divided lengthwise by a median septum. In Type I this septum is a rigid bony or cartilaginous spur; in Type II it is fibrous. Either way, the septum anchors the cord and can produce tethering.
- **Scarring after previous spinal surgery (secondary tethering / retethering).** Adhesions following repair of an open spina bifida (myelomeningocele) or after earlier lipoma or untethering surgery can re-anchor the cord. Some degree of re-adhesion after myelomeningocele repair is near-universal, though only a proportion of patients go on to develop symptoms.
- **Spina bifida occulta.** A failure of the posterior bony arch to fuse fully. This is extremely common, usually entirely incidental, and on its own does not cause tethered cord syndrome. Its significance is chiefly as a marker: when accompanied by overlying skin changes, it raises the suspicion that a true tethering lesion lies beneath.

How it tends to present in children and in adults

The way tethered cord syndrome declares itself differs strikingly between childhood and adult life, and this difference is one of its most characteristic features.

In children, the condition is often suspected from cutaneous (skin) markers over the lower back: a hairy patch, a dimple or sinus, a birthmark, a small fatty swelling. Other presentations include asymmetry of the feet or legs, foot deformity, a change in gait, scoliosis (curvature of the spine), problems with bladder control out of keeping with age, and weakness or altered sensation in the legs. Frequently the condition is identified during the investigation of a known dysraphic abnormality rather than presenting in isolation.

In adults, presentation is more often dominated by pain. This is characteristically diffuse and does not follow a single nerve root distribution — it may be felt across the lower back, into the legs, or in the perineal and anorectal region. A notable feature of the adult form, described in the landmark series by Pang and Wilberger, is that symptoms are frequently precipitated by a triggering event: a fall or injury, a bout of vigorous activity or stretching, or the gradual narrowing of the spinal canal with age, each of which adds tension to a cord that is

already taut. Progressive disturbance of bladder and bowel function, sensory and motor deficits in the legs, and sexual dysfunction may also occur. Adult onset is rarer than the paediatric form, and the anchoring filum in adults is less commonly grossly thickened than it tends to be in children.

How it is assessed (MRI conus level, urodynamics)

Assessment combines imaging, functional testing and the clinical course, and no single element is sufficient on its own.

Magnetic resonance imaging (MRI) is the first-line investigation. It demonstrates the level at which the conus terminates (normally around T12–L1; a conus lying below the L1–L2 disc is generally regarded as abnormal, and one below the mid-to-lower body of L2 is definitely so). It also shows the thickness of the filum, any fatty infiltration, the presence of a lipoma, a split-cord septum, or other features of dysraphism. A central and recurring point, however, is that a *normal-level conus* does not exclude the diagnosis* — this is the very crux of the controversy discussed below.

Urodynamic studies — formal tests of bladder storage and emptying — occupy a pivotal place. They can reveal detrusor overactivity, impaired bladder compliance, dyscoordination between the bladder and its sphincter, and signs of denervation. They serve both to support the diagnosis and to monitor change over time. Where imaging is unremarkable, much of the objective diagnostic weight may rest on urodynamic and electrophysiological abnormalities.

The documented clinical course is essential. Because the syndrome is defined by progression, evidence that symptoms or test results are worsening is what distinguishes a syndrome requiring attention from an incidental, stable finding.

The "occult tethered cord" debate

One area genuinely divides expert opinion: so-called occult tethered cord syndrome (OTCS), sometimes termed minimal or tight-filum syndrome.

The term describes patients — most discussion concerns children — who have symptoms suggestive of tethering, particularly a refractory neurogenic bladder or bowel, together with a conus that lies at a *normal* level and a filum of *normal* calibre on imaging. In other words, the usual radiological hallmarks of tethering are absent, yet a clinical picture consistent with traction is asserted to be present.

The argument *for* recognising and treating this entity, advanced by proponents such as Selden, draws on the Yamada paradigm: if traction can injure the cord by impairing its metabolism, then in principle a relatively normal-looking but inelastic filum could exert harmful tension even with a normally positioned conus, and section of that filum might relieve otherwise intractable symptoms in carefully selected patients.

The argument *against*, put by sceptics including Steinbok, Drake and others, is that the evidence supporting this is weak: it rests largely on uncontrolled case series, the diagnosis is difficult to define objectively, and improvement after surgery may not reliably reflect a genuine treatment effect. These commentators have called for properly designed randomised trials before the practice can be regarded as established.

The honest position, and the one reflected here, is that occult tethered cord syndrome remains an area of uncertainty and ongoing debate rather than settled practice.

How it is distinguished from related conditions

Because the same images and terms recur in this field, it is worth setting out plainly how the syndrome differs from the findings and lesions it is associated with.

- **A low-lying conus** is a radiological observation, not a diagnosis. It may support a suspicion of tethering, but a low conus without progressive symptoms is not tethered cord syndrome, and asymptomatic low-lying conus is

common.

- **Spina bifida occulta** is a frequently incidental bony variant. By itself it does not constitute the syndrome; its main value is as a clue to a possible underlying tethering lesion, especially when skin changes are present.

- **Diastematomyelia**, a spinal lipoma and a tight or fatty filum are *causes* — the anchoring lesions. They tether the cord. The syndrome is the functional damage that results when that tethering produces progressive injury; the lesion can be present without the syndrome.

- **Tethered cord syndrome itself** is the clinical entity, defined by progressive neurological, urological and orthopaedic dysfunction attributable to traction, and identified through the combination of cause, clinical picture and evidence of progression rather than by any one scan.

In short, the lesions answer the question "what is anchoring the cord?", whereas the syndrome answers the question "is that anchoring causing progressive harm?".

What the evidence shows about management (untethering; retethering risk) — impersonal

The published evidence on management is summarised here in general, impersonal terms; it does not constitute advice and individual circumstances vary widely.

Surgical untethering — release of the anchoring lesion, or section of the filum terminale — is the established approach for symptomatic, progressive tethered cord syndrome with a clear tethering lesion. Case series report symptomatic improvement in roughly two-thirds to three-quarters of patients. Pain and spasticity tend to respond best; urological and urodynamic improvement is reported in around half to three-quarters of cases but is less reliable, and established deficits frequently do not fully normalise. A consistent theme across the literature is that earlier intervention, before injury becomes fixed, is associated with better outcomes.

Prophylactic untethering of asymptomatic lesions — for example, spinal lipomas discovered before any symptoms appear — is debated. Some centres report that aggressive total or near-total resection of a lipoma, with reconstruction of the neural placode and an expansile duraplasty, markedly reduces later progression and retethering compared with partial resection; this is technically demanding and not universally adopted. Other groups advocate conservative observation of asymptomatic conus lipomas.

Retethering — secondary tethering after surgery — is a major and recurring issue. It is especially relevant after repair of an open spina bifida, where some degree of re-adhesion is near-universal, and after lipoma surgery. Repeat operation can stabilise or improve symptoms but carries cumulative risk, and scoliosis has been associated with earlier retethering. Long-term paediatric cohorts suggest that the risk of symptomatic retethering tends to fall with increasing age.

Spinal column-shortening osteotomy is an alternative strategy, particularly for adults and for retethering. By shortening the bony column, cord tension is reduced *without* direct manipulation of the cord or dura, which can lower the risks of cerebrospinal fluid leak and neurological injury. It is increasingly studied as an option in complex or recurrent cases.

For occult tethered cord syndrome, as noted above, management is genuinely controversial, and the literature does not support treating it as settled practice.

Outlook

The outlook in tethered cord syndrome depends heavily on the underlying cause, the duration and severity of dysfunction before any intervention, and whether the condition is being treated for the first time or has recurred.

In broad terms, the literature suggests that pain and spasticity are the symptoms most likely to improve, that bladder and bowel function improve less predictably, and that deficits which have already become fixed tend not

to recover fully even when the tethering is relieved. The recurring message that earlier treatment is associated with better results reflects the underlying biology: stretch-induced changes appear reversible at first and become entrenched with time.

Tethering is not always a single, once-and-for-all problem. The possibility of retethering, particularly in those with complex dysraphism or a history of previous surgery, means that long-term follow-up — with periodic clinical review and, where appropriate, repeat functional testing — is a feature of how the condition is monitored over a lifetime. The trajectory varies considerably from one person to another, which is precisely why the syndrome is assessed individually rather than by formula.