Tethered Cord Guide

Unravelling Tethered Cord for EDS Patients

Written by EDS patient advocate Christie Cox, reviewed by Dr. Pradeep Chopra and Dr. Sunil Patel for medical accuracy.

Understanding Tethered Cord Syndrome and Occult Tethered Cord

Unravelling the tethered cord has proven to be an elusive topic. Basically, there is not a great deal of credible insight or medical research available on it. To start simply, we begin with basic anatomy and symptoms most commonly seen by doctors to help you better understand the complex medical condition with a challenging diagnostic journey.

In short, Tethered Cord Syndrome (TCS) revolves around the undue stretching of the spinal cord resulting in numerous neurological complications and pain. TCS can impact functioning as this part of the body serves as a relay station between the brain and the rest of the body and is responsible for several autonomic functions involved in such fundamental processes as breathing, sleeping, and sensory and motor functions including so it is critical to get evaluated by an expert:

- Respiratory function
- Sleep and arousal
- Sensory and vestibular balance
- Motor functions (including regulation of processes like chewing, salivation)
- Origin of Cranial Nerves: Trigeminal nerve, Abducens nerve, Facial nerve Vestibulocochlear nerve

Back to Basics: The Anatomy of The Human Spine

- The vertebral column plays a pivotal role in sustaining an upright spine while shielding the spinal cord from injuries. It comprises thirty-three vertebrae.
- The upper twenty-four vertebrae have intravertebral discs, rich in collagen- a known trouble spot of faulty protein in hEDS. These discs normally functioning grant flexibility to the vertebral column.
- The spinal canal envelops the spinal cord in cerebrospinal fluid, safeguarding it from damage and providing essential nutrients. The outermost layer of the meninges, making up the spinal canal, is the dura mater.
- The spinal cord acts as the communication bridge between the brain and the body's nerves.

Tethered Cord Syndrome & Its Association with EDS

Tethered Cord Syndrome occurs when the filum terminale adheres to either scar tissue or the dura lining of the spinal canal. "Dura," is one of the three layers of membranes (meninges) that surround and protect the brain and spinal cord. While it can emerge at any spinal canal point, its prevalence is higher at the lumbosacral level (low back). Patients with conditions like Spina Bifida, Spina Bifida Occulta, and some patients with hypermobility and/or Ehlers-Danlos Syndromes (EDS) are more prone to tethered cord. Tethered Cord happens when the sticky fibrous tissue of the filum adheres to fatty/scar tissue or the dura lining of the spinal canal INSERT IMAGE. While this tethering can happen anywhere in the spinal canal, it is most common at the lumbosacral level often felt in the low back/sacrum/tailbone area. When the tethered filum pulls the spinal cord tightly enough that it causes neurological problems, it becomes known as Tethered Cord Syndrome (TCS).

Identifying Symptoms of Tethered Cord Syndrome

Detecting TCS can be extremely challenging due to its elusive nature. You may get different diagnosis from different medical experts based on their clinical diagnosis. While some individuals may exhibit clear signs of TCS, others may not. Hence, being familiar with all potential symptoms is crucial. But often the signs can go un-noticed as is the case with difficult to diagnose occult tethered cord.

Occult Tethered Cord Syndrome (OTCS) is a condition in which the spinal cord becomes attached to the spine, preventing it from moving freely within the spinal canal. "Occult" in this context means "hidden," suggesting that the tethering may not be immediately obvious on standard imaging studies like MRIs and even harder to diagnose – and prove to health insurance providers the necessity of treatment or surgical intervention. Patients with hypermobile Ehlers-Danlos Syndrome (hEDS), can sometimes present with neurologic symptoms that might be consistent with OTCS, although the connection between hypermobility and tethered cord remains a topic of study and debate.

In hypermobile individuals, or those with EDS, who suspect they may have OTCS, the following symptoms might be observed:

Symptoms:

- **Pain**: Chronic back pain, especially in the lower back. This pain may worsen with activity or straining, walking or stair climbing
- **Muscle Weakness** or numbness in the lower limbs
- **Urinary issues** such as

    - Incontinence-trouble holding it
    - Urgency- feeling like you should go immediately
    - Frequency-going often
    - Retention-difficulty relieving yourself without pushing or leaning forward while seated

- **Muscle Spasms**: This can include leg or foot cramps or spasms
- **Gait Abnormalities**: Changes in the way a person walks, such as foot drop or a shuffling gait
- **Deterioration of Motor Skills**: This can manifest as clumsiness or coordination issues
- **Orthopedic Abnormalities**: Such as scoliosis or foot deformities

However, as always, diagnosing OTCS—especially in the context of another complex condition like hEDS—can be challenging. It requires a comprehensive clinical evaluation and collaboration between various specialists, which are very rare and challenging to get appointment with due to long wait times.

New Diagnostic Criterion Published for Occult Tethered Cord

Reference for Clinicians: Klinge P. et al. (2024). *Clinical criteria for filum terminale resection in occult tethered cord syndrome.* (Symptom scale includes 15 items across neurological, pain, and bowel/bladder domains.) https://pubmed.ncbi.nlm.nih.gov/38489815/

Patient Symptom Checklist for EDS & Occult Tethered Cord Syndrome (OTCS)

This 15-item symptom scale is derived from Klinge et al., 2024, which defines a structured clinical profile used to identify and evaluate occult tethered cord syndrome often occuring and unseen in patients with Ehlers-Danlos Syndrome (EDS) and related connective tissue disorders. Symptoms are grouped into Neurological, Pain, and Bowel/Bladder domains. Each item counts as 1 point if present.

SECTION 1 — NEUROLOGICAL (5 items)

Check all that apply.

☐ **Increased muscle tone in legs** (stiffness, rigidity)
☐ **Hyperreflexia** (brisk 3+ reflexes in legs)
☐ **Foot clonus** (rhythmic jerking when foot is flexed)
☐ **Leg fatigue** (fatigue disproportionate to activity)
☐ **Leg paresthesia** (tingling, numbness, burning)

SECTION 2 — PAIN (5 items)

☐ **Leg pain** (one or both sides)
☐ **Sacral pain** (tailbone/sacrum region)
☐ **Low-back pain** (lumbar)
☐ **Muscle cramps** (legs, feet and/or low back)
☐ **Fluctuating pain pattern** (variable intensity, episodic, spurts)

SECTION 3 — BOWEL & BLADDER (5 items)

☐ **Urinary leakage** (incontinence/dribbling)
☐ **Urinary urgency** (strong, sudden urge)
☐ **Urinary frequency** (urinating more often than normal)
☐ **Urinary hesitation** (difficulty starting flow)
☐ **Bowel symptoms** (constipation/obstipation and/or bowel incontinence)

If Determined You Need Clinical Evaluation The Medical Specialists to Consult

- **Neurologist**: Can provide an initial evaluation of symptoms and guide towards further diagnostic tests.

- **Neurosurgeon with TCS experience**: Not all neurosurgeons are familiar with the subtleties of TCS, especially occult tethered cord. It's essential to seek a neurosurgeon who has experience with this specific condition. There are very few hypermobile aware neurosurgeons in the US that can evaluate, diagnose, and treat TCS including a few experts listed below (not inclusive list):

    - Petra Klinge, MD
    - Paolo Bolognese, MD
    - Fraser C. Henderson, Sr., MD
    - Jeffrey Greenfield, MD
    - Justin Virojanapa, DO
    - Sunil Patel, M.D (no longer doing surgery)

Diagnostics Imaging and Testing Typically Ordered for Clinical Evaluation

- Magnetic Resonance Imaging (MRI): The first diagnostic step is typically a lumbar MRI. While tethering might not always be directly visible, its impact on the spinal cord can be seen. MRIs can be done supine (lying down) or prone (face down), or both help detect tethering due to the spinal cord's positioning when standing/lying down on your back.
  Note on MRIs: Sometimes radiological criteria are not met or are ambiguous, yet an Occult Tethered Cord (characterized by the presence of symptoms with normal conus position and inconclusive findings of the filum) can still exist. Other signs of tethering that might be visible in a lumbar MRI include an enlarged foramen magnum, thick or fatty filum, presence of fatty tissue inside the canal, or the filum might be pulling to one side of the canal. A prone MRI of the lumbar region can be an invaluable tool for those where other MRIs indicate that the filum might be pulling to one side (usually the back side) of the canal. With prone MRIs, imaging is done while the patient is lying face down on their stomach (as opposed to facing up, like most supine MRIs). It can be challenging to locate a facility doing prone positioning for MRIs.
- Other Diagnostic Tests:
    - Electromyogram (EMG) Test and Nerve Conduction Study (NCS): Recommended for thorough evaluation of the nerve function in the lower limbs.
    - Urodynamic Study: Important for analyzing the function of the bladder and its associated structures for any neurological changes. These invasive tests can determine if the patient has symptoms of a neurogenic bladder measuring the amount of liquid your bladder can hold measured at intervals on when your nerves signal releases.
    - Cervical MRI: This can show the spinal cord being abnormally tight and potentially show signs that a tethered cord might exist on the other end of the spine. The cervical spinal cord can sometimes be seen as narrow from it being pulled tight. Some experts now recognize CCI can contribute to tethered cord but no research has been conducted to date.

Patience is Key to Diagnosis

Even with the correct imaging and every symptom listed, patients are often told they do not have Tethered Cord. Therefore, it is important to make sure that you have the images viewed by a neurosurgeon that is familiar not only with TCS and EDS. The combination of the images and the patient's symptoms should tell the neurosurgeon if surgical intervention is required.

Tethered Cord Treatment Options

1. **Conservative Approach**: Physical therapy, pain management, and medications can help alleviate some symptoms. Some patients can get relief from temporary measures such as steroid or regenerative therapy injections (prolotherapy or PRP) in the short term. However, when your medical team have determined true tethering, surgical intervention usually becomes necessity.
2. **Tethered Cord Release (TCR) Surgery**: Some patients are ultimately needed to be surgically treated with a Tethered Cord Release, involving the untethering of the spinal cord. An incision is made in the lumbar area of the low back.

   Surgical treatment is not without risk and does not guarantee relief of symptoms. There is usually a prescribed minimum of 24 hours flat on your back immediately post op (procedure called a laminectomy) during your hospital stay to reduce potential cerebral spinal fluid (CSF) leaks.
   Most patients describe the surgery as a painful recovery for only the first two weeks and "better than they ever remember feeling" (often because they have been tethered for much of their lives). In a large study by the NIH, up to 83% of adult patients report relief, 16% unchanged, and 1% report feeling worse. In children, the numbers are even better with 93% obtaining improved symptoms and 7% unchanged. Another study concludes a success rate of 48-100%.

Note: Shunt placement: In cases where a cyst or syrinx develops, a shunt might be placed to drain excess cerebrospinal fluid.

Risks to Consider Before Tethering Release Surgery

Here's an outline of the risks and complications associated with Tethered Cord Release (TCR) surgery, particularly in EDS patients:

1. Retethering: This occurs when the spinal cord adheres *again* to its surrounding structures after being surgically released with a higher likelihood of poor wound healing and scar tissue formation.

2. Cerebrospinal Fluid (CSF) Leak: CSF leak happens when there's an unintentional opening in the dura mater, the protective covering of the brain and spinal cord, leading to leakage of the CSF.

3. Intracranial Hypertension (IH): A condition where the pressure inside the skull (intracranial pressure) is increased. It's not directly a complication of surgery but can be associated with conditions like CSF leaks, Chiari and hypermobile EDS.

4. General Surgical Risks in EDS:

- Poor wound healing: The skin of EDS patients may not heal as efficiently as others resulting sometimes in wounds reopening or healing with widened or atypical scars.
- Excessive bleeding: Abnormal connective tissue can make vessels more fragile, leading to increased intraoperative and postoperative bleeding.
- Anesthetic complications: There's some evidence to suggest that EDS patients might be at a higher risk for complications with certain anesthetics.

Closing

The determination of if you have tethered cord or not is definitely a clinical diagnosis made only by a trained medical professional with expertise in this area. While this article might provide some symptomatic overviews and guidance on the types of tests that may be ordered by your neurosurgeon, trust this level of care to the doctors who know best. Hold onto hope if you are seeking treatment because surgical intervention for TCS can offer pain relief, improved quality of life, and improvements in walking for many EDS patients.

Graciously reviewed and approved for publication by the nonprofit EDS Awareness on ChronicPainPartners.com by Dr. Pradeep Chopra and Dr. Sunil Patel. Written and researched by patient advocate Christie Cox.

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Unravelling Tethered Cord for EDS Patients

"Unravelling Tethered Cord for EDS Patients" delves into the intricate world of Tethered Cord Syndrome (TCS), a condition characterized by the abnormal stretching of the spinal cord that can lead to debilitating neurological symptoms and pain. This essential guide provides a thorough understanding of TCS's anatomy, diagnostic challenges, and the critical evaluations needed from specialists, especially for those with hypermobile Ehlers-Danlos Syndrome. With expert insights and practical resources, this book empowers patients to navigate their complex medical journeys and advocate for the care they deserve.