

Spinal Conditions in EDS: A Spine Surgeon's Perspective on Diagnosis and Management

Victoria Daylor, Molly Griggs, MEd, Cortney Gensemer, PhD &

Sunil Patel, MD

Chair, Department Of Neurosurgery

Medical University Of South Carolina

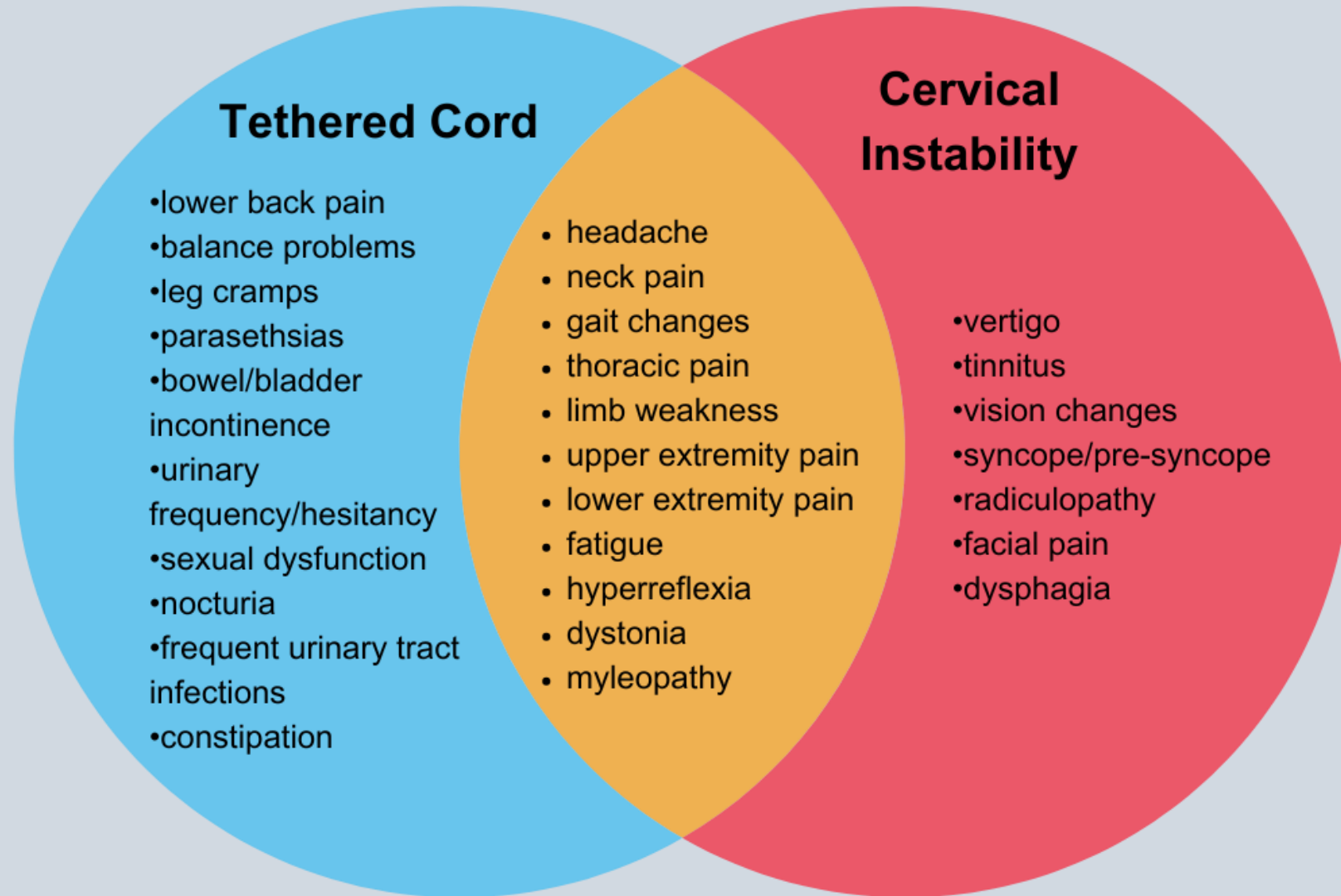
Understanding CI and TCS in hEDS

	Cervical Instability (CI)	Tethered Cord Syndrome (TCS)
Cause	Ligament laxity or trauma	Inelastic or thickened filum terminale restricting spinal cord movement
Types	Craniocervical instability (CCI): skull–C1 Atlantoaxial instability (AAI): C1–C2 Lower cervical instability (CI): below C2	Classical TCS (Type I): low conus, fatty/thickened filum Occult TCS (Type II): normal imaging, clinical diagnosis
Prevalence (hEDS)	~31.6% ¹	~6.7% (likely underdiagnosed) ¹
Diagnosis	Upright dynamic imaging + clinical correlation	Clinical evaluation ± imaging (MRI), may require urodynamic studies
Imaging Notes	Imaging may be normal despite instability	Imaging often normal in occult TCS
Treatment	Conservative: collar, rest, isometric PT Surgical: cervical or occipito-cervical fusion	Surgical: laminectomy and FT transection

1. Petrucci, T., Barclay, S. J., Gensemer, C., Morningstar, J., Daylor, V., Byerly, K., Bistran, E., Griggs, M., Elliot, J. M., Kelechi, T., Phillips, S., Nichols, M., Shapiro, S., Patel, S., Bouatia-Naji, N., & Norris, R. A. (2024). Phenotypic Clusters and Multimorbidity in Hypermobile Ehlers-Danlos Syndrome. *Mayo Clinic proceedings. Innovations, quality & outcomes*, 8(3), 253–262. <https://doi.org/10.1016/j.mayocpiqo.2024.04.001>

Overlapping Symptoms & Neurological Findings

Gensemer, C., Daylor, V., Nix, J., Norris, R. A., & Patel, S. (2024). Co-occurrence of tethered cord syndrome and cervical spine instability in hypermobile Ehlers-Danlos syndrome. *Frontiers in neurology*, 15, 1441866. <https://doi.org/10.3389/fneur.2024.1441866>



Case Study

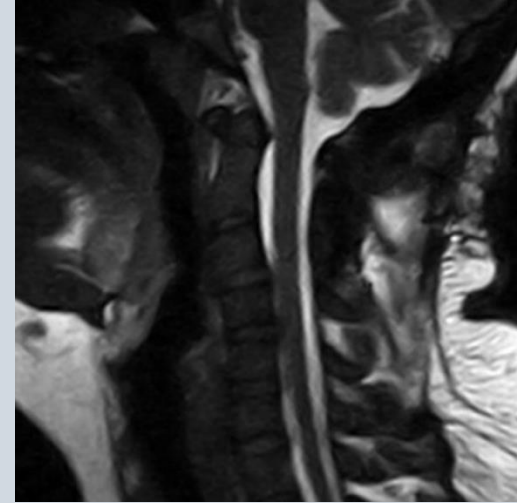
Female, 20's, with hEDS & POTS

Initial Presentation (CCI):

- Daily suboccipital headaches, neck pain, leg weakness, gait imbalance
- Supine MRI normal; clinical exam showed signs of myelopathy
- Temporary relief with rigid cervical collar
- Upright MRI confirmed CCI (clivo-axial angle 116° + medullary kinking)
- Underwent C0–C3 occipito-cervical fusion
- Post-op: gradual improvement in pain, POTS, and mobility
- Returned to college, graduated, began work as social worker



Preop medullary kinking



Postop reduced kinking



Slightly low-lying conus

Later Presentation (TCS):

- Four years later: New BLE numbness, pain, and urinary retention
- Longstanding urinary symptoms since childhood; neurogenic bladder confirmed
- MRI: Conus at mid-L2 (borderline), consistent with occult TCS
- Underwent L1–L2 laminectomy and FT resection

Post-Tethered Cord Release:

- 1 month: Improved urinary function and paresthesias
- 3 months: Discontinued catheter use, returned to work
- 6 months: 90% resolution of urinary symptoms, no back pain

Key Takeaways

- **Spinal instability and TCS are underdiagnosed in hEDS**, often missed by standard imaging and complicated by symptom overlap.
- **Dynamic upright imaging and detailed clinical history are critical** for identifying occult presentations, especially when standard MRI appears normal.
- **Surgical outcomes for TCS and CCI in hEDS are generally positive**, with TCS release often leading to significant symptom improvement and lower morbidity compared to fusion.
- **Treating TCS before cervical fusion may reduce symptom burden** or eliminate the need for fusion in select cases.
- **hEDS patients with spinal manifestations show unique pathophysiology**—including ligament laxity, altered FT structure, and inflammatory changes—highlighting the need for tailored diagnostic and treatment approaches.

Citations

Geneser, C., Daylor, V., Nix, J., Norris, R. A., & Patel, S. (2024). Co-occurrence of tethered cord syndrome and cervical spine instability in hypermobile Ehlers-Danlos syndrome. *Frontiers in neurology*, 15, 1441866. <https://doi.org/10.3389/fneur.2024.1441866>

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