Neuro-Cranio-vertebral Syndrome and Ehlers-Danlos Syndrome

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Introduction

There has recently been a trend to associate Ehlers-Danlos syndrome (EDS) with vertebral instability, believed to cause clinical and imaging changes that justify extensive cranio-cervical fusion. We propose a different line of argument, in which the relationship between the spinal manifestations and the clinical picture is based on an abnormal caudal traction of the central nervous system **(Fig 1).** This phenomenon is called the Neuro-Cranio-vertebral Syndrome (NCVS).

Methods

We conducted a retrospective study from the clinical records in our institution between 2015 and 2019 and selected 10 patients with a diagnosis of EDS and NCVS. We presented their images, signs and symptoms, and compared them to those seen in 373 patients with only NCVS. From the 10 patients with EDS, we report the postsurgical findings of 4 patients that underwent sectioning of the filum terminale (SFT), including significant amelioration in signs and symptoms ($p < 0.05$).

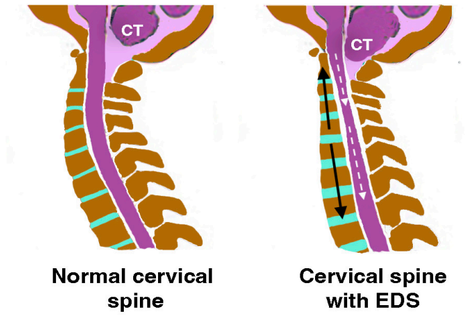

Figure 1. Illustration of the causative effect of EDS on Filum Disease. Black arrows: spinal elongation due to EDS. White arrows: abnormal caudal traction of the central nervous system. (CT=Cerebellar tonsils)

Results

We found that the 10 patients with EDS have more cranial and vertebral symptoms than patients with NCVS and also significant differences in the signs.

The patients that underwent SFT experienced significant improvements in signs and symptoms **(Fig 2)**.

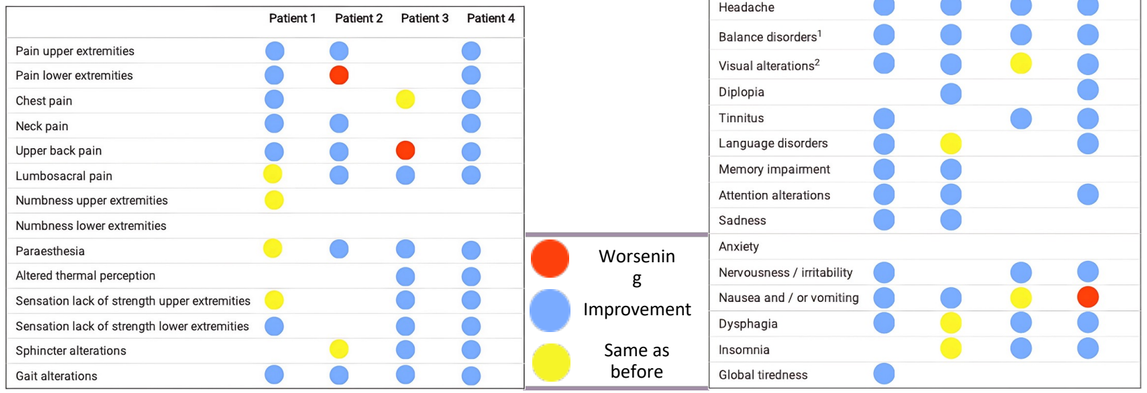

Figure 2. Evolution of vertebral symptoms in 4 patients diagnosed with EDS and NCVS/Filum Disease after SFT.

Discussion

This study offers new insights into the neurological symptoms of EDS, proposing an axial intramedullary tension mechanism as an alternative to cranio-cervical instability. Cervical subluxations in EDS are rare and typically linked to bone malformations. Similarly, Arnold-Chiari syndrome type I is a recognized comorbidity, with unique imaging findings in EDS patients.

We suggest that persistent tissue stress in the brainstem and spinal cord, rather than instability at movement extremes, explains many EDS symptoms. These include pain, fatigue, dysautonomia, headaches, cognitive issues, and peripheral neuropathies, all of which significantly affect daily life.

EDS-related connective tissue elasticity may prevent severe spinal deformities but allows gradual traction on the spinal cord, leading to unique neurological presentations. Some patients also experience CDP, an obsessive fear of spontaneous dislocation.

SFT shows promise in symptom relief, though further studies are needed to validate these findings and better understand EDS-related neurological mechanisms

Objectives

1. To analyze clinical and imaging findings in 10 EDS patients with NCVS and compare them to a control group.
2. To propose a mechanism explaining NCVS development in EDS patients.
3. To evaluate postoperative outcomes after SFT in these patients.
4. To highlight the psychological impact, specifically "cervical dislocation panic syndrome," (CDP) observed in some patients.

Conclusion

EDS and NCVS/Filum Disease Connection: Among 373 cases of NCVS and/or Filum Disease, 10 cases suggest EDS may play a causative role in FD.

Surgical Outcomes: SFT in four NCVS-EDS patients showed improvement of the clincal picture.

Reevaluation of Instability Theories: Symptoms attributed to cranio-cervical or atlanto-axoid instability in EDS may instead reflect NCVS. SFT should be the only option considered when encountering a patient with EDS.

Addressing Cervical Dislocation Panic: Clear communication with EDS patients about the distinction between cervical spine hypermobility and dislocation can reduce unnecessary panic and avoid misdiagnoses of cervical vertebral dislocation.

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