Study Identifies Abnormal Filum Terminale as Cause of Tethered Cord Symptoms in hEDS

A comprehensive study from Brown University has identified an abnormal filum terminale as the likely cause of tethered cord syndrome (TCS) symptoms in patients with hypermobile Ehlers-Danlos Syndrome (hEDS). hEDS is a heritable connective tissue disorder which affects 10% or more of Chiari patients. Many hEDS patients have symptoms that are similar to tethered cord, such as leg/back pain, leg weakness and urinary/bowel issues. However, unlike traditional TCS, for many hEDS patients, there is no MRI indication of tethered cord. For example, in traditional TCS, the conus medullaris – the tapered, lower end of the spinal cord – is often noticeably lower on MRI relative to the lumbar spinal vertebrae. In other cases the filum terminale – a thin strand of connective tissue between the conus and the tailbone – has visible fat composition on MRI. But for hEDS patients with TCS symptoms, there is often nothing on MRI to indicate a problem. This has led to controversy on how to treat these patients, with some surgeons choosing to operate based on symptoms, but others arguing against this.

To address this controversy, the researchers compared 78 patients with hEDS and TCS type symptoms (but no MRI findings) to 38 patients with typical TCS (MRI findings). The hEDS group was carefully screened and diagnosed by an experienced geneticist. Both groups underwent surgery to remove a small (2cm) section of the filum terminale. The group then examined the removed filum samples under a microscope, tested their elastic properties, and compared the surgical outcomes of the two groups. Since hEDS affects collagen structures, the group hypothesized that the structure of the filums for those patients would show abnormalities and thus justify surgical intervention to treat their symptoms.

Under the microscope, the team found disorganization of the collagen fibers in nearly 70% of the hEDS patients compared to only 10% of the typical TCS group. In addition, they found indications of damage to the collagen fibers in around 2/3 of the hEDS group compared to less than 1/3 of the TCS group. Next, they tested the elastic properties of the filum samples and found that the elasticity of the hEDS samples failed under about half the force of the TCS group. Taking the two findings together, the authors suggest that in a healthy person the filum terminale shields the nerves in the conus from mechanical stress through its ability to stretch. However, in hEDS patients, the abnormal collagen structure of the filum reduces this ability and results in damage to the filum and exposes the nerves in the conus to stress, leading to TCS symptoms.

Finally, the group found that the surgical outcomes for the two groups were very similar in terms of symptom improvement. While many patients experienced a significant improvement in symptoms, about 13% of each group had either a temporary or permanent worsening of symptoms.

Source: Diseased Filum Terminale as a Cause of Tethered Cord Syndrome in Ehlers-Danlos Syndrome: Histopathology, Biomechanics, Clinical Presentation, and Outcome of Filum Excision. Klinge PM, Srivastava V, McElroy A, Leary OP, Ahmed Z, Donahue JE, Brinker T, De Vloo P, Gokaslan ZL. World Neurosurg. 2022 Mar 17:S1878-8750(22)00329-1. doi: 10.1016/j.wneu.2022.03.038. Online ahead of print. PMID: 35307588

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