

An Introduction of Ehlers-Danlos Syndrome for the CST Practitioner

publication date: Dec 1, 2016 | author/source: Eloise Stager BA, LMT, CST

An Introduction of Ehlers-Danlos Syndrome for the CST Practitioner

by

[Eloise Stager, BA, LMT, CST](#)

Introduction

Ehlers-Danlos Syndrome (EDS) is a genetic, connective tissue disorder affecting collagen production. Since collagen is found throughout the body, all systems, structures and biomechanics can be affected. The most noticeable physical manifestation is joint hypermobility as described by the Beighton Criteria (6), a scoring system for joint hyperflexion and hyperextension (see fig 1). There are six major types of EDS recognized by the Ehlers-Danlos Society (3): Classical, Hypermobility, Vascular, Kyphoscoliosis, Arthrochalasia, and Dermatosporaxis. The severity of symptoms varies among the types and among individuals. The most common type, EDS-Hypermobility Type (EDS-HT), is the focus of this paper. EDS-HT is autosomal dominant, meaning that only one parent with Ehlers-Danlos Syndrome is needed to pass an affected gene to their children which will express itself with varying severity per individual. Unfortunately, there has not yet been a gene coded for Hypermobility Type, but some promising new research has been published in which multiple copies of the alpha-tryptase gene (TPSAB1) have been found in some individuals with a compilation of autonomic nervous system dysfunction (dysautonomia), mast cell activation disorder and hypermobility (11). Symptoms of EDS often worsen with the onset of puberty and tend to progress throughout life. Currently, there is no cure for Ehlers-Danlos Syndrome and conventional treatment is focused on palliative care, physical therapy and pain management.

Diagnostic Criteria

Diagnostic criteria for EDS-HT is an ongoing, international debate among specialists but includes clinical hypermobility assessments using the Beighton criteria, family history and whether or not there is multi-system involvement (12). Unfortunately, the Beighton Criteria is a flawed scoring technique, as it ignores hypermobility in the shoulders, knees, small finger joints, ankles, sacrum, vertebrae and circumferential

stability in the hips. In addition it does not consider mediolateral instability, gender or age. Women and children tend to be more hypermobile than men and decreased flexibility increases with age (6,7). Proper diagnosis is often significantly delayed until adulthood and patients are more likely to have spent a lifetime labeled as a hypochondriac, or misdiagnosed with chronic fatigue syndrome, fibromyalgia, multiple sclerosis, conversion disorder, anxiety, and/or depression.

In this author's opinion, there is an overwhelming lack of education about EDS for doctors and medical personnel. Personal discussions with colleagues and doctors about their individual training and knowledge of EDS-HT have been disappointing. In this author's experience, many doctors freely admit that their medical school education did little to prepare them or educate them about connective tissue disorders. This educational vacuum heavily contributes to the reasons EDS is not as well-known as it should be and why doctors are reluctant to diagnose and treat it, placing patients at risk, and prolonging the potential suffering.

Symptoms and Complications

The name, EDS-Hypermobility Type, is misleading. The joints, are of course affected, but so is every system in the body due to the lack of collagen. These include the structure and function of the craniosacral system, autonomic and central nervous systems, sensory organs, proprioception, cardiovascular system, respiratory system fascial system, integumentary system, gastrointestinal and reproductive systems, lymph system - EVERY SYSTEM is affected (10). Additionally, there is some overlap with mitochondrial disease and possibly ion channel disorders (channelopathies).

One of the primary and most debilitating symptoms of EDS is widespread, unrelenting and varying pain, with episodes of acute, excruciating pain when there is a direct joint injury, subluxation or dislocation. For many, these occur daily with mild physical impact or sometimes no impact at all. Consequently, the ligamentous laxity in the joints causes compensatory muscle recruitment for additional stabilization, which leads to muscle fatigue and small muscle fiber spasm, causing full body pain and exhaustion. Many people with EDS-HT require assistive devices to stabilize joints. Bruising or mild bleeding tendencies are common and variable due to the fragility of the venous structures; the skin is often soft, mildly stretchy, and delicate.

Of importance is the direct involvement of the central nervous system. Dysautonomia or autonomic nervous system dysfunction, is estimated to occur in varying degrees in 100% of people with EDS. It affects both the sympathetic and

parasympathetic branches with a tendency towards sympathetic hyperactivity. Patients may experience heart rate, blood pressure and temperature dysregulation, pre-syncope, syncope (fainting), gastroparesis (slow digestion), Reynaud's syndrome, anxiety/depression, poor sleep/insomnia, and trouble concentrating.

It is unclear why the autonomic nervous system is affected. Dysautonomia occurs in other illnesses as well, however, due to the high percentage of collagen in the dural system, we may be able to extrapolate a craniosacral model (1) as to why the central nervous system is jeopardized. EDS causes laxity in the dural membrane reducing structural integrity of the falx cerebri, falx cerebelli, tentorium, cauda equinae, filum terminale, cranial nerves, cerebrum, cerebellum, spinal cord, brain stem, ventricular and intracranial vascular systems, nerve roots, cerebral spinal fluid circulation, and general dural tensesgrity. Several conditions may develop due to a loss of dural integrity: Chiari 1 Malformation, dural ectasia, cerebral spinal fluid leaks, tentorium slumping, intracranial hypertension, nerve impingement, and venous and vascular drainage issues.

Depression and anxiety can be primary results of autonomic nervous system dysregulation, rather than secondary to the burden of the disorder (13). As expressed previously, the autonomic nervous system regulates and controls appropriate response and function of the parasympathetic and sympathetic branches of the ANS. Interference of proper signaling between the sensory and motor neurons of the ANS may contribute to a heightened state of sympathetic activity which may trigger a flight or fight response, often interpreted as a panic attack. The result for the patient is very often a misdiagnosis of an anxiety disorder.

The person with EDS is often erroneously labeled as depressed at a young age and prescribed a host of medications which are ineffective. In prepubescent children, Ehlers-Danlos symptoms may originally manifest as panic attacks, behavioral outbursts (due to sympathetic nervous system hyperactivity), poor proprioception, reduced muscle tone, early repetitive injuries and joint pain. Although hypermobility may be present, it is frequently overlooked. The adult with EDS is often prescribed opioid based medications and are at risk to be flagged as "drug seekers" or addicts as their pain increases despite pain medication, or perhaps, because of it. Alternative and Complementary medicine is cost prohibitive for most people with EDS, despite the benefits.

People with EDS are tired. Tired physically, emotionally, and spiritually. They are depleted in every aspect of their physical and emotional self. They are faced with the knowledge that there is no cure or effective treatment for their condition.

Hopes and dreams of having a normal life with normal activities are dashed and they frequently lose the ability to do activities that they once enjoyed. Their calendars are filled with doctor's appointments and medical tests. The young EDS patient faces the disheartening knowledge that they will likely live in pain for their entire lives and activities that their friends can do will not be possible for them to do. Fatigue and pain will control the activities of the day which will be modified to sustain energy for the next day... and the next. The person with Ehlers-Danlos has a very, very difficult life. For some, they cannot withstand the pain and uncertainty of their situation and end their lives prematurely. For others, they hold their heads high and bravely face their pain and fatigue every day.

Possible Presentation in the CranioSacral Clinic

Many people with EDS may not realize they have it. They may present to the craniosacral therapist with an extensive and complicated medical history with their primary complaint being chronic pain. They may have been diagnosed with fibromyalgia or chronic fatigue syndrome and prescribed several medications; they will likely have fatigue, anxiety and depression. A clinic intake form which includes health history questions for each system, injury history and surgical history will likely yield impressive but daunting results. It is especially important to note any history which includes repetitive joint injuries, subluxations, dislocations, fainting, cardiovascular abnormalities (such as mitral valve prolapse and aortic root dilation), general chronic pain, gastrointestinal disturbances, headaches and unusual visual symptoms (retinal detachment, nystagmus, corneal injury, or pupil dislocation). Brain injury, brain fog, trouble concentrating, and poor memory occur frequently. Numbness and tingling in extremities, nerve pain, Chiari 1 Malformation, craniocervical instability, and small fiber neuropathy are common. They may have or have had Complex Regional Pain Syndrome (CRPS), immune system dysfunction, and mast cell activation disorder. They may be on state medical assistance and unable to work and need assistive mobility devices.

Observations at CT Center for CranioSacral Therapy

Head Trauma - Based on my experience with EDS, as a patient, mother and therapist, I have observed unique characteristics of the EDS patient with regards to CranioSacral Therapy and have developed theories to explain them. Possibly the most thought provoking, is the common occurrence of post-concussive syndrome and/or brain injury¹⁷, without significant physical trauma to the head. I believe that *intracranial dural laxity allows for brain injury with mild trauma, and, repetitive mild trauma compounded over time, leads to a condition similar to, if not specifically,*

Chronic Traumatic Encephalopathy (15). I welcome an investigation to prove or disprove this theory.

Proprioception - Poor proprioception is common in EDS. It is widely accepted that proprioceptive awareness takes place in the large muscle groups and is a biomechanical system. Recent research has demonstrated that proprioception is perhaps, primarily sensed in the subcutaneous and myo-fascia rather than in deep muscle fibers. With this new paradigm, it is an appropriate consideration to attribute laxity of the fascia as the primary reason for poor proprioception in Ehlers-Danlos. Interestingly, this concept seems to extend into the "personality" of fascia itself, which can lead to unintended hyperextension in the tissue with manual therapy. The craniosacral concept of "Follow the tissues" to its end range may be detrimental to someone with EDS-HT as the proprioceptors in the fascia do not seem to register an appropriate end-point. At the time, the patient feels a sense of relief and reports that it feels good, only to recognize later that a strain or muscle spasm has occurred. Through experience, I have learned that a slight drag on the fascia increases its self-awareness and can reduce accidental hyperextension in the fascia. I am eager for further experiential observation by other craniosacral therapists as this information is on the leading edge of CST technique discovery to benefit those with Ehlers-Danlos Syndrome. The "drag" yields a similar result to weighted jackets for children with sensory processing dysfunction or when adding resistance to fast, repetitive movements seen in Regional Tissue Release. I suspect that the weight of the drag improves proprioception and alignment to the fine fibers of the fascia.

Hypermobility in the MindBodySpirit Triad– I have attempted to establish throughout this paper that the ever present symptoms in people with EDS-HT are generalized hypermobility and pain. This includes the structure (s) of MindBodySpirit. Consider this triad to be an integrated system, with the Inner Wisdom (IW) at the helm; for people with EDS-HT, this system appears to be hypermobile as well. It has been my clinical observation that BodyMindSpirit and IW do not integrate easily in people with EDS-HT. Each part of the triad has unique aspects which could complement each other, but instead "flop" against each other, inhibiting a tightly braided, integral system of wholeness, much like how EDS inhibits musculoskeletal and visceral structures. The client literally feels "all over the place". Pain is sensed within the triad as physical, emotional and spiritual with sparse reserves to rectify it. Despondency ensues and leads to further physical pain perpetuating the pain/depression cycle.

What Can You Do?

As craniosacral therapy practitioners, we have varied backgrounds as physical therapists, occupational therapists, massage therapists, psychologists, doctors, osteopaths, chiropractors, naturopaths, acupuncturists, rolfers, etc. We are in a unique position through education and training to recognize a connective tissue disorder in our clients and help them find resources to receive a diagnosis. If a patient presents with a known history of multi-system involvement, CRPS, Chiari 1 Malformation, Mast Cell Activation Disorder (hyper-allergic response), Gastroparesis (slow gastric motility), and/ or Postural Orthostatic Tachycardia Syndrome (rapid heart rate upon standing with feelings of faintness), they should be evaluated for hypermobility. These conditions, in particular, should be considered red flags. If the Beighton score is two or higher out of a total score of nine (2/9), and some of the above symptoms are present, please refer to a specialist. Proper care and treatment of EDS is imperative. Every person with EDS, regardless of type is strongly recommended to have a cardiac and neurological exam by a doctor knowledgeable in EDS. It is also important to note that **dysautonomia, mast cell activation disorder, Chiari 1 malformation and cardiac conditions occur more frequently than not.**

There are practical technique modifications that should be exercised depending on the symptoms presented. If Chiari 1, tethered cord or craniocervical instability are diagnosed in your client, then the following should be avoided: stillpoint induction; inferior and superior traction of the dural tube; the platform step of occipital base release; and intentional cranial compression (compression/decompression). Listen intently to your client; both physically and intuitively. Be an astute observer of the craniosacral rhythm, fascial glide, energy cysts, facilitated segments, intracranial dural restrictions, sphenobasilar lesions and dural releases. The client with EDS will likely have a very unique craniosacral therapy "feel" to their systems and structures. Heed fascial glide and cautiously approach "end of range" mobility in all structures, including the triad of mindbodyspirit. Concurrent physical therapy for core stabilization has been shown to be beneficial.

As therapists, we owe it to this group of individuals to listen, understand, respect, and assist in appropriate diagnosis and treatment. A dear friend and colleague said to me that EDS is similar to what fibromyalgia and Lyme disease were 10 years ago; a medical condition that no one believes in, but is real, emerging and medically important to understand. It is my hope that within five years Ehlers-Danlos Syndrome becomes well known and understood by all; that a genetic model is developed; and more importantly, a cure is found.

Resources/Bibliography

- (1) Upledger, J. E. (1997). *Your inner physician and you: Craniosacral therapy and somatoemotional release*. Berkeley, CA: North Atlantic Books.
- (2) Upledger, J. E. (1996). *Somatoemotional release and beyond*. Palm Beach Gardens, FL.: UI Pub.
- (3) Ehlers-Danlos Society, www.Ehlers-Danlos.com
- (4) Chronic Pain Partners, www.EDSAwareness.com
- (5) Dysautonomia International, www.DysautonomiaInternational
- (6) Beighton criteria. (2009). *Dictionary of Rheumatology*, 24-24. doi:10.1007/978-3-211-79280-3_114
- (7) Parvaneh, V. J., & Shiari, R. (2016). Proposed modifications to Beighton criteria for the diagnosis of joint hypermobility in children. *Indian Journal of Rheumatology*. doi:10.1016/j.injr.2016.03.009
- (8) Muldowney, K. (2015). *Living life to the fullest with Ehlers-Danlos syndrome: A guide for a person living with EDS to achieve a better quality of life*. Denver, CO: Outskirts Press.
- (9) Guimberteau, J. C., & Armstrong, C. (2015). *Architecture of human living fascia: The extracellular matrix and cells revealed through endoscopy*. Edinburgh: Handspring Publishing.
- (10) Ehlers-Danlos syndrome - Genetics Home Reference. (n.d.). Retrieved November 28, 2016, from <https://ghr.nlm.nih.gov/condition/ehlers-danlos-syndrome>
- (11) Alpha-Tryptasemia (n.d.). Retrieved November 28, 2016, from <https://www.niaid.nih.gov/research/hereditary-alpha-tryptasemia-faq>
- (12) Grahame, R., MD. (n.d.). *EDS Past, Present and Future*. Retrieved from <http://ehlers-danlos.com/2015-annual-conference-files/Grahame.pdf>
- (13) Eccles, J. A., Owens, A. P., Mathias, C. J., Umeda, S., & Critchley, H. D. (2015). Neurovisceral phenotypes in the expression of psychiatric symptoms. *Frontiers in Neuroscience*, 9. doi:10.3389/fnins.2015.00004
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4322642/>
- (14) *The Midbrain, Pons, Medulla, and Reticular Formation*. (n.d.). Retrieved November 28, 2016, from <http://www.csuchico.edu/~pmccaffrey/syllabi/CMSD320/362unit6.html>
- (15) Omalu, B. (2014). Chronic Traumatic Encephalopathy. *Progress in Neurological Surgery Concussion*, 38-49. doi:10.1159/000358761

(16) Wortzel, H. S., Brenner, L. A., & Arciniegas, D. B. (2013). Traumatic Brain Injury and Chronic Traumatic Encephalopathy: A Forensic Neuropsychiatric Perspective. *Behavioral Sciences & the Law*, 31(6), 721-738. doi:10.1002/bsl.2079

(17) Traumatic Brain Injury: Hope Through Research. (n.d.). Retrieved November 28, 2016, from http://www.ninds.nih.gov/disorders/tbi/detail_tbi.htm

Figure 1: Assessing Hypermobility: Beighton's Criteria

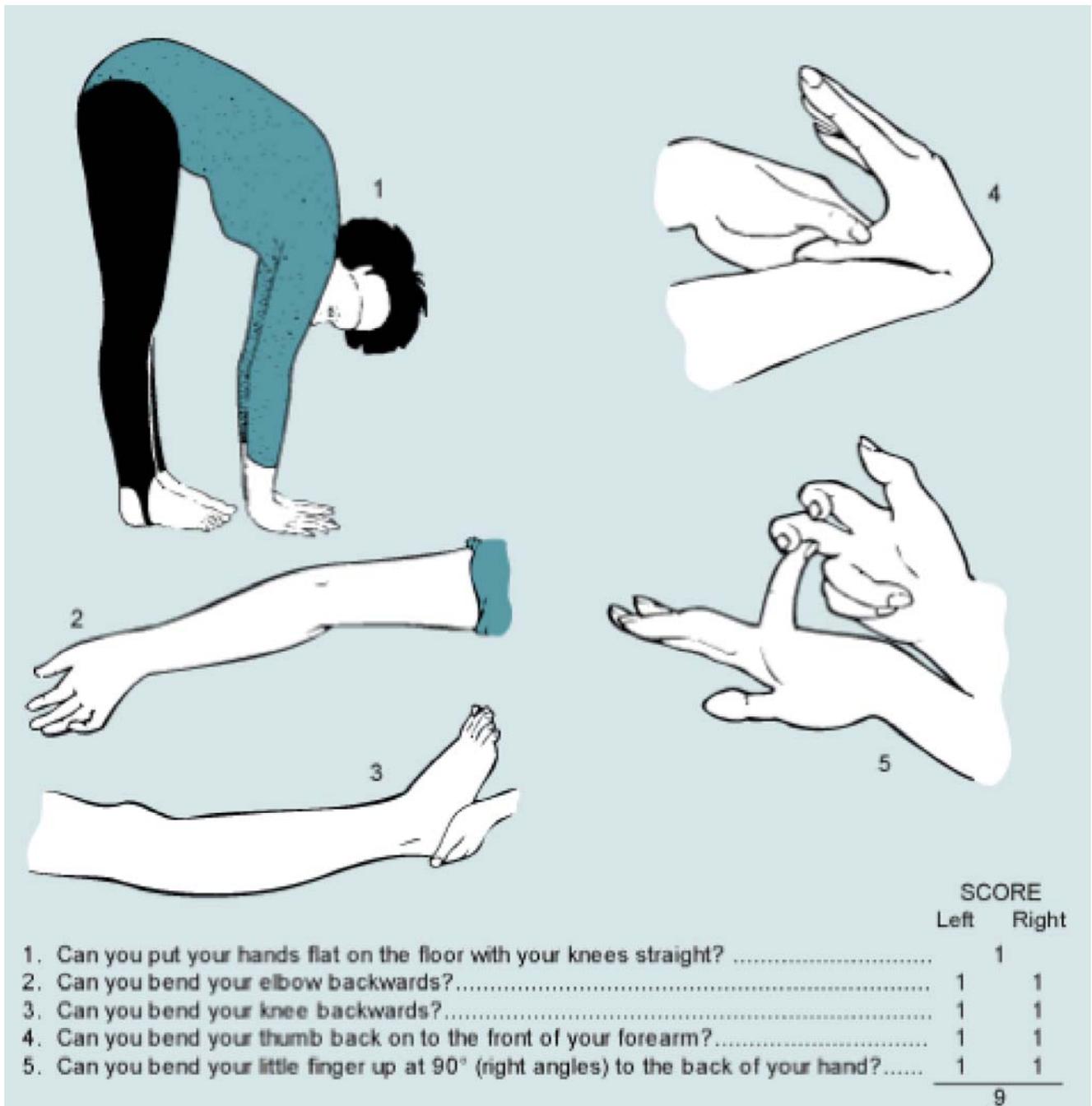


Figure 1. Beighton's modification of the Carter and Wilkinson scoring system. Give yourself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points.

Related Articles:

- [Mission](#)
- [Eloise Stager](#)