

EDS and Family: How to Welcome Them to Your Practice



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Learning Objectives

1. Identify clinical signs/symptoms of Ehlers-Danlos Syndrome (EDS) and some of its associated conditions.
2. Describe the role of central sensitization in EDS-related pain.
3. For a given patient, recommend a management plan.

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EDS is a rare inherited disorder.....



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GG is a 45 year old woman who presents with chronic widespread musculoskeletal pain, fatigue, headaches, and fainting. She has many allergies and does not tolerate many medications. She is at the point that she thinks she will have to quit her job as she cannot stand for several hours in the retail setting without incapacitating pain. By the time she gets home from work she just goes to bed due to exhaustion.

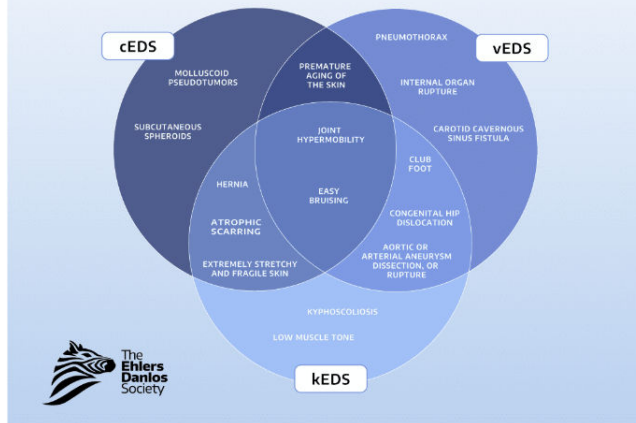
Her history includes several shoulder and knee dislocations dating back to her teen years when she was a dancer and cheerleader. She has had to have these reduced in the ED in the past. She notes widespread muscle pain in her shoulder girdle and upper back, very sensitive to touch. She feels that several other joints are “loose” and she also has chronic low back pain. She notes that her mother and grandfather are both also “double jointed”.

Does she have EDS?

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THE EHLERS-DANLOS SYNDROMES (EDS)

EACH TYPE OF EDS HAS ITS OWN COMBINATION OF SIGNS AND SYMPTOMS. SOME SIGNS AND SYMPTOMS ARE COMMON IN MULTIPLE TYPES OF EDS, WHILE OTHERS ARE ONLY ASSOCIATED WITH ONE SPECIFIC TYPE.



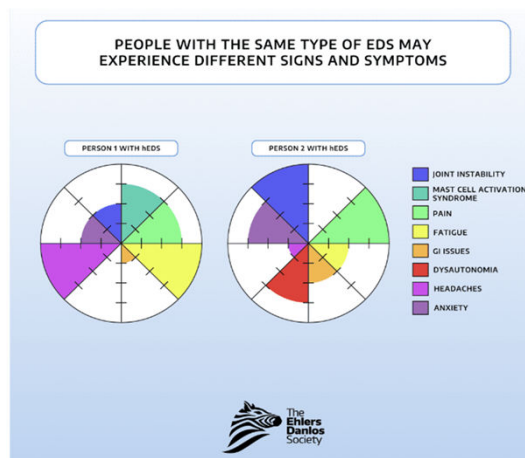
- 13 different types
 - Hypermobile EDS (hEDS)
 - Autosomal dominant
 - Gene(s) unknown
 - Classical EDS (cEDS)
 - Autosomal dominant
 - COL5A1, COL5A2, COL1A1
 - Vascular EDS (vEDS)
 - Autosomal dominant
 - COL3A1, COL1A1
 - Kyphoscoliotic EDS (kEDS)
 - Autosomal recessive
 - PLOD1, FKBP14
- Overlapping symptoms but almost all types of EDS have some degree of joint hypermobility, tissue fragility and skin hyperextensibility as well as fatigue and commonly pain

<https://www.ehlers-danlos.com/what-is-eds/>; Malfait F, et al. The 2017 International classification of the Ehlers-Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C:8-26

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Other Members of the EDS “Family”

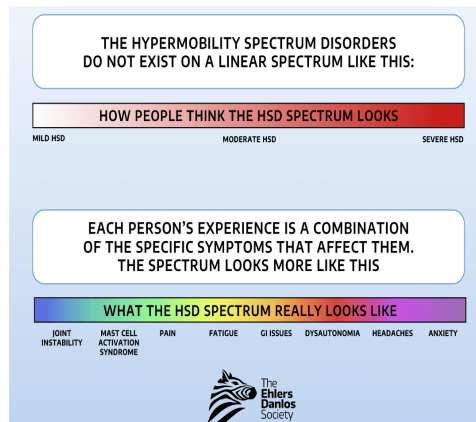
Condition	EDS only	EDS+SpA
MCAS	85.7%	13%
POTS	78.6%	60.9%
IBS	85.7%	60.9%
Cervical instability	21.4%	17.5%



<https://www.ehlers-danlos.com/what-is-eds/>; Wang TJ, et al. Acta Orthopædica Mexicana 2026 Mar-Apr;40(20):85-88

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Hypermobility Spectrum Disorders



Types of HSD

Generalized (G-HSD)

Peripheral (P-HSD)

Localized (L-HSD)

Historical (H-HSD)

The family

Hypermobility spectrum disorders (HSD) are also characterized by joint hypermobility, instability, injury and pain as well as possible fatigue, headaches, GI problems and autonomic dysfunction.

<https://www.ehlers-danlos.com/what-is-hsd/>

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Back to GG

1. How do we diagnose her EDS?
2. Should we send her for genetic testing?
3. Refer to a specialist?

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 **THE BEIGHTON SCORING SYSTEM**
Measuring joint hypermobility

A positive Beighton score is any score **greater than or equal to**:

- **6/9** points in **children before puberty**
- **5/9** points from **puberty up to the age of 50**
- **4/9** points for those **50 years of age and over**

A. 5th FINGER / PINKIES
Test **both sides**: Rest palm of the hand and forearm on a **flat surface** with the palm side down and fingers out straight.
Can the **fifth finger** be bent/lifted upwards at the knuckle to go back **beyond 90 degrees**?
If yes, add **one point** for each hand.

B. THUMBS
Test **both sides**: With the arm out straight, the palm facing down, and the wrist then fully bent downward, can the thumb be pushed back to touch the forearm?
If yes, add **one point** for each thumb.

C. ELBOWS
Test **both sides**: With arms outstretched and palms facing upwards, does the elbow extend (bend too far) upwards **more than an extra 10 degrees** beyond a normal outstretched position?
If yes, add **one point** for each side.

D. KNEES
Test **both sides**: While standing, with knees locked (bent backwards as far as possible), does the lower part of either leg to extend more than 10 degrees forward?
If yes, add **one point** for each side.

E. SPINE
Bend forward, can you place the palms of your hands **flat on the floor in front of your feet without bending your knees**?
If yes, add **one point**.

<https://www.ehlers-danlos.com/what-is-hsd/>

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Postural Orthostatic Tachycardia Syndrome (POTS)

SYMPTOMS

- Dizziness/lightheadedness
- Syncope
- Fatigue
- Brain fog
- Dyspnea
- Nausea, bloating, vomiting
- Headache
- Palpitations/tachycardia
- Tachycardia
- Tremor

MANAGEMENT

- Compression garments
- Exercise and physical activity (low impact and isometric exercises)
- Increase sodium (up to 10,000 mg/day)
- Increase water intake
- Avoid alcohol and caffeine
- Medications: midodrine, fludrocortisone, beta-blockers

<https://www.ninds.nih.gov/health-information/disorders/postural-tachycardia-syndrome-pots>

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Mast Cell Activation Syndrome (MCAS)

SYMPTOMS

- Hives/urticaria
- Flushing
- Angioedema
- Rash
- Itching
- Nausea/vomiting/diarrhea
- Bloating/abdominal pain
- Wheezing/dyspnea
- Nasal congestion
- Coughing

Must have at
least 2 of
these

MANAGEMENT

- Antihistamines
- Leukotriene modifiers
- Mast cell stabilizers (cromolyn)
- Epi-pen
- Omalizumab

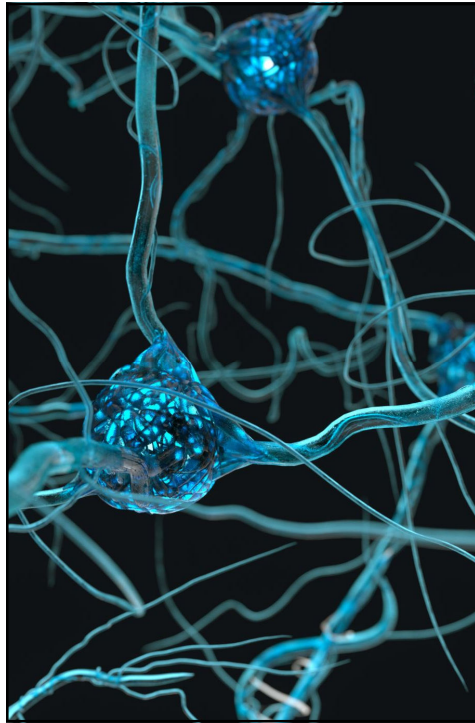
<https://www.aaaai.org/conditions-treatments/related-conditions/mcas>

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GG scored 6/9 on the Beighton scale but she has diffuse muscle pain as well as joint pain and back pain.

Why does she have extra-articular widespread pain?

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Central Sensitization

Augmentation of responsiveness of central neurons to input from unimodal and polymodal receptors.



- Altered sensory processing in the brain, malfunctioning of descending anti-nociceptive mechanisms, increased activity of pain facilitatory pathways, long term potentiation of neuronal synapses in the ACC



- Increased responsiveness of dorsal horn neurons in the spinal segments of the primary source of nociception (temporal summation of second pain or wind up)

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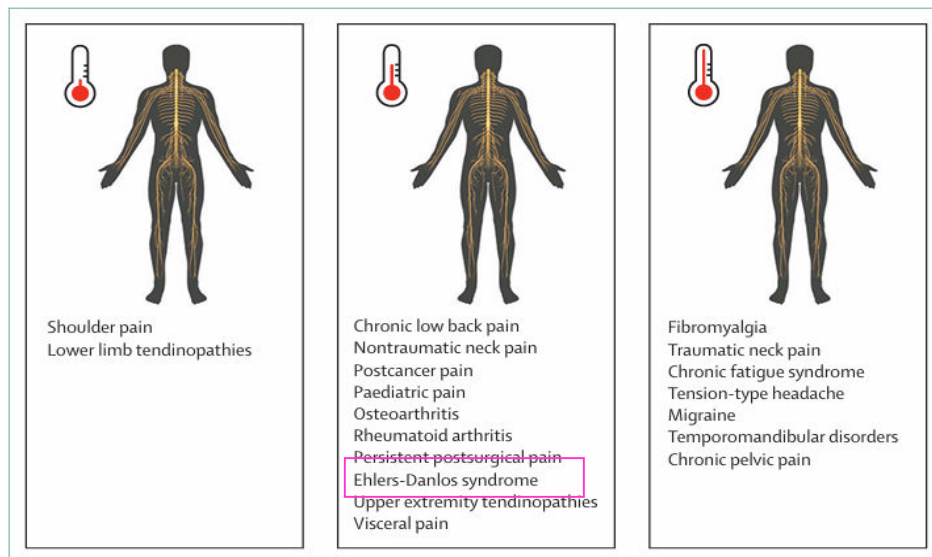


Figure 1: Medical diagnoses related to central sensitisation shown on the central sensitisation continuum
The height of the thermometer reading indicates the severity of central sensitisation.

Nijs J, et al. Lancet Rheumatol 2021;3:e383-92

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Measuring Central Sensitization:

Central Sensitization Inventory

<30 – subclinical
 30-39 – mild
 40-49 – moderate
 50-59 – severe
 ≥60 – extreme

Neblett R, et al. J Pain 2013 Mar 13;14(5):438-445

Table 2. Central Sensitization Inventory

Name: _____ SS#: _____
 Date: _____

Please circle the best response to the right of each statement.

1. I feel ~~unrefreshed~~ when I wake up from sleeping.	Never	Rarely	Sometimes	Often	Always
2. My muscles feel stiff and achy.	Never	Rarely	Sometimes	Often	Always
3. I have anxiety attacks.	Never	Rarely	Sometimes	Often	Always
4. I grind or clench my teeth.	Never	Rarely	Sometimes	Often	Always
5. I have problems with diarrhea and/or constipation.	Never	Rarely	Sometimes	Often	Always
6. I need help in performing my daily activities.	Never	Rarely	Sometimes	Often	Always
7. I am sensitive to bright lights.	Never	Rarely	Sometimes	Often	Always
8. I get tired very easily when I am physically active.	Never	Rarely	Sometimes	Often	Always
9. I feel pain all over my body.	Never	Rarely	Sometimes	Often	Always
10. I have headaches.	Never	Rarely	Sometimes	Often	Always
11. I feel discomfort in my bladder and/or burning when I urinate.	Never	Rarely	Sometimes	Often	Always
12. I do not sleep well.	Never	Rarely	Sometimes	Often	Always
13. I have difficulty concentrating.	Never	Rarely	Sometimes	Often	Always
14. I have skin problems such as dryness, itchiness, or rashes.	Never	Rarely	Sometimes	Often	Always
15. Stress makes my physical symptoms get worse.	Never	Rarely	Sometimes	Often	Always
16. I feel sad or depressed.	Never	Rarely	Sometimes	Often	Always
17. I have low energy.	Never	Rarely	Sometimes	Often	Always
18. I have muscle tension in my neck and shoulders.	Never	Rarely	Sometimes	Often	Always
19. I have pain in my jaw.	Never	Rarely	Sometimes	Often	Always
20. Certain smells, such as perfumes, make me feel dizzy and nauseated.	Never	Rarely	Sometimes	Often	Always
21. I have to urinate frequently.	Never	Rarely	Sometimes	Often	Always
22. My legs feel uncomfortable and restless when I am trying to go to sleep at night.	Never	Rarely	Sometimes	Often	Always
23. I have difficulty remembering things.	Never	Rarely	Sometimes	Often	Always
24. I suffered trauma as a child.	Never	Rarely	Sometimes	Often	Always
25. I have pain in my pelvic area.	Never	Rarely	Sometimes	Often	Always
Subtotal					

Total = _____

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GG has a CSI score of 67, indicating
 extreme central sensitization.

Now what?

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Management

- Neuroscience education
- Joint protection and stability
 - Regular low impact exercise
 - Strengthening and stabilization (PT)
 - Mobility aids, braces, splints (ring), compression garments
- Pain Management
 - Analgesics
 - Heat/cold
- Lifestyle
 - Nutrition, hydration
- Mental health/support
 - Mindfulness, meditation, deep breathing
 - Identify and manage triggers (e.g. stress)

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Joint Stabilization: Physical Rehabilitation

- Retrospective evaluation of physical rehab in hEDS patients
- Measures included 6-minute walk test, kinesiophobia, fatigue, pain and quality of life
- Significant improvement in walked distance, which was maintained at 6 weeks ($p = .001$)
- Significant improvement in kinesiophobia ($p=.033$)
- Significant improvement in fatigue ($p=.01$)
- Significant improvement in quality of life (e.g. vitality) ($p=.001$)

Hakimi A, et al. Am J Med Genet Part A 2020;182A:2263-2271

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Cognitive Behavioral Therapy

- Scoping review of psychological interventions in EDS, HSD or both (10 studies – case and cohort studies)
- Multiple different approaches including cognitive behavioral therapy, acceptance commitment therapy, intensive interdisciplinary pain treatment, etc.
- Could not draw conclusions

Song JZ, et al. Orphanet J Rare Dis 2023 Aug 31;18(1):254

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Pain Pharmacotherapy



- Bottom up – addressing localized joint pain
 - COX-2 selective NSAIDs, used PRN
 - Topical NSAIDs for superficial joints (e.g. fingers)
 - Acetaminophen or OTC products
- Top down – addressing central sensitization
 - SNRI's
 - Gabapentinoids

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Nutrition

Goals

1. Reduce autonomic dysfunction
2. Reduce immune reactions
3. Normalize gut bacteria
4. Support digestion, absorption, and metabolism

Limit added sugars and refined carbohydrates

Limit processed foods

Limit saturated and trans fats

Avoid sugar substitutes

Consider going gluten-free

Extra sodium with POTS!

<https://hypermobilityclinic.org/nutrition-and-ehlers-danlos-syndrome/>

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Neuroscience Education

- Systematic review of neuroscience education in people with chronic musculoskeletal pain and central sensitization
- Outcomes compared to usual treatment for pain, disability and psychosocial factors
- Overall, showed that pain neuroscience education improves pain, disability and psychosocial factors, particularly when done 1:1 with medium or long-term sessions with reinforcements (vs. remote sessions)

Lepri B, et al. Int J Environ Res Public Health 2023 Feb 24;20(5):4098

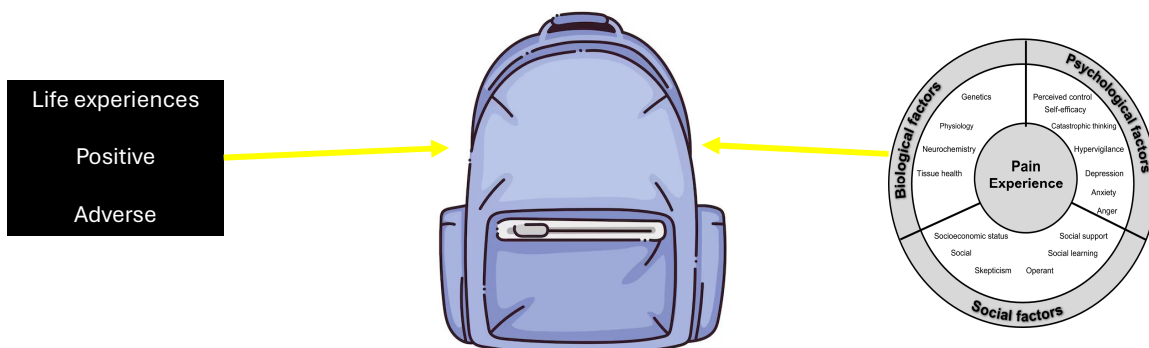
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Examples of scripting

- Repeated “microtraumas” due to joint instability send repeated pain messages to the brain for processing. With repetition, we know that the nervous system “learns” pain, like muscle memory. The more the joint is affected, the more “practice” the pain pathway gets until it doesn’t really need a stimulus to create pain.
- Over time, pain becomes an indicator of risk and need for protection, not necessarily biological tissue damage. Slight injury or no injury can feel very painful depending on the context and what else is in our “backpack”.
- With EDS, both of these processes can be going on

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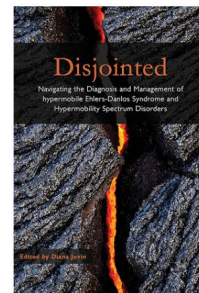
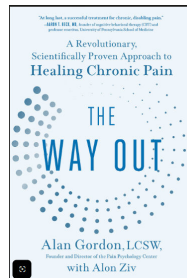
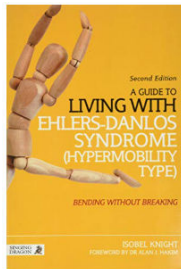
The Back Pack



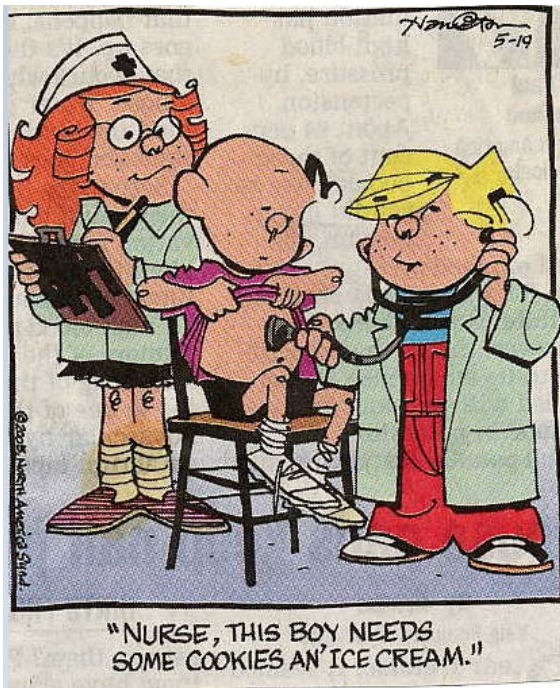
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Resources

- Lorimer Moseley on YouTube “Why Things Hurt” and “Pain, the Brain and Your Amazing Protectometer”
- Also “The Explain Pain Handbook: Protectometer” by Moseley and Butler
- www.tamethebeast.org
- “The Way Out” by Alan Gordon
- <https://override.health>
- The Ehlers-Danlos Society (www.ehlers-danlos.com)



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Questions?

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