Hypermobile Ehlers-Danlos Syndrome (hEDS)

A Patient Guide for Rheumatologists

WHAT IS hEDS? hEDS is a heritable disorder of connective tissue, the structural 'glue' of the body, causing joint instability, skin fragility, and systemic effects. Severity varies widely, from mild laxity and intermittent bracing to wheelchair use and complex multisystem involvement.

~1 in 500 people affected	Avg. 10+ years to diagnosis	3:1 to 4:1 diagnosed are female	No cure: management-focused

HOW HEDS AFFECTS THE BODY — SYSTEMIC INVOLVEMENT:

Patient has checked applicable symptoms

Neurological

- ☐ Migraines & headaches
- ☐ Brain fog/cognitive fatigue
- ☐ Small fiber neuropathy
- ☐ Proprioception deficits
- ☐ Anxiety/depression (often neurological in origin)

Gastrointestinal

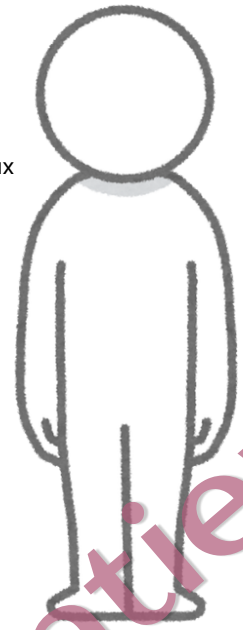
- ☐ IBS
- ☐ Gastroparesis/ delayed emptying
- ☐ GERD & acid reflux
- ☐ Food intolerances

Immune / MCAS

- ☐ MCAS — mast cell overactivation
- ☐ Flushing, hives, itching
- ☐ GI distress & food reactions
- ☐ Chemical/environmental sensitivity

Musculoskeletal

- ☐ Joint hypermobility & instability
- ☐ Subluxations & dislocations
- ☐ Chronic widespread pain
- ☐ Muscle fatigue & weakness
- ☐ Cervical instability (may contribute to headache, cranial nerve symptoms, or myelopathy)



Cardiovascular

- ☐ POTS — heart rate spikes on standing
- ☐ Blood pooling & dizziness
- ☐ Palpitations

Dermatological

- ☐ Soft, velvety, hyperextensible skin
- ☐ Stretch marks without weight change
- ☐ Easy bruising
- ☐ Poor wound healing

Genitourinary

- ☐ Pelvic floor dysfunction
- ☐ Bladder urgency/frequency
- ☐ Chronic pelvic pain
- ☐ Menstrual irregularities

Fatigue & Sleep

- ☐ Profound fatigue
- ☐ Non-restorative sleep
- ☐ Post-exertional malaise
- ☐ Chronic widespread pain at rest

DO

- Recognize hEDS as a legitimate connective tissue disorder
- Screen for POTS, MCAS, and GI comorbidities, including orthostatic vitals
- Coordinate with neurology, cardiology, and pain management
- Validate symptom burden even when labs are normal
- Consider PT specializing in hypermobility

DON'T

- Dismiss as fibromyalgia without first ruling out hEDS; fibromyalgia and hEDS frequently coexist and both diagnoses may apply
- Over-rely on Beighton score: it misses many hEDS patients
- Assume normal inflammatory markers rule out significant disease
- Attribute symptoms to anxiety or deconditioning
- Avoid interpreting pain medication requests as drug-seeking before undertreated hEDS pain has been ruled out
- Attribute orthostatic symptoms to anxiety before ruling out POTS

ORDER / REFER

- Medical genetics referral if vascular or other EDS subtype cannot be excluded; ophthalmology evaluation may be indicated as part of that workup
- Tilt table test or NASA Lean Test as in-office POTS screen
- Allergy/Immunology (MCAS)
- GI motility evaluation if indicated
- PT specializing in hypermobility
- Pain management for central sensitization

Why Rheumatology Matters for hEDS: hEDS has no abnormal inflammatory markers, no diagnostic imaging finding, and no blood test. Diagnosis is clinical, based on the 2017 diagnostic criteria requiring generalized joint hypermobility, systemic features, and the exclusion of other heritable connective tissue disorders. Many patients arrive at rheumatology after years of being told their labs are normal and therefore nothing is wrong. Rheumatologists are uniquely positioned to make or exclude the diagnosis, and to distinguish hEDS from overlapping conditions including fibromyalgia, lupus, seronegative inflammatory arthritis, and other hypermobility spectrum disorders.

COMMON MISDIAGNOSES IN hEDS PATIENTS PRESENTING TO RHEUMATOLOGY

Often Diagnosed As	Consider Instead/Also	Key Differentiator
Fibromyalgia	hEDS with central sensitization	Joint hypermobility history; systemic involvement; fibromyalgia and hEDS frequently coexist
Seronegative inflammatory arthritis	hEDS joint instability	Normal inflammatory markers; hypermobility on exam; subluxations rather than synovitis
Lupus (SLE)	hEDS with MCAS or autonomic involvement	Normal ANA or low titer; symptoms driven by mast cell or autonomic dysfunction, not autoimmune inflammation
Hypermobility Spectrum Disorder (HSD)	hEDS meeting full 2017 criteria	Apply full diagnostic criteria; HSD is appropriate only when hEDS criteria are not met. It is not a default label
Psychosomatic/Functional	hEDS with normal labs	Clinical diagnosis; normal labs are expected and do not exclude hEDS
Chronic fatigue syndrome or ME/CFS	hEDS with POTS and post-exertional malaise	Orthostatic assessment; POTS frequently drives fatigue and PEM in hEDS patients
Allergic or autoimmune condition	MCAS in the context of hEDS	Normal standard allergy panels; episodic multisystem symptoms; responds to H1/H2 antihistamines and mast cell stabilizers

Cervical Instability: Connective tissue laxity in hEDS can affect the craniocervical junction and upper cervical spine. Instability at these sites may produce neurological symptoms including headache, dizziness, visual disturbances, and upper limb weakness. Avoid aggressive cervical manipulation. If neurological symptoms are present, consider referral to a neurosurgeon or neurologist with experience in connective tissue disorders before initiating manual therapy.

The Beighton Score: A Screening Tool, Not a Diagnostic Gate: The Beighton score measures hypermobility at nine specific joint sites and was developed as a research screening tool, not a diagnostic instrument for hEDS. A score below the threshold does not exclude hEDS for several reasons:

- Joint laxity decreases with age. Many patients who met criteria in their twenties score below threshold by their forties.
- Protective muscle guarding developed over years of managing instability can mask hypermobility on exam.
- Prior injury, surgical repair, or bracing at scored joints can artificially lower the score.
- hEDS affects connective tissue systemically. A patient with significant spinal, hip, or shoulder instability may score low on the nine standard sites.

The 2017 diagnostic criteria require generalized joint hypermobility confirmed by Beighton score OR historical evidence of hypermobility at five or more joints, meaning patient-reported history of hypermobility counts when current exam findings are limited. A thorough hypermobility history, systemic feature assessment, and exclusion of other heritable connective tissue disorders complete the diagnostic picture. The score is one input, not the decision.

The hEDS Trifecta: Frequently Co-Occurring Conditions

hEDS	+	POTS	+	MCAS
Joint instability Structurally abnormal connective tissue Systemic symptoms		Heart rate spikes on standing Dizziness & fatigue Brain fog & cognitive dysfunction Exercise intolerance		Mast cell overactivation Flushing, hives, itching GI distress & food reactions Chemical/environmental sensitivity

MY CURRENT MEDICATIONS & SUPPLEMENTS

WHAT HELPS:

WHAT MAKES IT WORSE:

PRIOR DIAGNOSES RECEIVED:

SPECIALISTS PREVIOUSLY SEEN

WHAT I NEED FROM TODAY'S APPOINTMENT

Referrals needed:	
Questions I have:	
Medication changes:	
My primary concern today:	
Other:	

CURRENT SYMPTOM SEVERITY: Rate 0–10 using the Wonkosk Pain Scale (pg. 4)

Joint Pain and Instability Severity:	
Fatigue Severity:	
GI Symptoms Type and Frequency:	
Heart Rate and Dizziness Triggers and Frequency:	
Additional Symptoms:	

Sources: Moffatt et al. 2017 (JJMD) | Tinkle et al. 2017 (JJMD) | Genetics in Medicine Open 2023 | ehlers-danlos.com | PMC10899487 | PMC10468568
This document was prepared to support a productive appointment with a provider who may not specialize in hEDS.
Rheumatology is often the critical diagnostic crossroads for this condition. This reference is intended to make that conversation more efficient.

MANKOSKI PAIN SCALE

Use this scale when rating your pain severity in CURRENT SYMPTOM SEVERITY

#	What the pain is like	Typical treatment	In my own words
0	No pain.	*No medication needed.*	*"I feel completely normal."*
1	Very minor annoyance -- occasional minor twinges.	*No medication needed.*	*"Hardly notice it."*
2	Minor annoyance -- occasional strong twinges.	*No medication needed.*	*"Annoying but manageable."*
3	Annoying enough to be distracting.	*Mild OTC painkillers may help.*	*"Hard to ignore; affects my focus."*
4	Can be ignored if very focused, but still distracting.	*Mild OTC painkillers relieve pain for 3-4 hours.*	*"Getting in the way of tasks."*
5	Can't be ignored for more than 30 minutes.	*Mild OTC painkillers reduce pain for 3-4 hours.*	*"Stops me mid-task."*
6	Can't be ignored. Can still go to work and participate in social activities.	*Stronger prescription pain relief needed; works 3-4 hours.*	*"Present all the time. I push through."*
7	Difficult to concentrate; interferes with sleep. Can still function with effort.	*Stronger painkillers only partially effective.*	*"Hard to function. Sleep is disrupted."*
8	Physical activity severely limited. Can read/converse with effort. Nausea possible.	*Strongest painkillers minimally effective.*	*"Mostly bed-bound. May feel nauseated."*
9	Unable to speak. Crying out or moaning uncontrollably. Near delirium.	*Strongest painkillers only partially effective.*	*"Cannot communicate. Losing control."*
10	Unconscious. Pain causes passing out.	*Strongest painkillers only partially effective.*	*"Passed out or on the verge of it."*

Mankoski Pain Scale developed by Andrea Mankoski, 1995. Adapted for patient communication. Not a clinical diagnostic tool.

IMPORTANT NOTE FOR HEDS PATIENTS & PROVIDERS:

People with hEDS often have an altered pain baseline due to central sensitization -- a process in which the nervous system becomes increasingly sensitized to pain signals over time.

A "5" for this patient may be what others feel as a "8."
Please do not compare severity numbers to those of patients without chronic illness.

This scale helps us communicate:
it is not a measure of tolerance, willpower, or how "bad" things really are.