

# Hypermobile Ehlers-Danlos Syndrome (hEDS)

## A Patient Guide for Healthcare Providers

**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. hEDS severity varies widely, from mild joint laxity to wheelchair use, feeding tubes, and complex multisystem involvement.

~1 in 500 people affected

Avg. 10+ years to diagnosis

3:1 to 4:1 diagnosed are female

No cure: treatment is 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

### 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 (can cause neurological issues)

### Cardiovascular

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

### Dermatological

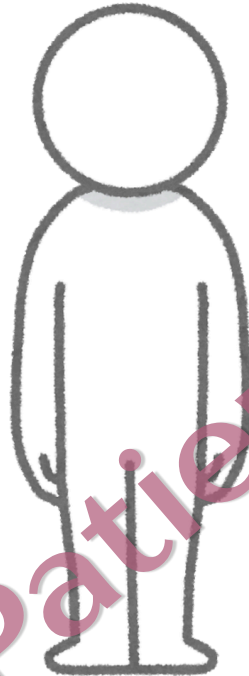
- Soft, velvety, hyperextensible skin
- Stretch marks without weight change
- Easy bruising
- Poor wound healing

### 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



## The hEDS Trifecta: Frequently Co-Occurring Conditions

hEDS  
Joint instability  
Structurally abnormal connective tissue  
Systemic symptoms

+

POTS  
Heart rate spikes on standing  
Dizziness & fatigue  
Brain fog & cognitive dysfunction  
Exercise intolerance

+

MCAS  
Mast cell overactivation  
Flushing, hives, itching  
GI distress & food reactions  
Chemical/environmental sensitivity

**Why Primary Care Matters for hEDS:** The primary care provider is the most consequential person in an hEDS patient's medical journey – not because they are expected to be an hEDS specialist, but because they control access to the specialists who are. A referral made early changes outcomes. A referral withheld, or a case closed with a functional label, can add years to a diagnostic odyssey that already averages over a decade.

hEDS patients frequently arrive at primary care after being dismissed elsewhere. They have researched their condition out of necessity and learned to anticipate skepticism. A PCP who validates the condition, coordinates across specialties, and documents symptoms consistently is providing the structural support that makes the rest of the patient's care possible. This condition has no cure. Management is the goal, and management requires a coordinated team.

## MSK

- Treat as a multi-system condition
- Coordinate across specialties
- Utilize the patient's experience
- Screen for PTSD, MCAD, and GI issues
- Use the most recent techniques
- Promote MSK therapy in the short to enable already accommodations, and continuity across providers
- Recognize that functional capacity varies day to day - a patient who appeared well at their last visit may be significantly limited today
- Use the patient's self knowledge actively - Most MSK patients have researched their condition out of necessity and are a wealth source of clinical context

## MSK

- Screen as "gut issues"
- Recommend high impact exercise
- Assess normal labs - no problem
- Check common comorbidities
- Consider a higher care level
- Address pain aspects to behavior rather than undertreated chronic illness
- Address cognitive symptoms to depression or anxiety before using all diagnostics
- Use a patient's ability to work or appear functional as evidence that symptoms are manageable
- Close the case after a single specialist where a normal finding - MSK requires coordinated multi-system evaluation, not sequential diagnosis

## GISSR / MSK

- GI table test or MSK test not as in-office screen
- Rheumatology referral
- GI study evaluation
- GI specialist is hypermobility
- Allergy / Immunology (MCAD)
- Most all medical testing if MCAD is suspected - serum tryptase, 24 hour urine  $\alpha$ -methylglutamate, and urine prostaglandin
- GI coordinate with Allergy/Immunology
- Pain management consultation
- Genetic referral if MCAD subtype is unclear or family history suggests a more complex presentation - confirmed MSK does not mean genetic testing
- Psychology referral using a trauma informed framework - to support patients managing chronic illness, not to explain every physical symptom

## COMMON MISDIAGNOSES IN MSK PATIENTS PRESENTING TO PRIMARY CARE

Often Diagnosed As	Consider Instead/Also	Key Differentiator
Anxiety / panic disorder	MSK with PTSD and autonomic dysfunction	Postural heart rate change; symptoms worsen on standing not purely situational
Fibromyalgia	MSK with central sensitization	Not hypermobility history; systemic involvement both may occur
Depression (primary)	MSK with undertreated chronic pain	Most symptoms secondary to unmanaged physical illness, treat the physical cause
Hypochondria / somatic symptom disorder	MSK with normal labs	Clinical diagnosis expected to have normal labs, symptom pattern is physiologically consistent
Chronic fatigue syndrome / ME-CFS	MSK with PTSD and post-exertional malaise	Difficult to assess, PTSD frequently drives fatigue in MSK
Deconditioning	MSK with exercise intolerance	Symptoms predominate inactivity; exercise intolerance is physiological, not motivational
Strong pain or fatigue (stress)	Early MSK presentation	Pain severity, systemic symptoms, and family history warrant monitoring rather than dismissal

PLEASE COMPLETE THIS PAGE BEFORE YOUR APPOINTMENT AND BRING IT WITH YOU

**MY CURRENT MEDICATIONS & SUPPLEMENTS**

**WHAT HELPS:**

**WHAT MAKES IT WORSE:**

**WHAT I NEED FROM TODAY'S APPOINTMENT**

Referrals needed:

Questions I have:

Medication changes:

My primary concern today:

Other:

**CURRENT SYMPTOM SEVERITY: 1-10 Refer to Workbook Pain Scale (pg 4)**

Joint pain/tenderness severity:

Fatigue severity:

All symptoms type and frequency:

Heart rate/blood pressure triggers and frequency:

Additional symptoms:

**ADDITIONAL NOTES**

## MARKOSKI PAIN SCALE

Use this scale when rating your pain severity as 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.	Most OTC painkillers may help.	"Hard to ignore; affects my focus."
4	Can be ignored if very focused, but still distracting.	Most OTC painkillers relieve pain for 2-4 hours.	"Getting in the way of tasks."
5	Can't be ignored for more than 30 minutes.	Most OTC painkillers reduce pain for 2-4 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 2-4 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 reach/move 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."

Markoski Pain Scale developed by Andrea Markoski, PhD. 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 '9'.  
Please do not compare severity numbers to those of patients without chronic illness.

The scale helps us communicate:  
It is not a measure of tolerance, willpower, or how 'bad' things really are.