Hypermobile Ehlers-Danlos Syndrome (hEDS)

A Patient Guide for Orthopedic Surgeons & Orthopedists

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

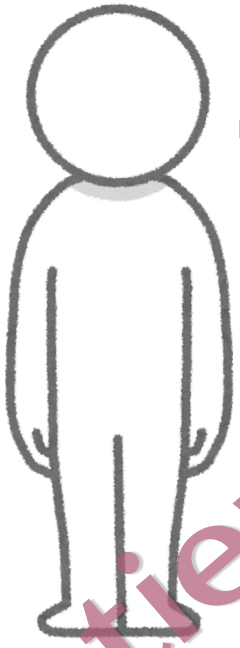
- ☐ IBS
- ☐ Gastroparesis/ delayed emptying
- ☐ GERD & acid reflux
- ☐ Food intolerances

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 (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

Fatigue & Sleep

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

Genitourinary

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

DO

- Recognize joint instability as the primary driver of pain and injury in hEDS: not deconditioning, anxiety, or drug-seeking
- Prioritize stabilization and proprioception-based rehabilitation before considering surgical intervention
- Coordinate with physical therapy experienced in hypermobility: standard PT protocols can cause harm in this population
- Account for tissue fragility when planning any procedure; sutures, anchors, and repairs are at elevated risk of failure
- Screen for POTS and MCAS before elective surgery; both significantly affect anesthetic and perioperative risk
- Recognize that pain severity is often underreported relative to objective findings due to central sensitization

DON'T

- Attribute widespread joint pain and instability to anxiety, psychosomatic causes, or malingering
- Recommend aggressive strengthening programs without hypermobility-aware PT oversight: overloading unstable joints worsens instability
- Perform surgery as a first-line intervention without a documented conservative management trial
- Assume standard tissue strength: suture pullout, anchor failure, and repair breakdown are significantly more common in hEDS
- Use epinephrine-containing local anesthetics without first screening for MCAS and autonomic instability
- Dismiss recurrent subluxations or dislocations as patient behavior or non-compliance

CONSIDER / REFER

- Physical therapy with a provider experienced in hypermobility and joint stabilization before and after any intervention
- Bracing and orthotic evaluation for unstable joints: custom options are often more effective than off-the-shelf
- Preoperative cardiology or autonomic evaluation if POTS is suspected or confirmed
- Allergy/Immunology consultation preoperatively if MCAS is suspected: medication and anesthetic sensitivities must be mapped
- Rheumatology and pain management for multidisciplinary care coordination and central sensitization management
- Caution with corticosteroid injections: repeated use may further weaken compromised connective tissue; discuss alternatives with pain management

MUSCULOSKELETAL MANIFESTATIONS IN hEDS — WHY THEY OCCUR

Structural Mechanisms

- **Defective collagen:** Abnormal collagen composition reduces tensile strength and elasticity throughout ligaments, tendons, joint capsules, and cartilage
- **Joint capsule laxity:** Insufficient passive restraint allows excessive range of motion, microtrauma, and recurrent subluxation or dislocation
- **Proprioceptive deficit:** Impaired joint position sense reduces the neuromuscular feedback needed to prevent injury and stabilize joints dynamically
- **Chronic compensation and poor repair:** Muscles chronically overwork to substitute for ligamentous instability, and abnormal collagen repair mechanisms slow recovery and increase re-injury risk

Common Orthopaedic Diagnoses / Presentations

- Recurrent joint subluxations and dislocations (shoulder, patella, hip, wrist, ankle)
- Chronic joint pain without clear structural pathology on imaging
- Ligamentous laxity and multi-joint instability
- Early-onset osteoarthritis from chronic microtrauma
- Tendinopathy and enthesopathy from chronic compensatory loading
- Cervical instability (may produce radicular or myelopathic symptoms)
- Recurrent soft tissue injuries (sprains, strains) with prolonged recovery
- Failed prior orthopaedic repairs or unusually slow post-surgical healing

SURGICAL CONSIDERATIONS IN hEDS — WHAT TO KNOW BEFORE YOU OPERATE

Tissue and Repair Integrity
Connective tissue quality is fundamentally altered in hEDS. Standard surgical assumptions about tissue strength, suture holding capacity, and anchor fixation do not apply. Ligament and tendon repairs are at significantly elevated risk of failure, stretching, or re-rupture. Surgeons should consider augmentation techniques, slower rehabilitation progression, and extended protected weight-bearing. Wound healing is frequently delayed; dehiscence and widened scarring are common even with meticulous closure.

Anesthetic and Perioperative Risk hEDS patients carry several perioperative risk factors requiring proactive planning. Cervical instability poses intubation risk; careful airway management and positioning are essential throughout. POTS increases hemodynamic instability risk under anesthesia; preoperative hydration, compression, and anesthesia team awareness are critical. MCAS patients may react to anesthetic agents, latex, prep solutions, dyes, and adhesives; obtain a baseline serum tryptase (during a non-reactive period) and full sensitivity history before scheduling; coordinate with Allergy/Immunology. Epinephrine-containing local anesthetics and nitrous oxide may both be poorly tolerated due to adrenergic sensitivity and mast cell activation risk.

Pain Management Considerations Pain in hEDS is complex and often involves central sensitization, meaning standard pain scales will underrepresent actual severity and standard analgesic approaches may be insufficient. NSAIDs and opioids may both be poorly tolerated; NSAIDs due to MCAS overlap, opioids due to mast cell degranulation risk. Multimodal pain planning in coordination with pain management is strongly recommended before elective surgery.

COMMON MISDIAGNOSES IN hEDS PATIENTS PRESENTING TO ORTHOPEDICS

Often Diagnosed As	Consider Instead/Also	Key Differentiator
Anxiety/psychosomatic pain	hEDS with central sensitization	Beighton score (standardized hypermobility assessment); multi-joint history; skin findings
Recurrent sprain (isolated)	Ligamentous laxity (hEDS)	Pattern of bilateral, multi-joint involvement
Rotator cuff tear (traumatic)	hEDS instability with secondary impingement	Atypical trauma mechanism; bilateral shoulder symptoms
Patellofemoral syndrome	hEDS patellar instability	Recurrent subluxation history; ligamentous laxity
Failed surgical repair (unexplained)	Undiagnosed hEDS	Tissue quality findings intraoperatively; prior repair history
Malingering/drug seeking	Undertreated hEDS pain	Central sensitization; inadequate prior pain management
Idiopathic scoliosis	hEDS-associated spinal instability	Early onset; progressive curve pattern atypical for adolescent idiopathic scoliosis; concurrent multi-joint hypermobility; full connective tissue evaluation warranted

Beyond Instability: Why Standard Orthopedic Rehab Can Harm hEDS Patients

Rehabilitation protocols designed for structurally normal connective tissue are frequently inappropriate for hEDS. High-repetition loading, aggressive range-of-motion work, and standard post-surgical timelines can worsen joint instability, accelerate microtrauma, and increase pain. The goal in hEDS rehabilitation is stabilization and motor control, not mobility or flexibility. Physical therapists unfamiliar with hypermobility may inadvertently push patients toward end-range motion that further stretches already compromised ligamentous structures. Referral to a PT with documented hypermobility experience is strongly recommended before and after any orthopedic intervention. The Muldowney Protocol, a structured rehabilitation approach developed specifically for hypermobility, is one established starting point.

When Surgery Is Necessary: What to Expect in hEDS

Patients with hEDS who undergo orthopedic surgery face higher rates of repair failure, longer recovery timelines, increased post-surgical pain, and greater likelihood of requiring revision procedures. This is not a reflection of patient behavior or effort; it is a direct consequence of the underlying connective tissue disorder. Shared decision-making should include explicit discussion of these elevated risks, conservative alternatives, and hypermobility-aware post-surgical rehabilitation. When surgery is necessary, tissue augmentation, slower rehabilitation progression, and extended monitoring are appropriate. Outcomes improve when the full clinical picture is addressed: autonomic dysfunction, MCAS overlap, central sensitization, nutritional status, and GI dysmotility, all of which may increase perioperative risk.

MY CURRENT MEDICATIONS & SUPPLEMENTS

WHAT HELPS:

WHAT MAKES IT WORSE:

WHAT I NEED FROM TODAY'S APPOINTMENT

My primary concern today:

Questions I have:

Medication changes to discuss:

Referrals needed:

Other:

CURRENT SYMPTOM SEVERITY: Complete this section using the [illegible] Pain Scale (pg. 4)

Joint pain severity (please specify joints):

Subluxation and/or dislocation frequency:

Functional limitations (mobility, grip, weight-bearing):

Pain at rest:

Pain with activity:

Additional symptoms:

Sources: [illegible]
[illegible]

This document was created to provide an orthopedic-focused clinical reference for providers less familiar with hEDS.
Joint instability in this condition has identifiable structural and physiological drivers that this reference is designed to make quickly accessible.

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.