Hypermobile Ehlers-Danlos Syndrome (hEDS)

A Patient Guide for Dentists & Oral Surgeons

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. Dental and surgical procedures carry specific risks in hEDS that require modified technique and heightened awareness.

~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

Check any symptoms that apply to you before your appointment.

Cardiovascular

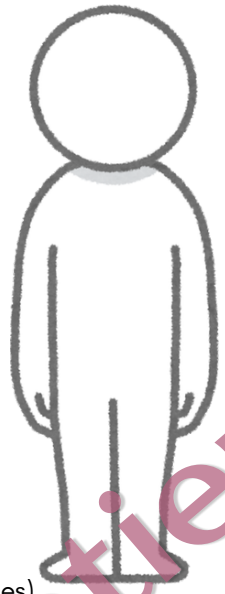
- ☐ POTS — heart rate spikes on standing
- ☐ Blood pooling & dizziness
- ☐ Palpitations

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)



Dermatological

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

Neurological

- ☐ Migraines & headaches
- ☐ Brain fog/cognitive fatigue
- ☐ Small fiber neuropathy
- ☐ Proprioception deficits
- ☐ Anxiety/depression

Fatigue & Sleep

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

DO

- Limit mouth opening duration and use bite blocks cautiously to protect the TMJ
- Use gentle tissue handling: oral tissues are fragile and tear more easily
- Allow extra healing time and monitor wound sites closely post-procedure
- Ask about MCAS before using epinephrine-containing anesthetics: reactions are possible
- Document anesthesia response and share with patient for future providers
- Consider shorter appointments with breaks to reduce jaw fatigue and subluxation risk
- Ask about cervical instability and use positioning supports or breaks as needed during extended procedures

DON'T

- Assume standard local anesthetic doses will be sufficient; they frequently are not in hEDS
- Dismiss a patient who reports feeling pain during a procedure: anesthesia resistance is real and documented
- Force or hold the jaw open for extended periods
- Assume normal healing timelines: wound dehiscence and delayed healing are documented risks in this population
- Use epinephrine-containing anesthetics without first screening for MCAS
- Attribute post-procedural pain complaints to anxiety or drug-seeking behavior
- Perform elective oral surgery without discussing modified anesthesia and wound care protocols

CONSIDER / REFER

- Higher or more frequent local anesthetic dosing when resistance is encountered
- Epinephrine-free anesthetic formulations if MCAS is present or suspected
- Pre-procedural bite guard or jaw support for patients with known TMJ instability
- Extended post-operative follow-up for wound monitoring
- Oral and maxillofacial specialist for complex extractions or jaw surgery
- TMJ specialist if jaw dysfunction, clicking, or subluxation is present; cervical spine specialist if instability is reported
- Pain management coordination for post-surgical pain in patients with central sensitization

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

Anesthesia Resistance in hEDS: A Real and Documented Risk Local anesthetic resistance is one of the most consistently reported experiences among hEDS patients in dental settings, and one of the most frequently dismissed. The proposed mechanism involves abnormal connective tissue affecting anesthetic diffusion, as well as sodium channel variants associated with hEDS. A patient who reports feeling pain during a procedure is not being dramatic. When standard dosing fails, increase the dose, change the injection site, allow more time, or consider an alternative anesthetic. Proceeding without adequate anesthesia causes acute harm and compounding medical trauma. Document the patient's response and share it with them. This information is critical for every future dental and surgical encounter.

Surgical Precautions for Oral Surgeons: hEDS affects the structural integrity of all connective tissue, including oral and mucosal tissue. Wounds may dehisce, bleed more than expected, or heal more slowly than expected. Sutures may not hold as expected in fragile tissue. Use technique and materials accordingly. Post-operative pain may be significantly higher than expected due to central sensitization, and standard post-op analgesic protocols may be insufficient. Coordinate with the patient's pain management provider before elective procedures. If the patient has MCAS, pre-medicating with H1 and H2 antihistamines before surgery is worth discussing with their immunologist or allergist.

COMMON MISDIAGNOSES IN hEDS PATIENTS PRESENTING TO DENTISTRY

Often Diagnosed As	Consider Instead/Also	Key Differentiator
Anxiety or psychosomatic pain	Anesthesia resistance in hEDS	Documented mechanism; sodium channel variants and abnormal tissue diffusion affect local anesthetic efficacy
Drug-seeking behavior	Undertreated post-procedural pain with central sensitization	Altered pain baseline is expected in hEDS; standard analgesic protocols are frequently insufficient
TMJ disorder (idiopathic)	hEDS with joint hypermobility	TMJ instability, clicking, and subluxation are direct connective tissue manifestations; assess for systemic hypermobility
Poor oral hygiene or patient non-compliance	hEDS tissue fragility and delayed healing	Wound dehiscence and slow healing are documented risks independent of patient behavior
Allergic reaction (unspecified)	MCAS triggered by epinephrine-containing anesthetic	Episodic flushing, hives, or systemic reactions; screen for MCAS before using epinephrine

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 Workbook Pain Scale (pg. 5)
Tooth pain severity:
Jaw pain/TMJ frequency and severity:
Additional Symptoms:

Sources: [illegible]

This document was created to alert dental and surgical providers to specific hEDS-related risks that require modified technique. Anesthesia resistance, tissue fragility, and delayed healing are documented and clinically significant in this population.

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 Andrew 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 "9."
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.