Hypermobility 114: Hospitalization with HSD, POTS, MCAD

Leslie N Russek, PT, DPT, PhD, OCS
Clarkson University

Who Am I?

- Professor Emeritus, Physical Therapy, Clarkson University.
- Staff PT, St. Lawrence Health System, Potsdam NY.
  - Clinical specialties: hypermobility, chronic pain, fibromyalgia, headaches, temporomandibular disorders
- Facilitator of the North America Allied Health Professionals ECHO
- Member of:
  - The Allied Health Working Group of the International Consortium of Ehlers-Danlos Syndromes and Hypermobility Spectrum Disorders
  - The National Academy of Sciences, Engineering and Medicine Committee on Selected Heritable Connective Tissue Disorders and Disability.
- Author of ”Chronic Pain” chapter in *Physical Rehabilitation* textbook for PT students
- Lrussek@Clarkson.edu
- https://webspace.clarkson.edu/~lrussek/
- I do free weekly Zoom lectures for people with HSD (see website)

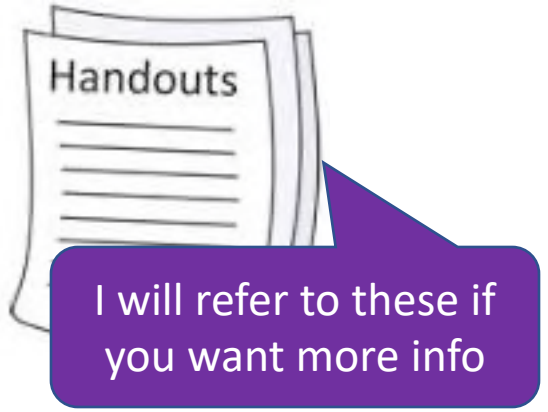
I do not have any conflicts of interest to report

Hypermobility Lecture Series Schedule

I will refer to these if you want more info

- HSD 101: Basics of HSD/hEDS and self-care
- HSD 102: POTS and POTS self-care, basics of MCAS
- HSD 103: Pain management in HSD/hEDS
- HSD 104: Safe exercise selection and progression with HSD/hEDS
- HSD 105: Posture and joint protection
- HSD 106: Gut issues in HSD/hEDS, POTS, MCAS
- HSD 107: Fatigue in HSD/hEDS and POTS
- HSD 108: Headaches, migraines, & TMJ pain associated with HSD, POTS and MCAS
- HSD 109: Breathing disorders in HSD
- HSD 110: Lumbar instability
- HSD 111: Conservative management of cervical instability
- HSD 112: The vagus nerve
- HSD 113: The role of fascia
- NEW - HSD 114: Hospitalization with HSD, POTS, MCAD

Relevant Handouts Available



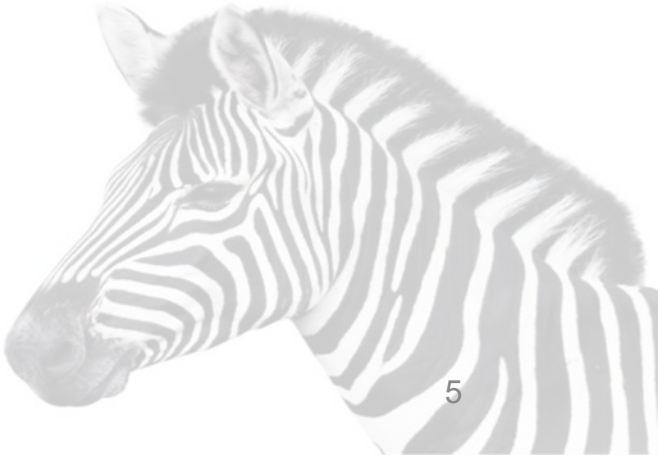
- https://webspace.clarkson.edu/~lrussek/research.html

- **Surgical precautions for people with HSD/hEDS. Most of the info in today's session.**
- **Hypermobility**
  - Sleep Hygiene and Positioning. Sleep posture and sleep hygiene strategies.
  - Upper cervical Instability. (UCI). My handout based on the recent article I co-authored.
    - You can access the full article at Full text of UCI article.
    - A comprehensive patient guide to EDS cervical instability by EDSawareness. (not created by me)
- **POTS**
  - Overview of POTS symptoms and causes.
  - Checklist for POTS self-care management.
- **MCAS**
  - Suggestions for managing MCAS.
  - How to check your medications for (MCAS) sensitivities.
  - MCAS in the Emergency Room. (not created by me)

Disclaimers

The information in this presentation is for general purposes, only, and may or may not apply to your situation.
This presentation is based on published medical literature, but cannot be used to diagnose or treat individual patients.
This information is best used to start a discussion with your health care providers to determine if/how any of these concerns may apply to you.

I cannot diagnose or make specific treatment recommendations in this lecture.



Objectives

By the end of this session, participants should be able to:

1. Identify concerns that can arise in the hospital for patients with HSD, POTS, or MCAD.
2. Begin a discussion with healthcare providers about concerns due to HSD, POTS or MCAD.

3. HSD: Hypermobility Spectrum Disorders and hypermobile Ehlers-Danlos Syndrome
4. POTS: Postural Orthostatic Tachycardia Syndrome, in this presentation including other forms of dysautonomia, such as orthostatic intolerance
5. MCAD: Mast Cell Activation Disorders

Outline

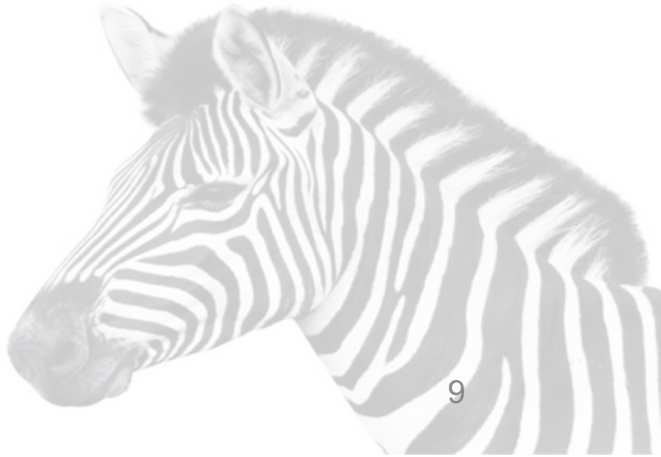
- Why is it important to know about HSD, POTS, MCAD in the hospital?
- When should you suspect HSD, POTS, MCAD?
- Specific conditions and systems
  - Migraines and headaches
  - Neurological issues
  - Cardiovascular issues
  - Immune system issues, MCAD, anaphylaxis
  - Musculoskeletal issues
  - Skin issues
  - Bleeding issues
  - Respiratory Issues
  - Obstetric issues
- Surgical precautions
- Resources for you and your providers

Recognizing Signs and Symptoms

- It can be helpful to recognize what signs & symptoms might be due to HSD, POTS, or MCAS
- You may be able to avoid unnecessary testing
- It may suggest appropriate treatment approaches
  - For example:
    - A migraine due to POTS may resolve with an IV saline drip
    - A migraine due to MCAS may resolve with appropriate antihistamine medication
    - A migraine due to cervical instability may require cervical stabilization with a brace to decrease stress to the brainstem
- It can reduce the chance that your symptoms are considered psychogenic (due to psychological causes and not physical)

Hypermobility: HSD

- Musculoskeletal pain or injury in response to minor trauma
- Easy/nontraumatic dislocation/subluxation
- Hyperextensible/fragile skin, easy bruising, slow wound healing
- GERD, vomiting, gastroparesis, IBS, prolapse, hernia, median arcuate ligament syndrome, superior mesenteric artery syndrome
- Frequent falls, clumsiness
- Upper cervical instability: syncope, non-epileptic/pseudo seizures, acute cognitive and visual changes[8]
- Increased prevalence of Chiari I Malformation, CSF leaks, idiopathic intracranial hypertension, tethered cord, Eagle syndrome[13]
- Mitral valve prolapse, varicose veins, easy bruising
- Dysfunctional breathing
- Urogenital prolapse, incontinence, hernias
- Hernias and prolapse at multiple locations



Dysautonomia, POTS, Orthostatic Intolerance

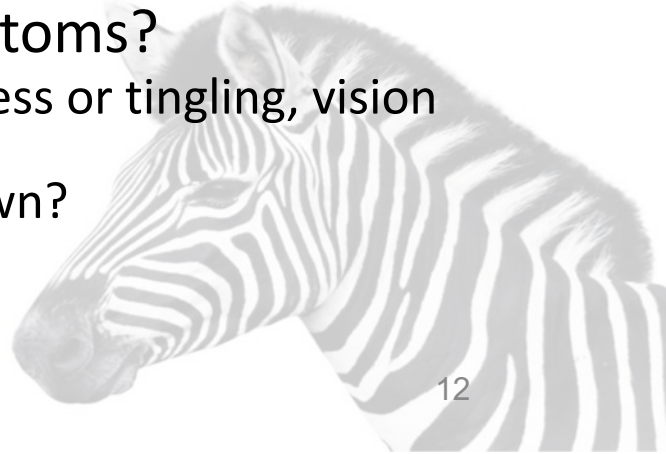
- Cardiac irregularity, tachycardia (fast heartrate, HR), bradycardia (slow HR). Tachycardia may present as anxiety or panic attack.
- Orthostatic intolerance, low blood pressure (BP)
- Dizziness, presyncope (almost passing out), syncope (passing out)
- Migraine
- Severe fatigue, exercise intolerance
- Edema or swelling in the feet and hands
- Raynaud's: cold, chalky white hands
- Acrocyanosis: purple blotchy feet/hands
- Dumping syndrome/rapid gastric emptying (diarrhea), nausea, vomiting

Mast Cell Activation Disorder/Syndrome

- Anaphylaxis: severe allergic reactions
- Migraine
- Intolerance to medication, sometimes the excipients rather than the active ingredient.
- Intolerance to odors, including perfumes, colognes, chemical cleaners.
- Food intolerances.
- IBS (irritable bowel), SIBO (small intestine bacterial overgrowth) SIBO, leaky gut
- Adhesive sensitivity for tape or sensors.

Migraines and Headaches

- Migraines are very common in POTS and MCAD flares.(Wig, 2019, Conti, 2019)
  - Are you experiencing an MCAD flare? (GI, skin, breathing, allergy symptoms?)
  - Have you been exposed to anything that could trigger MCAD?
    - E.g.: chemicals, new medications, foods, smoke, perfumes etc.
  - Are you experiencing a POTS flare? (lightheaded, tachycardia, anxious?)
  - Any reason why your POTS might flare?
    - E.g., heat, dehydration, MCAD flare, cervical instability, etc.
- Cervical instability can trigger migraines and headaches (Henderson, 2017)
  - Have you been in positions or activities that would stress the neck?
    - E.g., washing hair, dental work, car-rides, prolonged leaning forward, etc.
- Do you have any other new or worse neurological signs or symptoms?
  - Seizures, dystonia (uncontrolled muscle spasticity), passing out, numbness or tingling, vision changes, voice changes
  - Do you have sudden headaches with position changes, such as lying down?



Central Neurological System Problems

- Signs & Symptoms:
  - Vision/hearing changes
  - Sensory changes
  - Weakness/loss of coordination
  - Tics, tremors or jerks
  - Balance disturbance
  - Speech problems (dysphagia)
  - Nausea/vomiting
  - Non-epileptic seizures
  - Syncope (fainting)
  - Bowel/bladder dysfunction
  - Depersonalization (feeling unreal, disconnected from your body)

- Possible neuro conditions:
  - Upper cervical instability
  - Chiari malformation
  - Tethered cord
  - Cerebrospinal fluid leak
  - Tarlov cysts
  - Idiopathic intracranial hypertension
  - Arachnoiditis

(Henderson, 2017)

Neurological Signs & Symptoms

- Pseudo seizures/non-epileptic seizures, fainting and dystonia (muscle spasticity) may be due to cervical instability. (Russek, 2022)
  - Cervical instability may be aggravated by an MCAD flare
  - MRI done with you lying down and the neck in neutral cannot diagnose cervical instability
- Fainting caused by POTS-related syncopal events can mimic seizures, especially when they include myoclonic jerking (spastic twitching of muscles). (Rugg-Gunn, 2009)
- People with HSD are more vulnerable to a variety of neurological conditions:
  - Craniocervical and atlantoaxial instability (CCI and AAI), Chiari I malformation, CSF (cerebrospinal fluid) leaks, idiopathic intracranial hypertension, Eagle syndrome, tethered cord and Tarlov cysts. (Henderson, 2017)
- You may be anxious because of these symptoms, but this doesn't mean your symptoms are caused by anxiety
- Functional Neurological Disorders (FND) is a condition where the nervous system malfunctions

Cardiovascular Signs & Symptoms

- Dysautonomia can lead to orthostatic intolerance. HR may be abnormally high or blood pressure may drop (orthostatic hypotension). Dysautonomia can be aggravated by: bedrest, surgery, anesthesia, COVID. It most commonly presents in adolescent and adult women.(Vernino, 2021, Raj, 2020)
- Chest pain: Costochondritis is common because the joints between ribs and sternum can be lax. Costochondritis symptoms mimic a heart attack. (https://www.ncbi.nlm.nih.gov/books/NBK532931/ Subluxed ribs may also cause crushing chest pain that may feel like a heart attack. Pectoralis and scalene trigger points caused by excessive chest breathing pattern can also sometimes feel like a heart attack.(Bagcier, 2020)
- Pectus excavatum (inward pointing breastbone) can restrict space for the heart to expand and restrict rib movement, leading to palpitations that are worse lying down. (Tocchioni, 2013)
- Excessive bleeding is more common in people with hEDS. This includes bleeding skin, bruising, heavy menstrual bleeding, hematoma formation, bleeding from the gums, excessive bleeding during surgery. (Wiesmann, 2014; Kumskova, 2022)

Immune Sensitivity

- Medication sensitivity and reactions to medications that are usually well tolerated. People tend to be sensitive to excipients in the medication (not the active ingredient); dyes and alcohol preservatives are common problems.(Schofield, 2019)
- The web site https://dailymed.nlm.nih.gov/dailymed/ provides useful info about excipients. Patients with MCAD may report being “allergic” to many medications. While this might not be a true allergy, not tolerate many medications. (MCAD)
- Intolerance to smells, including perfume, cologne, chemical cleaners. In patients with odor sensitivity, the room and all providers who enter the room need to be fragrance free. Exposure to smells may trigger migraines or cardiac irregularities. (MCAD)

Common Emergency Room Conditions

- Anaphylactic reactions due to MCAD
- Non-epileptic seizures or fainting due to cervical instability or POTS
- Anxiety or panic attacks due to POTS, MCAD, HSD

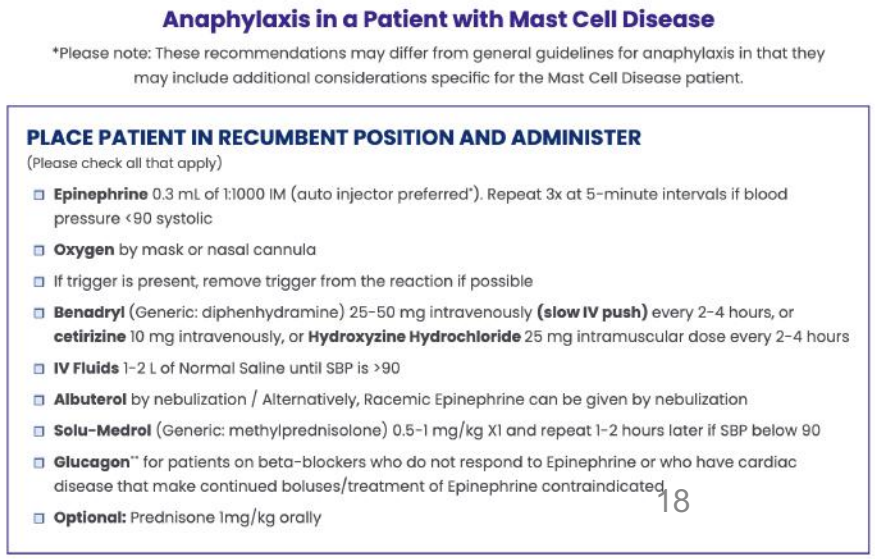
Anaphylaxis Associated with MCAD

Anaphylaxis and Mast Cell Disease

Anaphylaxis is an acute, life-threatening, systemic reaction that results from the sudden, rapid, systemic release of mediators from mast cells and basophils. Anaphylaxis symptoms present as new or worsening symptoms including:

- Mouth: itching, swelling of lips and/or tongue
- Throat: itching, tightness, closure, hoarseness
- Skin: itching, hives, redness, swelling, flushing
- Gut: nausea, vomiting, diarrhea, cramps
- Lung: shortness of breath, cough, wheeze
- Heart: weak pulse, dizziness, passing out

- Symptoms of anaphylaxis: https://tmsforacure.org/anaphylaxis/
- ER MCAD crisis response plan: medications to avoid or use: https://tmsforacure.org/wp-content/uploads/2023/06/TMS_ER-Protocol-2022_fillable.pdf



Anaphylaxis in a Patient with Mast Cell Disease

*Please note: These recommendations may differ from general guidelines for anaphylaxis in that they may include additional considerations specific for the Mast Cell Disease patient.

PLACE PATIENT IN RECUMBENT POSITION AND ADMINISTER

(Please check all that apply)

- ☐ **Epinephrine** 0.3 mL of 1:1000 IM (auto injector preferred*). Repeat 3x at 5-minute intervals if blood pressure <90 systolic
- ☐ **Oxygen** by mask or nasal cannula
- ☐ If trigger is present, remove trigger from the reaction if possible
- ☐ **Benadryl** (Generic: diphenhydramine) 25-50 mg intravenously **(slow IV push)** every 2-4 hours, or **cetirizine** 10 mg intravenously, or **Hydroxyzine Hydrochloride** 25 mg intramuscular dose every 2-4 hours
- ☐ **IV Fluids** 1-2 L of Normal Saline until SBP is >90
- ☐ **Albuterol** by nebulization / Alternatively, Racemic Epinephrine can be given by nebulization
- ☐ **Solu-Medrol** (Generic: methylprednisolone) 0.5-1 mg/kg X1 and repeat 1-2 hours later if SBP below 90
- ☐ **Glucagon**** for patients on beta-blockers who do not respond to Epinephrine or who have cardiac disease that make continued boluses/treatment of Epinephrine contraindicated
- ☐ **Optional:** Prednisone 1mg/kg orally

MCAS Crisis Medications

The Mast Cell Disease Society

Medications to Use and Avoid

Quick Reference Guide

Medications *to avoid* or *use with caution* in patients with mast cell disease in emergency situations

Please note: *Some of the Medications to Avoid may be given if absolutely necessary, if given with a prep to stabilize mast cells. Please refer to one of our mast cell experts for instructions.*

Medication Type	Avoid or Use With Caution	Medications That Are Typically Tolerated
General Medications	• alcohol • amphotericin b • dextran • dextromethorphan • polymyxin B • quinine • vancomycin IV • alpha-adrenergic blockers • beta-adrenergic blockers	• calcium channel blockers • centrally acting alpha 2 adrenergic stimulants • aldosterone antagonists • Oral doses of Vancomycin may be tolerated in some cases.
Pain Medications	• opioid narcotics (may be tolerated by some individuals) • Toradol (ketorolac) • Non-steroidal anti-inflammatory drugs (unless the patient is already taking a drug from this class)	• fentanyl [may require adjunct treatment with Zofran (ondansetron)] • tramadol
General Anesthetics	• atracurium • doxacurium • rocuronium • mivacurium	• pancuronium • vecuronium
Local Anesthetics	• benzocaine • chloroprocaine • procaine • tetracaine	• bupivacaine • lidocaine • mepivacaine • prilocaine • levobupivacaine • ropivacaine
Intraoperative Induction Medications		• ketamine • midazolam • propofol
Inhaled Anesthetics		• sevoflurane

https://tmsforacure.org/wp-content/uploads/2023/06/TMS_ER-Protocol-2022_fillable.pdf#emergencyalert

Musculoskeletal Issues

- Trauma that might not be sufficient to cause injury in a non-hypermobile person may cause injury in HSD/hEDS. For example, a patient may sublux or dislocate a joint rolling over in bed or sneezing. Non-traumatic dislocations and subluxations are common and can be very painful, even if the joint has relocated. (HSD) (Wiesmann, 2014)
- Joints become more unstable as bedrest deconditions muscles that normally provide support. Patients may report hips, shoulders or ribs 'slipping out' or other sharp pain associated with subluxations. (HSD)
- Subluxed ribs may cause crushing chest pain that may feel like a heart attack. (HSD)

Skin Issues

- Fragile skin requires specialized procedures for suturing.(HSD)(Castori, 2012)
- Fragile skin can lead to skin breakdown more quickly than in non-hypermobile patients.(HSD)(Doolan, 2023)
- Skin can be more easily damaged by tape (including EKG pads, tape used to stabilize IV needles. Skin is also vulnerable to abrasion.(Doolan, 2023)
- Mast cell activation disorder (MCAD) can lead to severe tape/adhesive allergies, including to EKG electrodes.(Mihele, 2023) (MCAD)

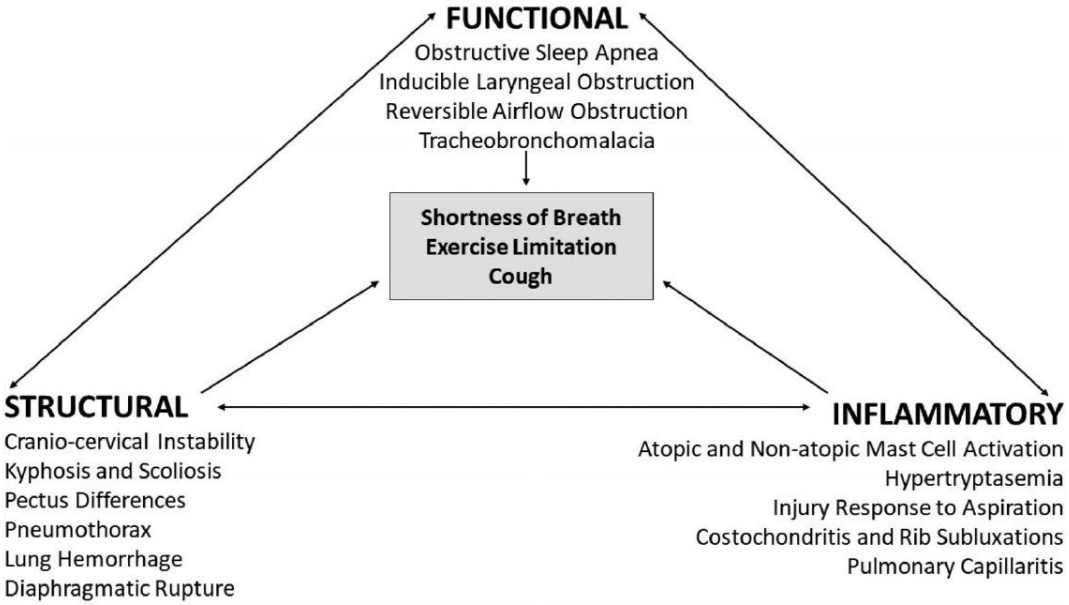
Bleeding Issues

- Excessive bleeding is more common in people with hEDS. This includes bleeding skin, bruising, heavy menstrual bleeding, hematoma formation, bleeding from the gums, and excessive bleeding during surgery
- The International Society of Thrombosis and Haemostasis bleeding assessment tool (ISTH-BAT) can be used to assess for bleeding disorders. Research shows that 62% of patients with HSD had abnormal ISTH-BAT scores indicating bleeding disorders and high risk of hemorrhagic complications
- Women with heavy menstrual bleeding should be screened for bleeding disorders.
- Patients with hEDS/HSD are more likely to have negative Von Willebrand Disease and hemophelia tests in spite of having a bleeding disorder

Wiesmann, 2014; Kumskova, 2023; Wright, 2023; Kendel, 2023

Respiratory Issues

- An acute POTS attack may present with difficulty breathing. (POTS)
- MCAD reactions causing sinus congestion can lead to increased mouth breathing. Mouth breathing contributes to TMJ pain, headaches, tinnitus.
- Pectus excavatum (inward pointing breastbone) can restrict space for lungs to expand, and can restrict rib movement. (Tocchioni, 2013)



Bascom R, Dhingra R, Francomano CA. Respiratory manifestations in the Ehlers-Danlos syndromes. *Am J Med Genet C Semin Med Genet*. Dec 2021;187(4):533-548. doi:10.1002/ajmg.c.31953

Obstetric Issues

- Complications with delivery appear to be more common in HSD/hEDS. This includes uterine torsion, cervical incompetence, preterm labor, severe perineal tearing, and failure of sutures for episiotomies and C-sections. Separation of the pubic symphysis and coccyx dislocation may also occur.(Gilliam, 2020)
- MCAS issues may also complicate pregnancy.(Dorff, 2019)

Surgical Precautions

- Orthopedic surgery (e.g. rotator cuff or ACL repair) is only effective 34% of the time in patients with hEDS; this is 50% as often as non-hypermobile pts. Therefore, it is important that conservative management (with an hEDS knowledgeable provider) be fully explored before resorting to surgery.
- Orthopedic surgeries in hEDS are more likely to have complications, with one study reporting 91% complication rate.
- Surgeons should take hEDS into account when planning surgery.
- Spinal surgery complications rate is higher in HSD/hEDS than in the general population.
- Gastrointestinal surgeries in hEDS are more likely to have complications, but fewer complications than vascular EDS. Typical complications include arterial perforation or tears, bowel perforation or tears, recurrent hernias or increased bleeding.

Rombaut, 2011; Yonko, 2021; Homere, 2020; Chi, 2023; Kulas Søborg, 2017; Burcharth, 2012)

Surgical and Anesthetic Precautions:

Hypermobility Spectrum Disorder (HSD) and Hypermobile Ehlers Danlos Syndrome (hEDS)

The main feature of HSD/hEDS is ***laxity of connective tissue***, including skin, ligaments, blood vessels and nerves. This can cause ***potentially fatal problems*** for these patients when unconscious, and/or having surgery.

BEWARE THE UNCONSCIOUS PATIENT!	***In the unconscious HSD/hEDS patient, a little force may displace any joint.*** ***Treat unconscious HSD/hEDS patients with full spinal stabilization*** as if they have a spinal injury. If you don't, then you may cause one! ***Use NO traction on limbs.*** ***Use extreme care with the chest:*** the ribs easily dislocate front or back.
BEWARE THE LARYNGOSCOPE!	***Use extreme gentleness, with minimal, if any, anterior traction on the laryngoscope. The jaw may dislocate*** on one or both sides. Manipulation of the laryngoscope can also damage the cricopharyngeal muscle and its nerves, the esophagus and the cervical spine.
BEWARE NECK MOTION!	***Keep patient's head in neutral position throughout.*** Movement of unstable subcranial joints may cause spinal cord damage during incautious patient handling during anesthesia. Consider a soft collar.
LOCAL ANESTHESIA	HSD/hEDS patients are often resistant to local anesthetics: ***they may need much larger doses than other patients, and these may need to be repeated during a procedure.*** Ropivacaine may work better than lidocaine or bupivacaine.
SURGICAL TECHNIQUE	Use minimal force when cutting or moving tissues. Cut blood vessels may contract poorly: **electrocautery is appropriate**. Tissue healing may be prolonged. **Close layers without tension using slowly-absorbable or non-absorbable sutures**. Reinforce them with steri-strips etc. as appropriate.
BLEEDING & BRUISING	These are due to fragile small blood vessels, not an intrinsic blood disorder, so **elaborate clotting tests are rarely indicated**. Be alert for slowly-accumulating, deep hematomas.
POST-OPERATIVE PAIN	Painful polyneuropathy is common in HSD/hEDS. Post-operative pain may be more severe and more prolonged than normal. **Be liberal with analgesics**.
CARDIO-VASCULAR INSTABILITY	HSD/hEDS patients are subject to hypotension and/or tachycardia due to low blood volume, and defective venoconstriction. ***Liberal IV fluids usually can address this.***
GI DYSFUNCTION	Poor GI motility is routine in HSD/hEDS, worse after surgery. **Minimize constipating agents, and use laxatives pre-emptively**. Consider pro-motility agents.
CARDIAC RESCUCITATION	Some HSD/hEDS patients have **loose costosternal joints,** sometimes palpably displaced. For them, **chest compressions could in theory be very dangerous, causing rib detachments, a flail chest and even heart or lung puncture** by freed anterior ribs. There is no consensus on whether cardiac resuscitation should include chest compressions in patients with clear evidence of rib displacements.

Alan Spanos, MD, (919) 967-2927, alan.spanos@yahoo.com.

This document is online at *www.AlanSpanosMD.com*. It was updated March 2019.

For more information, see the Ehlers Danlos Society at ehlers-danlos.com.

Handout for patients to share with surgeons and anesthesiologists

https://edswellness.org/wp-content/uploads/2019/07/7-Surgical-Anesthetic-Precautions.pdf

Surgical Issues Related to Positioning

- Patients with possible cervical instability require careful neck positioning during surgery, especially in cases of intubation. In some cases, use of a rigid cervical collar during surgery is a wise precaution.(Castori, 2012)
- Intubation may cause subluxation of the temporomandibular joint or damage to the disc.(Wiesmann, 2014)
- Joints and tissues not being operated on may be stressed or damaged by positioning. For example, shoulder hyperabduction may cause a brachial plexus injury.(Wiesmann, 2014)

Surgical Issues: Tissue Fragility

- Shear forces may damage skin, for example when rolling or transferring patients. Gripping patients for rolling or transferring may cause bruises.
- Tissue healing is delayed; therefore recovery is slower and rehab may need to proceed slower.
- Intubation may cause damage to fragile tracheal mucosa. A smaller endotracheal tubes may be less damaging.
- There is increased risk of bleeding due to vascular fragility. See Wiesmann, 2014, for extensive discussion of operative bleeding.
- Tourniquets may cause bruising and hematoma formation.
- Any surgery with devices moved within the body (laparoscopy, colonoscopy, etc.) have increased chance of damaging tissues.
- Special procedures recommended for skin sutures: closer together, leave sutures in longer.

(Wiesmann, 2014; Ericson, 2017; Burcharth, 2012; Castori, 2012)

Surgical Precautions due to POTS

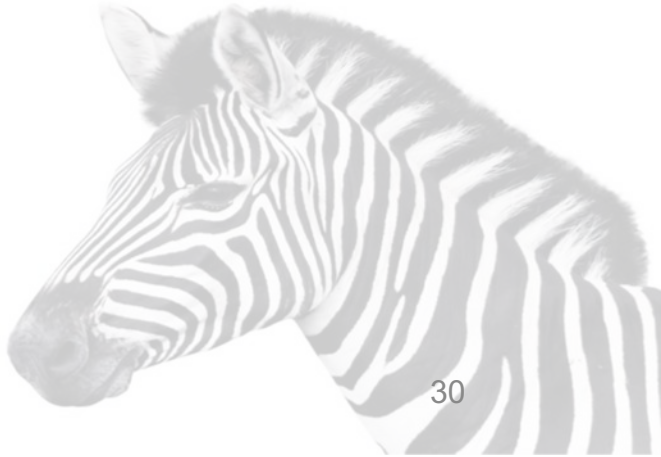
- Orthostatic intolerance (e.g. POTS) may lead to abnormal response to anesthesia.
- Orthostatic intolerance may result in poor regulation of blood pressure after surgery, interfering with getting patient upright after surgery; this may interfere with physical therapy. Since anesthesia can cause POTS flares, the patient may be more POTS-reactive after surgery than before.
- Consider increased hydration before and after surgery.
- Use hypotensive agents, sympathomimetics, catecholamines, and vasodilators with caution. Use sedating drugs sparingly.

(Ruzieh, 2018)

Surgical Precautions due to MCAD

- Reactions to medications, or increased reactivity if MCAD medications were discontinued for surgery.
- Reactions to adhesives in tape, EKG pads, etc

(Lacerna, 2021)



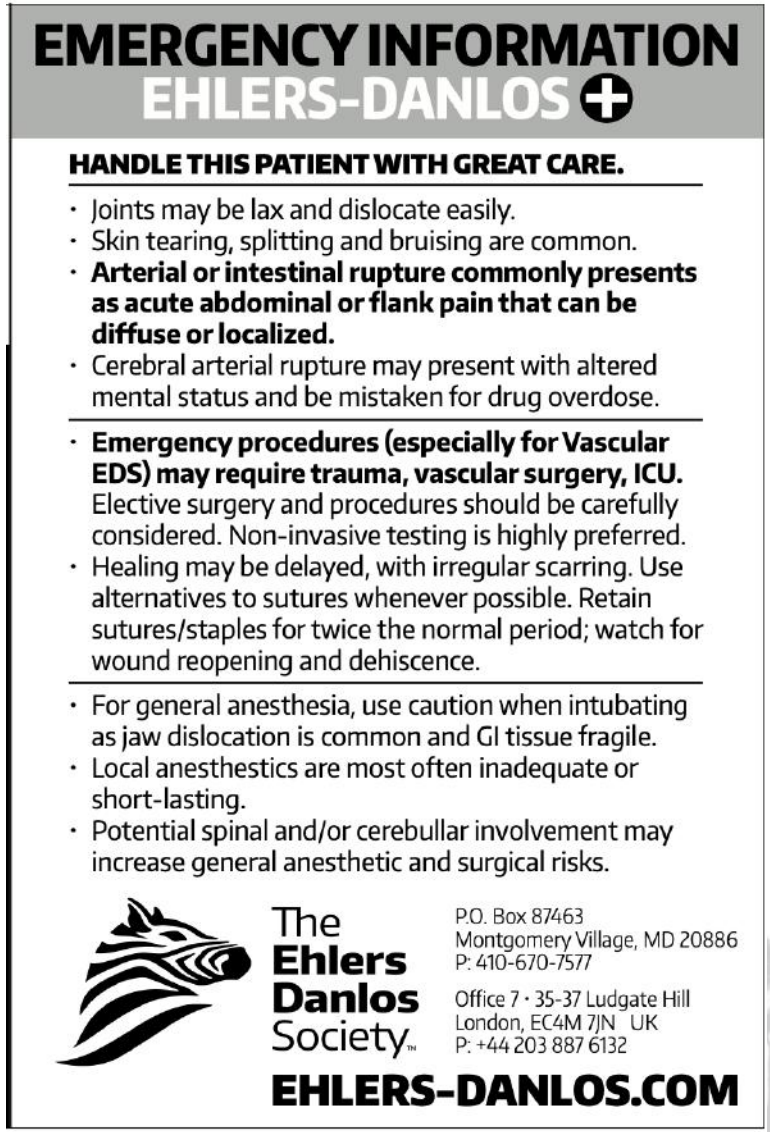
Summary

- There are lots of reasons why people with HSD/hEDS, POTS and MCAD may present with medical issues that require treatment in the hospital
- Discuss HSD, POTS, MCAD with providers when having for surgery
- Be aware of special issues you may need when receiving emergency care
- Communicate with your care providers if you have HSD/hEDS, POTS, or MCAD

Resources

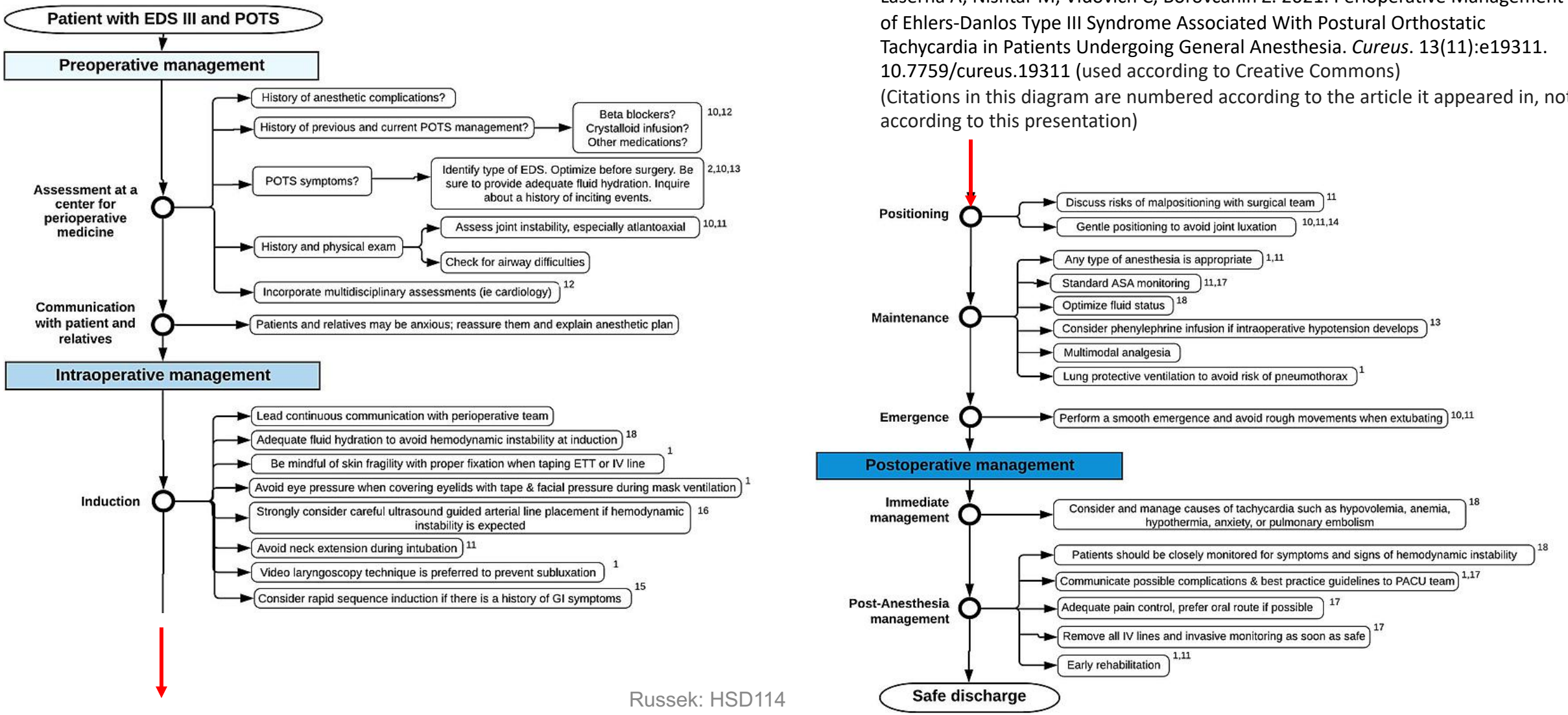
- A one-page sheet of precautions to share with doctors: https://edswellness.org/wp-content/uploads/2019/07/7-Surgical-Anesthetic-Precautions.pdf
- Web site with info to share with your doctors: https://hypermobilityclinic.org/surgical-and-anesthetic-precautions-for-hypermobile-and-eds-patients/
- It includes a link to the article with surgical flow chart: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4223622/

Handle With Care!



This emergency card, can be accessed on the Ehlers-Danlos Society website: https://www.ehlers-danlos.com/wp-content/uploads/2022/11/walletcard2017combined.pdf

Recommended Surgical Precautions –



Laserna A, Nishtar M, Vidovich C, Borovcanin Z. 2021. Perioperative Management of Ehlers-Danlos Type III Syndrome Associated With Postural Orthostatic Tachycardia in Patients Undergoing General Anesthesia. *Cureus*. 13(11):e19311. 10.7759/cureus.19311 (used according to Creative Commons)

(Citations in this diagram are numbered according to the article it appeared in, not according to this presentation)

Surgical Concerns

Chopra P, Bluestein L. Perioperative Care in Patients with Ehlers Danlos Syndromes. *Open Journal of Anesthesiology*. 2020;10:13-2913. doi:10.4236/ojanes.2020.101002. Reproduced according to open access license. Available at https://www.scirp.org/pdf/ojanes_2019123013435518.pdf .
Citation numbers on this page refer to citations in that article, not this lecture.

Table 1. Considerations for dealing with patients with EDS.

Head and neck (6)	Chiari malformation
	Brain stem compression
	Idiopathic intracranial hypertension
	Atlantoaxial instability
	Craniocervical instability
	Epilepsy
	Intracranial aneurysm
	Temporomandibular joint dysfunction (7)
	Headaches and migraines (8)
	Intermittent compression of vertebral arteries
	Compression of upper cervical nerve roots from C0 to C2 hypermobility (6))
Spine (6)	Segmental kyphosis and instability
	Tethered cord syndrome
	Tarlov cyst syndrome (8)
	Meningeal ecstasias/cysts
	Spontaneous cerebrospinal leak (9)
Cardiovascular	Structural defects such as mitral valve prolapse and aortic root dilatation (hEDS) (11, 17, 18)
	Dysautonomia (10)
	Increased peripheral pooling (12)
	Severe progressive cardiac-valvular aortic valve and mitral valve problems (cvEDS)
Pulmonary	Tracheomalacia
	Rib subluxations
	Pulmonary bullae
	Decreased pulmonary volumes secondary to kyphoscoliosis
	Obstructive sleep apnea (13)
GI (19)	Gastroparesis
	Intestinal dysmotility (14)
	Visceroptosis (15)
	Hollow visceral (intestines, uterus) rupture
	Rectal/uterine prolapse (16)
	Visceral fragility (dEDS and vEDS)
Urinary	Neurogenic bladder (6, 17)
	Interstitial cystitis (20)
Musculoskeletal (8)	Joint subluxations and dislocations (8)
	Myopathy (25)
	Fatigue (22)
	Poor joint proprioception (23)
	Muscle weakness, axonal polyneuropathy, atrophy of muscles of hands and feet (24)
Hematologic	Bleeding disorders (20)
	Mast cell activation syndrome (20, 21)
	Vascular fragility
Neurological	Dysautonomia (22)
	Postural orthostatic tachycardia syndrome (22,)
	Neuropathy (26, 27)
	Small fiber peripheral neuropathy (28)
	Entrapment neuropathy (24, 27, 28)
	Unpredictable response to local anesthetic

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Thank You!

Questions?