HSD/EDS, MCAD, POTS/OI In The Hospital

Disclaimer: All medical decisions regarding patient care should be made by a qualified medical provider. This document should not replace proper medical care by qualified providers.

Abbreviations: Hypermobility Spectrum Disorder (HSD); hypermobile Ehlers-Danlos Syndrome (hEDS); mast cell Activation Disorder (MCAD); postural orthostatic tachycardia syndrome, (POTS); orthostatic intolerance (OI).

EMERGENCY INFORMATION

If the patient reports having HSD/hEDS, MCAD, or POTS/OI, or if you suspect one of those conditions:

1. If the patient is having an **anaphylactic reaction**, especially to medications, this may be an MCAD reaction. See https://tmsforacure.org/wp-content/uploads/2023/06/TMS_ER-Protocol-2022_fillable.pdf for an Emergency Room Response Plan from The Mast Cell Disease Society.(MCAD)
   a. Grading System for Anaphylaxis Severity in Mast Cell Activation Disorder, shown in table below. (https://www.mastcellhope.org/education/emergency-treatments/)

Grade I	Grade II		Grade III		
Local reaction (No systemic Reaction)	Mild to moderate systemic reaction (Systemic reaction without cardiovascular and/or respiratory involvement)		Severe systemic reaction = anaphylaxis (Systemic reaction with cardiovascular and/or respiratory involvement)		
Grade I	**Grade II A**	**Grade II B**	**Grade III A**	**Grade III B**	**Grade III C**
Local reactions: e.g. •Redness •Swelling •Pruritus	Skin: e.g. •Urticaria •Angioedema •Flush	Skin plus GI-Tract: e.g. •Urticaria •Angioedema •Flush plus e.g. •Abdom. pain •Vomiting •Diarrhoea	Respiratory: •Cough •Wheezing •Stridor	Severe Respiratory: e.g. •Objective dyspnoea •Accessory muscles and/or Severe Cardio-vascular: •Shock	Reanimation: •Respiratory arrest and/or •Cardio-vascular arrest
	or GI-Tract: e.g. •Abdom. pain •Vomiting •Diarrhoea		or Cardio-vascular: •Tachycardia •Lowered blood pressure		

2. If the patient is having **syncopal events or non-epileptic seizures (aka pseudo-seizures**), consider POTS/OI[1,2] or cervical instability.[3] Do not attribute to psychogenic causes without appropriate assessment. Although the patient may be anxious, these reactions are not simply anxiety or panic attacks.
   a. POTS patients may be significantly improved after IV fluids while waiting for diagnostic testing. Do not tell them that there was nothing wrong, as their physiological response was real.
   b. Upper cervical instability (UCI) can also lead to non-epileptic seizures or syncopal events.[3] Note that supine imaging will usually not detect cervical instability or compression of vital structures, so cannot be used to rule out cervical instability.[4]
   c. Fant, 2017 provides a useful guide for evaluating pediatric syncope in the ER.[5]
3. Patients with **upper cervical instability** might not tolerate any neck movement from neutral. Use caution with positioning and intubation. Quote from Russek, 2022[3]: "Upper cervical instability (UCI) can result in myelopathy, cranial nerve neuropathy, brainstem compression, vertebrobasilar artery compromise and compromised venous or cerebrospinal fluid outflow. Vertebrobasilar artery problems are likely associated with AAI, while cervical medullary syndrome, cranial nerve problems and cerebrospinal fluid obstruction are likely associated with AOI. Symptoms of UCI include: headaches, neck or facial pain, dizziness, vertigo, nausea, paresthesias, dyspnea, dysphonia, vision changes (blurred or tunnel vision, visual aura), hearing changes, dysphagia, choking, sleep apnea, memory deficits, and pre-syncopal episodes. Signs associated with UCI include: long-tract findings such as hyper-reflexia, positive Babinski and Hoffman's signs, loss of abdominal reflex, dysdiadochokinesia, as well as bowel/bladder problems, gait/balance deficits, weakness of arms and legs, sleep apnea and syncopal episodes. Dysautonomia is more likely to be present and severe with UCI and cervical myelopathy." (see full article for citations)
4. **Anxiety or panic attacks**: POTS, MCAD and HSD can all cause signs and symptoms that may lead to anxiety and panic. Assessment should look for potential medical/physical causes or contributing factors for anxiety.
   a. POTS can cause excessive sympathetic nervous system activation, including tachycardia.[6]
   b. MCAS can present as anxiety and panic. [7]
   c. HSD may be associated with a variety of acutely painful issues that may contribute to anxiety and panic.

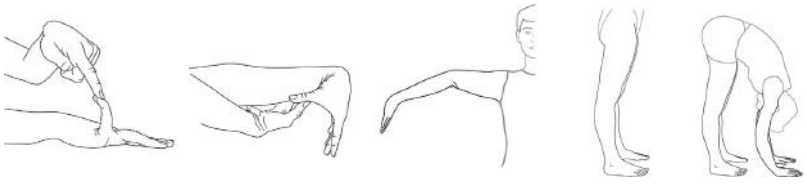
Basics of Hypermobility Spectrum Disorder (HSD); hypermobile Ehlers-Danlos Syndrome (hEDS); mast cell Activation Disorder (MCAD); postural orthostatic tachycardia syndrome, (POTS); orthostatic intolerance (OI)

HSD/hEDS are heritable connective tissue disorders associated with widespread connective tissue fragility and hyperelasticity. Although hypermobile joints are the most visible finding, connective tissue and collagen are present in most tissues. Therefore, patients with HSD/hEDS can present with multisystem involvement including fragile skin and poor tissue healing, dysautonomia (including postural orthostatic tachycardia syndrome, POTS, and orthostatic intolerance), headaches/migraines, gastrointestinal dysfunction, urogenital issues, immune reactivity including mast cell Activation Disorder (MCAD), fatigue, central sensitization. While many healthcare providers think HSD/hEDS is a rare condition and they do not treat these patients, experts contend that the condition is underrecognized and underdiagnosed.[8-10] Because people with HSD/hEDS often have signs and symptoms driving them to seek health care, the prevalence of symptomatic hypermobility is reported as 30% in primary care, 39% in pain clinics, and 37% in rheumatology clinics.[10] These patients WILL be in the hospital at some point. HSD/hEDS frequently exists as a 'trifecta' along with POTS and MCAD, and additional atypical presentations and precautions are associated with POTS and/or MCAD. The complex, multisystem presentation has led to the recommendation "When you can't connect the issues, think connective tissues."

Common signs and symptoms associated with each component of the "trifecta" [2,11,12]

HSD/hEDS	Dysautonomia (POTS/OI)	Mast Cell Activation Disorder (MCAD)
• Musculoskeletal pain or injury in response to minor trauma • Easy/nontraumatic dislocation/subluxation • Dysautonomia, POTS, orthostatic intolerance • MCAD • 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	• Cardiac irregularity, tachycardia, bradycardia. Tachycardia may present as anxiety or panic attack. • Orthostatic intolerance, low BP • Dizziness, presyncope, syncope • Migraine • Severe fatigue, exercise intolerance • Peripheral/dependent edema • Raynaud's or acrocyanosis (purple blotchy feet/hands) • Dumping syndrome/rapid gastric emptying (diarrhea), nausea, vomiting	• Anaphylaxis • Intolerance to medication, sometimes the excipients rather than the active ingredient. • Intolerance to odors, including perfumes, colognes, chemical cleaners. • Food intolerances. • IBS, SIBO, leaky gut • Adhesive sensitivity for tape or sensors. • Migraine

The Beighton Scale, a 9-point scale shown below (1 pt for each side, and the spine) is the standard tool for identifying generalized joint hypermobility (GJH). In adults, ≥5/9, children ≥6/9 indicates GJH. Patients may also earn 1 point for historical hypermobility (≥2 Yes responses on 5PQ). However, these are not the only joints that can be hypermobile, unstable, or symptomatic. There are other hypermobility grading criteria that include other joints.[9] If it is not appropriate to assess the Beighton Score, a score of ≥2 on the 5-Point Questionnaire (5PQ below) can also identify people with hypermobility. Do not rule out HSD/hEDS simply because the patient does not currently meet the Beighton criteria.



5-Point Questionnaire

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
4. As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
5. Do you consider yourself "double-jointed"?

Issues affecting people with HSD/POTS/MCAD that might arise in the hospital, organized by system:

Musculoskeletal issues

- Trauma insufficient 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) [14]
- 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)

Cardiovascular/dysautonomia

- Dysautonomia can lead to orthostatic intolerance: HR may be abnormally high (postural orthostatic tachycardia, POTS) or blood pressure may drop (orthostatic hypotension). Dysautonomia can be precipitated or exacerbated by: bedrest, surgery, anesthesia, COVID. It most commonly presents in adolescent and adult women. While POTS is the most common form of dysautonomia, tachycardia is not always present. [15] POTS treatment guidelines: https://www.onlinecjc.ca/article/S0828-282X(19)31550-8/fulltext) Vernino, 2021[2] provides a good overview of POTS. (POTS)
- POTS can lead to syncopal events, which may include myoclonic jerking that looks like a seizure.[16](POTS)
- Although an upright tilt table test (UTTT) is the gold standard, the NASA Lean Test is accurate and can be performed in 15 minutes bedside in any patient who can stand for 10 minutes. https://batemanhornecenter.org/wp-content/uploads/2016/09/NASA-Lean-Test-Instructions-1.pdf (POTS)
- For patient self-care strategies, see POTS Self-Care Checklist at https://webspace.clarkson.edu/~lrussek/research.html. (POTS)
- 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.[17] (HSD)
- Pectus excavatum can restrict space for heart expansion,and restrict rib movement, leading to palpitations.[18]
- Aortic dissection can occur in some forms of EDS, but is not more common in HSD/hEDS.[19] (HSD)

Bleeding disorders

- 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. [14,20,21, 22](HSD)
- 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.[20] (HSD)
- 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.[21] (HSD)

Immune sensitivity

- Emergency Room MCAD Response guidelines by TMSforacure: https://tmsforacure.org/wp-content/uploads/2023/06/TMS_ER-Protocol-2022_fillable.pdf (MCAD)
- 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. [23] 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)
- MCAD can be assumed if the patient scores ≥14 on the Validated MCAD Questionnaire available at: https://merridycasson.com/wp-content/uploads/2021/07/Allergy-Reactivity-MCAS.pdf (MCAD)

Neurological issues

- Craniocervical and atlantoaxial instability (CCI and AAI), Chiari I malformation, CSF leaks, idiopathic intracranial hypertension, Eagle syndrome, tethered cord and Tarlov cysts are all more common in HSD than in the general population.[13](HSD)
- Upper cervical instability may compress the brainstem, cranial nerves or vascular structures serving the brain and may block CSF flow into the spinal cord. (HSD) [24] Patients may present with:
    - Pseudo seizures/non-epileptic seizures, drop attacks and dystonia. These are real, physical problems and should not be treated as psychogenic.
    - People with HSD often experience instability flares with MCAD flares, so ask about MCAD if the patient has a recent increase in seizure-like episodes.
- Syncopal events may mimic seizures, especially when they include myoclonic jerking. [16](POTS)
- Migraines/headaches
    - Migraines are extremely common in POTS, and some sources suggest that all patients with migraine be evaluated for POTS. [25] POTS is often a component of Long-COVID.[26](POTS)
    - Migraines are extremely common in MCAD due to histamine release in the brain causing neuroinflammation.[27]
    - Craniocervical and atlantoaxial instability (CCI and AAI), Chiari I malformation, CSF leaks, idiopathic intracranial hypertension, Eagle syndrome are all more common in HSD than in the general population. Consider these possibilities when evaluating severe headaches, especially when positional. [13] (HSD)
    - Use caution if/when providing Botox injections to manage migraines, as muscle trigger points often develop in an attempt to provide stability to an unstable cervical spine. By relaxing muscles, Botox may remove the only source of stability these patients may have.[28] The long-term solution is for physical therapy to educate patients about proper posture and body mechanics, and provide motor control training to improve function of the deep stabilizing muscles. (HSD)
- Cognitive issues/brain fog are common in all three conditions. Other aggravating factors include over-breathing, chest breathing, poor quality/quantity of sleep, and medication sensitivities. (HSD/POTS/MCAD)
- Functional Neurological Disorders seem to be more common in people with EDS. It is a neural processing disorder, not a psychological condition; it is not always a response to trauma. Patients present with unusual and inconsistent neurological signs and symptoms, including pseudoseizures, tremors, limb weakness, episodic paralysis, sensory and visual disturbance.[29] Excellent overview at https://www.fndaction.org.uk/wp-content/uploads/2022/10/Information-guideance-sheet-for-medical-professionals.pdf. (HSD/MCAD)
- Supine MRI or CT with the neck supported in midrange might not detect instability associated with gravity or excessive motion. Flexion, extension and rotation MRI/CT and upright MRI are sometimes preferred. (HSD)

Gastrointestinal issues

- Multiple GI issues are common in all three conditions. [30-32](HSD/POTS/MCAD)
- Median arcuate ligament syndrome (MALS) and superior mesenteric artery syndrome (SMAS) both cause post-prandial vomiting and such severe pain after eating that patients may refuse to eat, leading to weight loss. This may look like an eating disorder, but inability to eat is due to intense pain. MALS and SMAS are more common in hEDS and POTS than in the general population.[33,34]
- People with gastroparesis and delayed gastric emptying due to HSD might benefit from starting colonoscopy prep earlier than most patients due to slower transit times.(HSD) (anecdotal)

Dermatologic issues

- Fragile skin requires specialized procedures for suturing.[35] See chart, below. (HSD)
- Fragile skin can lead to skin breakdown more quickly than in non-hypermobile patients.[36] (HSD)
- Skin can be more easily damaged by tape or abrasion.[36] (HSD)
- MCAD can lead to severe tape/adhesive allergies, including to EKG electrodes.[37] (MCAD)

Anxiety/panic/depression

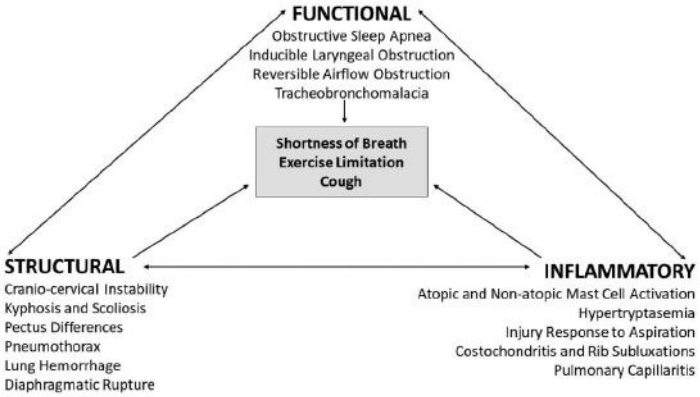
- The hyperadrenergic subtype of POTS is frequently associated with anxiety/panic episodes and patients have frequently been first diagnosed with anxiety or panic disorder due to the tachycardia and sympathetic flare associated with acute POTS attacks.[2] If anxiety/panic is significantly reduced by having the patient lie supine and/or receive IV fluids, consider POTS. (POTS)

Urogenital

- Urogenital issues are more common in HSD [38] (HSD)

Respiratory issues

- See chart, right.[39] (HSD)
- An acute POTS attack may present with difficulty breathing. (POTS)
- MCAD reactions causing sinus congestion can lead to increased mouth breathing.
- Pectus excavatum (sunken breastbone) can limit space for lungs and restrict rib movement.[18]



Obstetric

- MCAS issues may complicate pregnancy.[40]
- Increased risk of bleeding may complicate delivery.[21]
- 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.[38]

Surgical Precautions and Considerations in HSD, POTS and MCAD

Surgeons should consider precautions in people with hypermobile Ehlers-Danlos Syndrome (hEDS)/ Hypermobility Spectrum Disorder (HSD). Connective tissue, skin and vascular tissues are generally more fragile in hEDS compared to non-hEDS patients. However, skin and vascular tissues are more robust in hEDS compared to classical and vascular EDS, and surgery is less risky in hEDS compared to other forms of EDS.

Potential complications and decreased surgical success rate

- 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.[41,42]
- Orthopedic surgeries in hEDS are more likely to have complications, with one study reporting 91% complication rate.[42]
- Surgeons should take hEDS into account when planning surgery.[43,44]
- Spinal surgery complications rate is higher in HSD/hEDS than in the general population.[45]
- 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.[46,47]

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.[35,44]
- Intubation may cause subluxation of the temporomandibular joint or damage to the disc.[14]
- Joints and tissues not being operated on may be stressed or damaged by positioning. For example, shoulder hyperabduction may cause a brachial plexus injury.[14,44]

Issues related to tissue fragility

- Shear forces may damage skin, for example when rolling or transferring patients. Gripping patients for rolling or transferring may cause bruises.[14]
- Tissue healing is delayed; therefore recovery is slower and rehab may need to proceed slower.[48]
- Intubation may damage fragile tracheal mucosa. Smaller endotracheal tubes may be less damaging. [14,44]
- There is increased risk of bleeding due to vascular fragility. See Wiesmann, 2014,[14] for extensive discussion of operative bleeding.
- Tourniquets may cause bruising and hematoma formation. [14]
- Laparoscopic surgery may require higher pressures for peritoneal inflation, causing increased risk of pneumothorax. [14]
- Post-dural puncture headache (PDPH) may occur due to increased risk of CSF leak.[14]
- Special procedures recommended for skin sutures: closer together, leave sutures in longer.[35,47]

POTS issues that may occur with surgery:

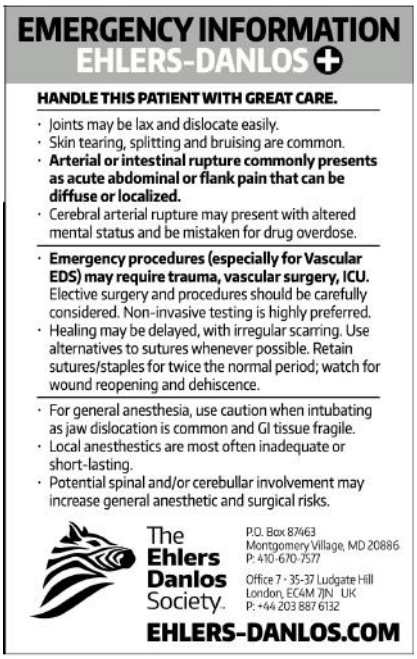
- Orthostatic intolerance (e.g. POTS) may lead to abnormal response to anesthesia. [49]
- Orthostatic intolerance may result in poor regulation of blood pressure after surgery, interfering with getting patient upright after surgery; this may interfere with physical therapy. Since anesthesia can cause POTS flares, the patient may be more POTS-reactive after surgery than before.[49]
- Consider increased hydration before and after surgery.[44]
- Use hypotensive agents, sympathomimetics, catecholamines, and vasodilators with caution. Use sedating drugs sparingly.

MCAD issues that may occur with surgery:

- Reactions to medications, or increased reactivity if MCAD medications were discontinued for surgery. See https://tmsforacure.org/wp-content/uploads/2023/06/TMS_ER-Protocol-2022_fillable.pdf.
- Reactions to adhesives in tape, EKG pads, etc. [44] Pretreatment of the skin with 10 minutes of Pepto Bismol or Milk of Magnesia can decrease skin reactivity.

Resource Materials

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



The following table, published in 2012, remains an excellent guideline for surgical precautions in hEDS.[35]

Table 1. Surgical and anesthetic recommendations in the JHS/EDS-HT

Evidence	Ref No.	Recommendation
Surgical procedure		
(1) Orthopedic surgery is paradoxically associated with pain worsening in JHS/EDS-HT; anecdotal observations suggest a low success rate for abdominal surgery in functional disorders	5	Consider more conservative treatments as an alternative to non-life-threatening operations
(2) Although soft tissue fragility is not severe in JHS/EDS-HT, delayed wound healing with consequent suture widening, suture dehiscence and postsurgical hernias are possible complications	1, 6	(a) Perform skin closure in two layers (cutaneous and subcutaneous) without excessive tension (b) Use generous sutures, deep stitches and steri-strips as reinforcement devices (c) Leave sutures twice as long as normally recommended
(3) Minor bleeding disorders are common in JHS/EDS-HT	7	Consider preoperative prophylaxis with desmopressin (1-deamino-8-D-arginine vasopressin), especially in patients with a positive history for mucosal bleeding (nose, gingivae, bowel, bladder, etc.) and/or easy bruising
(4) Episiotomy is associated with an increased risk for pelvic prolapses in JHS/EDS-HT women	8	Consider cesarean section as first-choice delivery procedure
Anesthetic procedure		
(5) Dysautonomia is a major feature in JHS/EDS-HT and may need special anesthetic considerations	9	(a) Consider to carry out appropriate investigations (e.g. tilt test) before any intervention in order to properly plan the anesthetic procedure, especially in patients with cardiovascular symptoms (b) In case of confirmed dysautonomia, consider prophylactic early fluid loading and phenylephrine infusion
(6) JHS/EDS-HT patients often display resistance to intradermal lidocaine infiltrations and topical EMLA cream	10, 11	Consider alternative anesthetic procedures or double the anesthetic dose
(7) Epi/peridural anesthesia may be hampered by severe spondylosis and/or scoliosis, and could be complicate by intraspinal hypotension due to increased meningeal weakness in JHS/EDS-HT	None[1]	Favor total anesthesia in case of major surgery
(8) Temporomandibular joint dysfunction and occipitoatlantoaxial instability may be more common in JHS/EDS-HT	13, 14	Perform intubation with care and consider the use of pediatric devices also in adults
Postsurgery recovery		
(9) Muscle deconditioning due to inactivity rapidly worsens chronic pain and fatigue in JHS/EDS-HT	15	Consider early physical therapy support in case of surgery with postoperative bed rest for >7 days

[1] Reports specifically describing such likely complications are lacking. However, mild scoliosis and premature spondylosis are commonly encountered in the JHS/EDS-HT clinic, while some preliminary studies indicate that generalized joint hypermobility is associated with orthostatic headache [12].

- A handout to print and share with your surgeon: https://edswellness.org/wp-content/uploads/2019/07/7-Surgical-Anesthetic-Precautions.pdf

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.

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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.

Considerations for surgical intervention for patients with EDS[50]

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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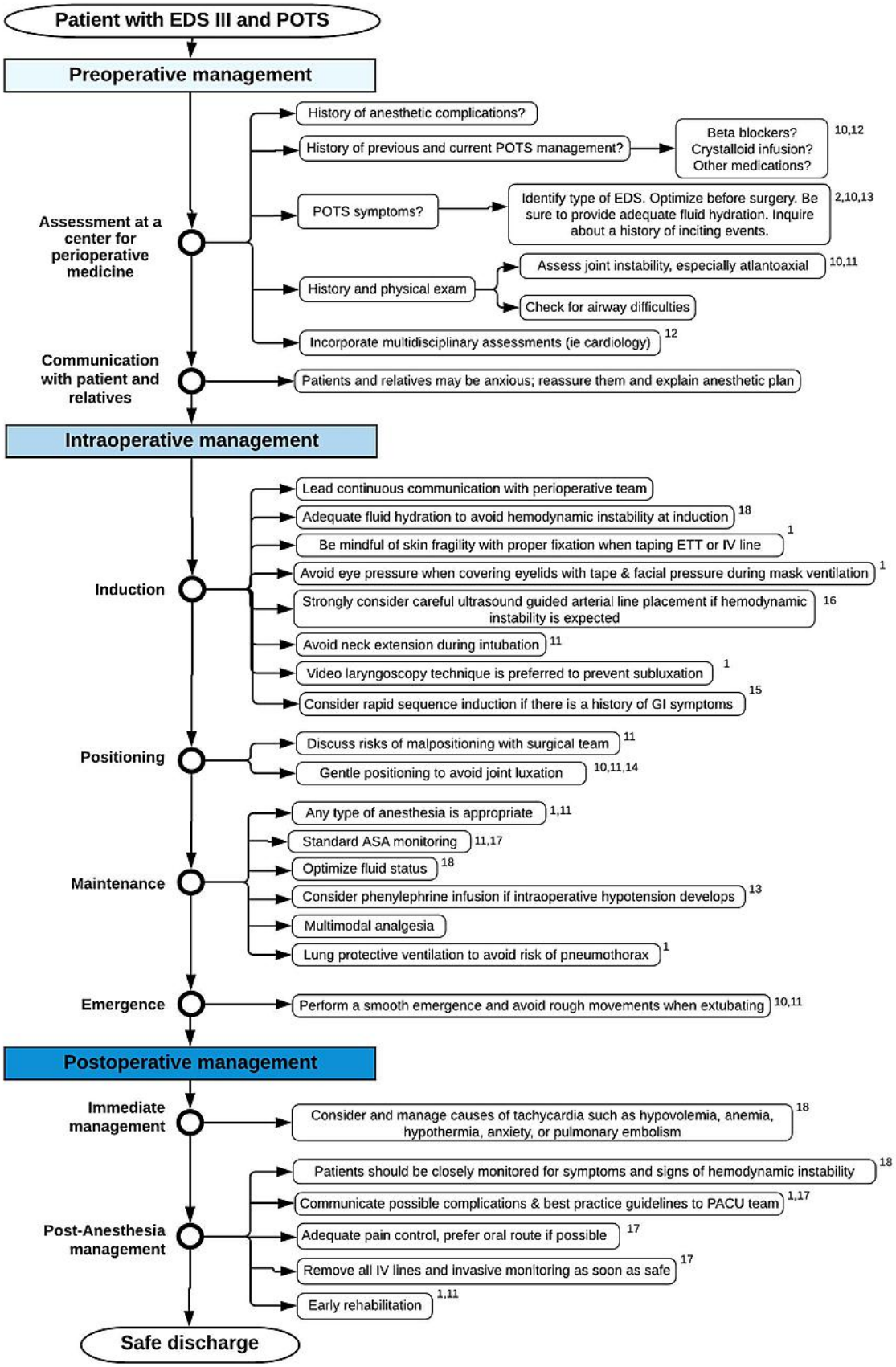
Perioperative flow diagram for the management of patients with EDS III and POTS[44]



POTS: Postural Orthostatic Tachycardia Syndrome, EDS III: Ehlers-Danlos Syndrome Type III, ASA: American Society of Anesthesiologists, PACU: Post-Anesthesia Care Unit, ETT: endotracheal tube, IV: Intravenous. (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)

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