Hypermobility 116: Excessive Bleeding in HSD/hEDS

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Who Am I?

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  - Clinical specialties: hypermobility, fibromyalgia, headaches, temporomandibular disorders
- Member: Ehlers-Danlos Society Medical and Scientific Board
- Chair: The Allied Health Working Group of the International Consortium of Ehlers-Danlos Syndromes and Hypermobility Spectrum Disorders
- Frequent presenter to professional and patient groups at national and international conferences
- Author of multiple review and research articles on hypermobility
- Author: “Pain Mechanisms in HSD” in Di Bon, *The Integral Movement Method for Hypermobility Management*
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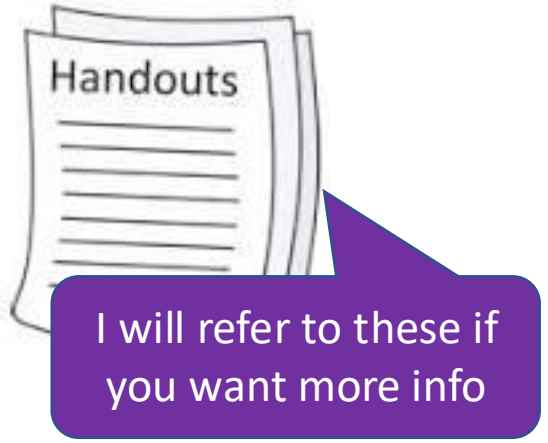
I do not have any conflicts of interest to report

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 104b: How to modify exercises so they work for you
- 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
- HSD 114: Hospitalization with HSD, POTS, MCAD
- HSD 115: Functional Neurological Disorder (FND)
- **HSD 116: Bleeding in HSD/hEDS**

Relevant Handouts Available



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

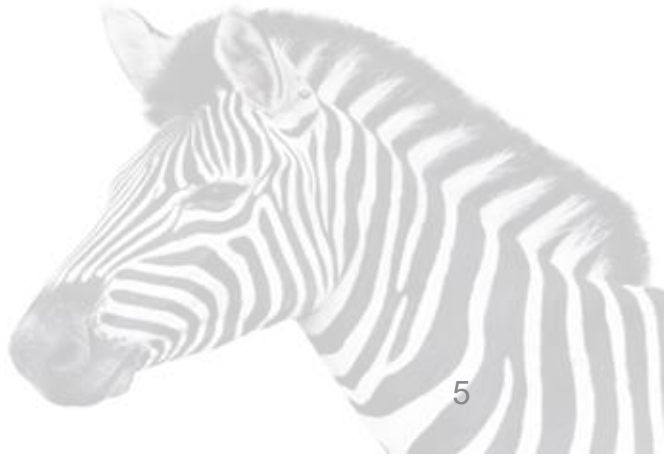
Nothing specifically on bleeding, but a few related topics:

- Surgical precautions for people with HSD/hEDS. Includes some info about bleeding. This is fairly technical and perhaps best for health care providers
- Chronic Pain Partners surgical planning booklet. Excellent patient information for EDS patients preparing for surgery
- **MCAS**
  - Suggestions for managing MCAS.
  - MCAS in the Emergency Room.

Disclaimers

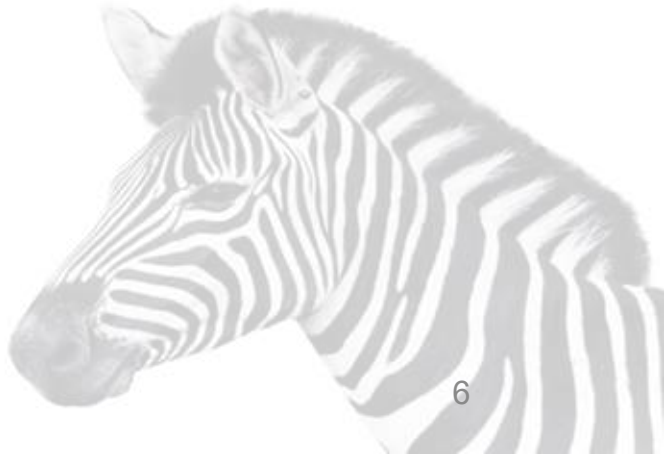
The information in this presentation is for general purposes, only, and may or may not apply to your situation.
This presentation cannot be used to diagnose or treat individuals.
This information is best used to start a discussion with your health care providers to determine if/how any of these concerns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

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



Alert

- This presentation will be discussing blood and bleeding, including menstrual bleeding.
- There are NO photographs of blood or bleeding.



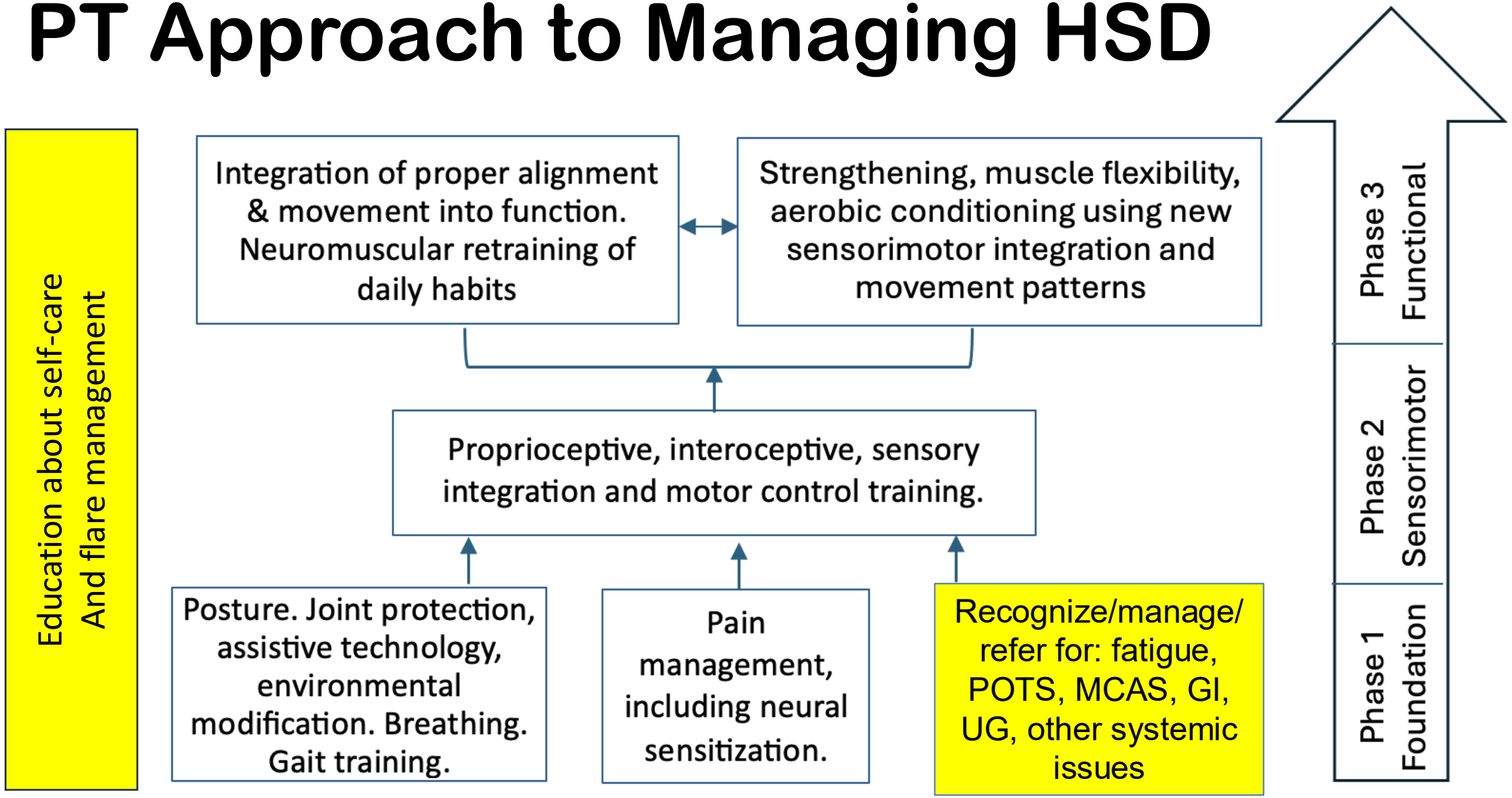
Objectives

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

1. Identify bleeding problems people with HSD/hEDS might have.
2. List several reasons for excessive bleeding in HSD/hEDS.
3. Assess their own bleeding tendencies using the Self-BAT
4. Initiate a discussion with health care providers about possible testing, precautions or treatment related to excessive bleeding.

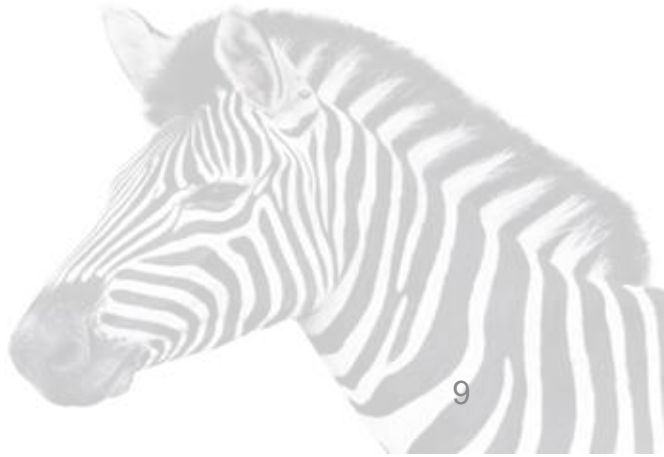
- HSD: Hypermobility Spectrum Disorders and hypermobile Ehlers-Danlos Syndrome
- Self-BAT: Self-administered Bleeding Assessment Tool

PT Approach to Managing HSD



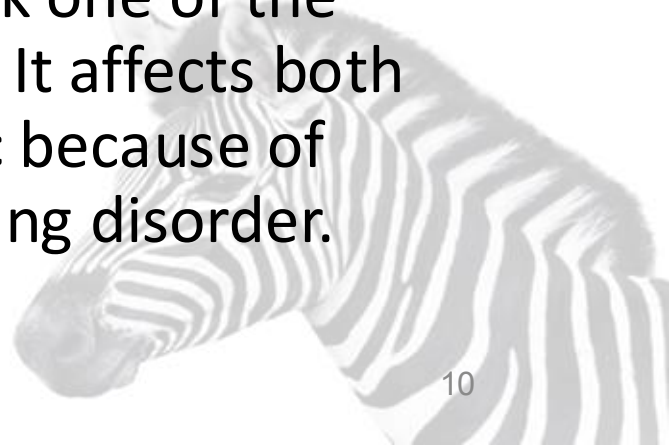
What is Excessive Bleeding?

- Bleeding may be due to injury, surgery, menstruation, dental work or spontaneous, such as a nose-bleed.
- If you bleed without a reason, bleed too much, or don't stop bleeding normally.
- If you are distressed by your bleeding.
- Only about a third of people with hEDS/HSD who have excessive bleeding get evaluated by a hematologist.



Genetic Bleeding Disorders

- Some bleeding disorders are genetic, others can develop in response to certain illnesses or as a reaction to some medications.
- Hemophilia
  - Hemophilia A is genetic, and has been assumed to affect only males, but women who are genetic 'carriers' may have a milder symptoms.
  - Hemophilia B is also genetic, but is often caused by a mutation, so the parents might not have had the abnormal gene. Again, women may have milder symptoms.
- Von Willebrand Disease (VWD) is also inherited. People lack one of the proteins needed for blood clots to form and stop bleeding. It affects both men and women, but women are often more symptomatic because of heavy menstrual bleeding. This is the most common bleeding disorder.

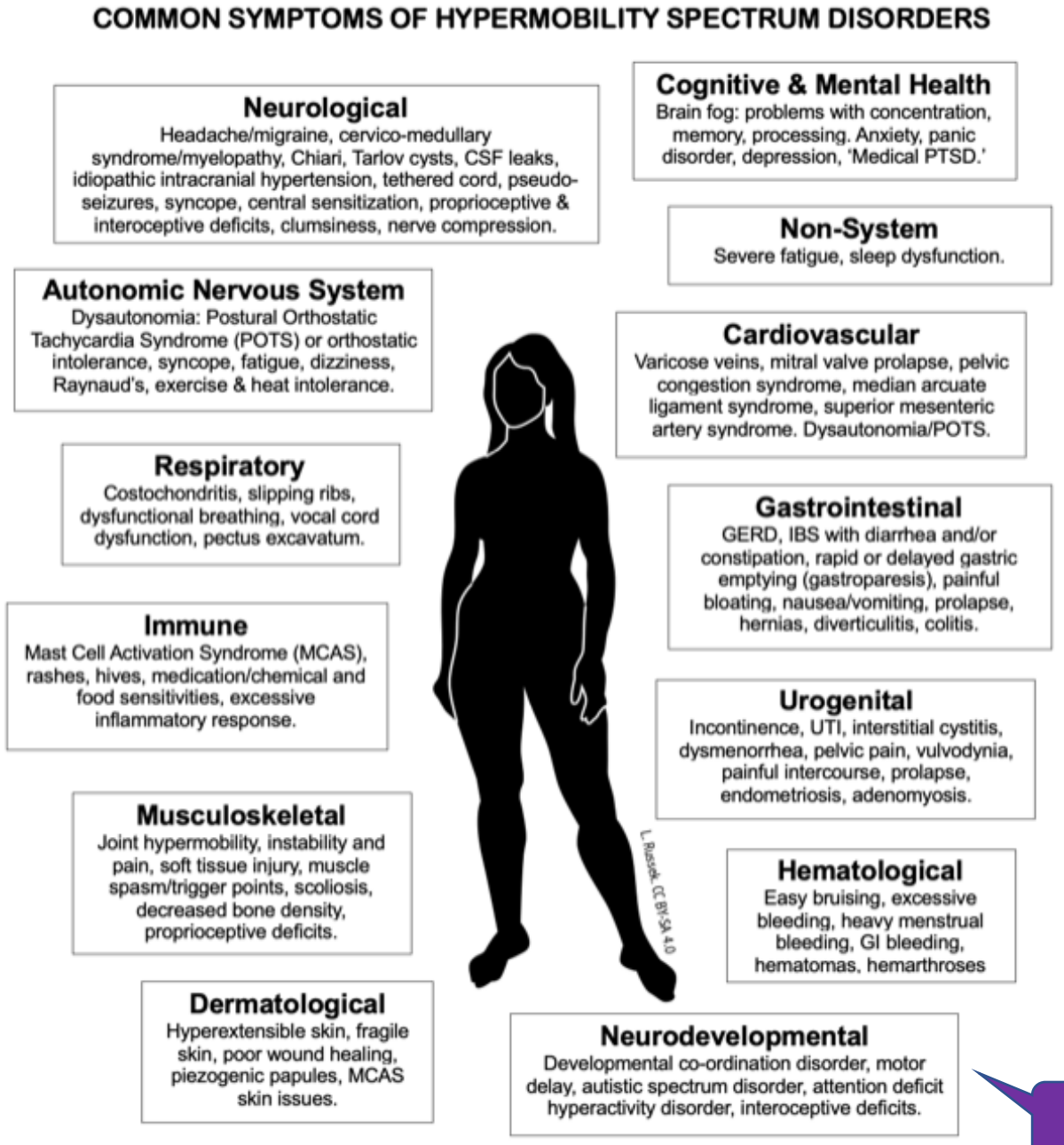


Other Reasons for Excessive Bleeding

- HSD/hEDS
- Blood thinner medications, such as warfarin, heparin, Eliquis, Zarelto, Pradaxa
- Medications that interfere with clotting, such as aspirin, NSAIDs, Plavix, sometimes SSRI/SNRI meds such as Prozac, Zoloft, Lexapro.
  - For list of meds & herbs: https://med.stanford.edu/content/dam/sm/ohns/documents/Sinus%20Center/Stanford_Medication_and_Herbs.pdf
- Hormonal abnormalities, uterine fibroids, polyps or some other uterine disorders.
- Nutritional deficiencies (e.g. severe vitamin K deficiency)
- Some organ diseases (e.g., liver or kidney) or bone marrow problems

How Common is Excessive Bleeding?

- Bleeding disorders are very common in HSD.
  - 62% of people with mixed EDS and 52% of those with hEDS have excessive bleeding (Kumskova, 2022)
  - 56.7% of people with HSD had platelet abnormalities. (Artoni, 2018)
  - In children with HSD, 75% had abnormal Bleeding Assessment Tool (BAT), but only 12.3% had been assessed for bleeding. (Kendel, 2023)
- HSD is very common in bleeding disorders, especially those with bleeding disorders of unknown cause (BDUC), i.e. not due to Von Willebrand or hemophilia.
  - 80% of female patients seen in a BDUC clinic are hypermobile. (Sanchez-Raya, 2018)
  - Only 55% of bleeding disorder specialists regularly screen for hypermobility in patients with BDUC. (Kelly, 2024)
- Providers in the bleeding disorders community recognize the prevalence of HSD and HSD has been identified as a BD research focus need (Sidonio, 2022)



Common HSD Problems

Bleeding issues:

- Easy bruising
- Heavy menstrual bleeding
- Excessive bleeding gums, nosebleeds
- Excessive post surgical bleeding
- Excessive bleeding during childbirth
- GI bleeding

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

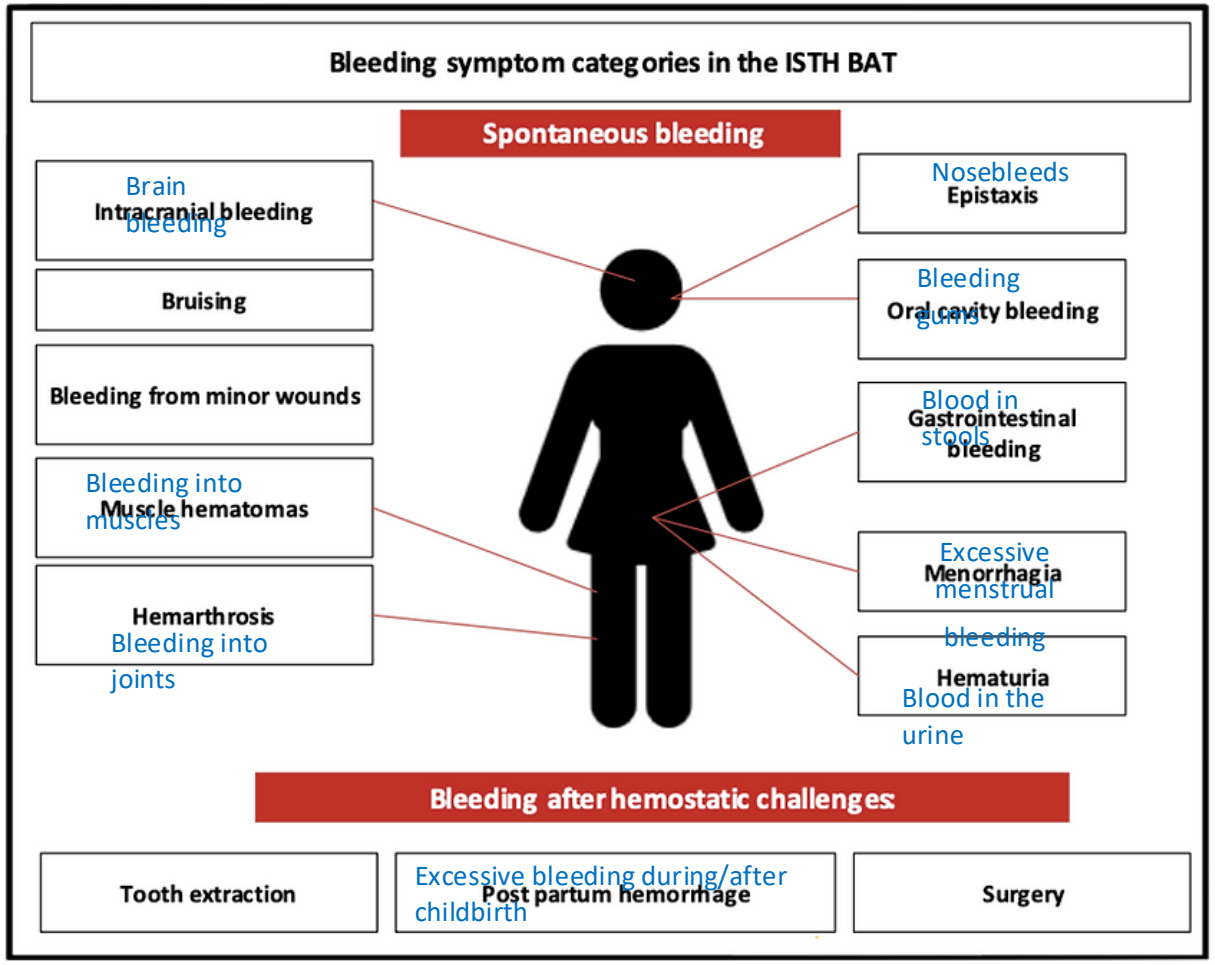
Women's Health issues:

- Pelvic floor pain/dysfunction
- Dyspareunia & vulvodynia
- Incontinence
- Pelvic organ prolapse
- Childbirth challenges

Pezaro, 2024; Boileau, 2024; Nazemi, 2023; Glayzer, 2021

Graphic of HSD

Do You Bleed Too Much?

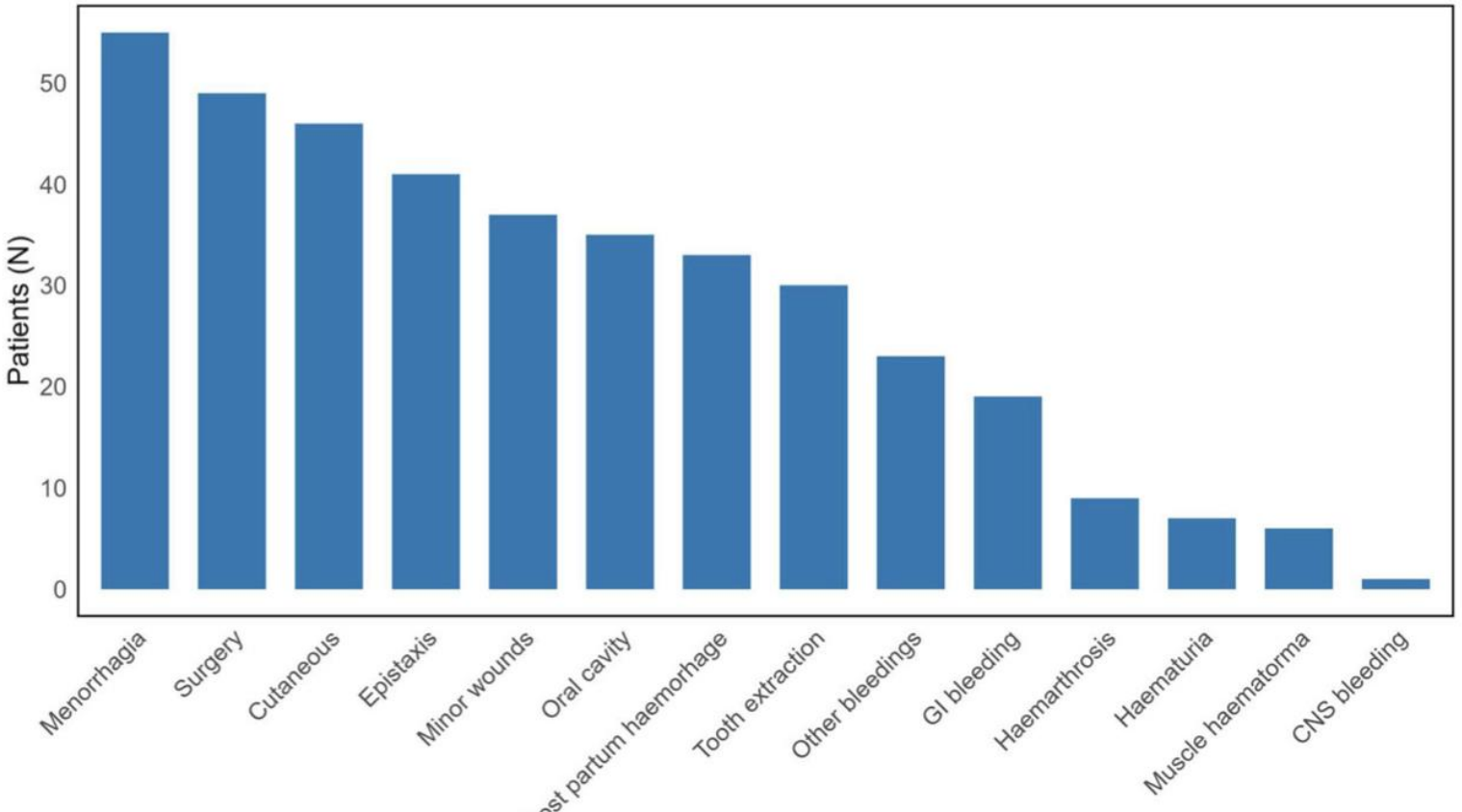


- ISTH-BAT (International Society on Thrombosis and Haemostasis Bleeding Assessment Tool) rates 14 types of bleeding. Intended to be completed by healthcare providers.
- The **Self-BAT** is a version that you can complete for yourself.
  - https://letstalkperiod.ca/self-bat/
  - Score >5 for women, >3 for men indicates possible bleeding disorder

Punt, 2019; DeYoung, 2026

https://pmc.ncbi.nlm.nih.gov/articles/PMC7354401/pdf/RTH2-4-761.pdf

Types of Bleeding in HSD



Leinøe, 2024

Menorrhagia = excessive menstrual bleeding
Cutaneous – from the skin
Epistaxis = nosebleeds
Hemarthrosis = bleeding into joints
Hematuria = blood in the urine
Hematoma = bleeding into a muscle
CNS bleeding = central nervous system

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

Heavy Menstrual Bleeding

- Heavy menstrual bleeding affects 76% of women with hEDS/HSD.
  - It is often the most distressing bleeding issue in hEDS/HSD.
- Severe menstrual bleeding can result in ER visits and hospitalization.
  - 14% had menstrual bleeding that was life-threatening or required surgery.
- Hysterectomies performed before EDS diagnosis in 78% of cases. This is about 50% more common than hysterectomies in VWD.
- Women with hEDS/HSD with heavy menstrual bleeding should be screened for hemostatic/platelet related bleeding disorders.

(Henderson, 2026; Cabrero Segurado, 2026; Jesudas, 2025; Kumskova, 2023)

Heavy Menstrual Bleeding (HMB or Menorrhagia)

SAMANTA Questionnaire	Score	
	Yes	No
1. Do you bleed for more than 7 days a month?	3	0
2. Do you have 3 or more days of heavier bleeding during menstruation?	1	0
3. In general, do you find your periods particularly uncomfortable because of their abundance?	3	0
4. On any of the heavier bleeding days do you stain your clothes at night; or would you stain them if you did not use double protection or change during the night?	1	0
5. During heavy bleeding days do you worry about staining the seat of your chair, sofa, etc.?	1	0
6. In general, on heavier bleeding days, do you avoid (as much as possible) certain activities, trips, or leisure plans because you have to change your tampon or pad frequently?	1	0

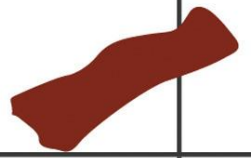
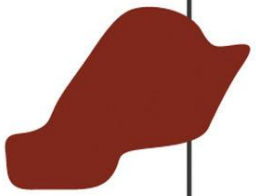
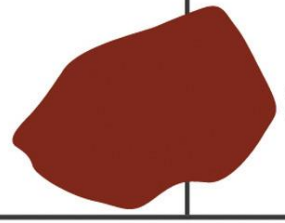
Total score: A value > 3 indicates that the woman may have heavy menstrual bleeding.

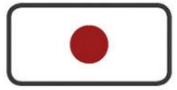
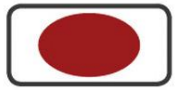
Perelló

- The 7-2-1 rule:
  - Periods lasting **7** or more days,
  - Changing pads/tampons every **2** hours or less
  - Passing blood clots larger than **1** inch (quarter-sized) or the size of a grape
- If you answer Yes to any of the above questions, you may have excessive bleeding

Pictorial Blood Loss Assessment Chart

Toilet	Score (ml of blood)
	1
	2
	3

Clots	Score (ml of blood)
	1
	2
	3

Sanitary towels	Type	Score (ml of blood)
	Brand	kotex
	At daytime	1
	At night	1
	At daytime	2
	At night	3
	At daytime	3
	At night	6
	At daytime	4
	At night	10
	At daytime	5
	At night	15

Tampon	Type	Score (ml of blood)
	Brand	Tampax
	Regular	0,5
	Super	1
	Super plus	1
	Regular	1
	Super	1,5
	Super Plus	2
	Regular	1,5
	Super	3
	Super Plus	6
	Regular	4
	Super	8
	Super Plus	12

Perelló, 2021

Bleeding Issues & HSD

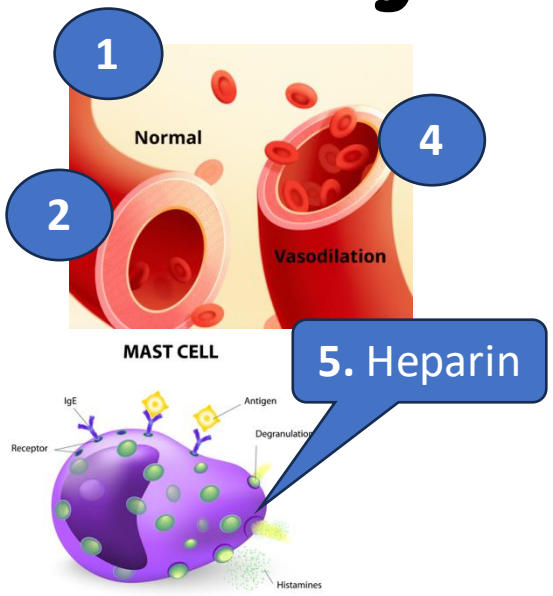
- People with HSD have high ISTH-BAT indicating bleeding disorders and high risk of hemorrhagic (excessive bleeding) complications.
- BAT scores were actually higher in hypermobile patients (in bleeding disorder clinic) than non-hypermobile. (Leinøe, 2024)
- HSD is more likely in people with bleeding disorders of unknown cause (BDUC), i.e. not due to Von Willebrand or hemophilia.
  - Sometimes patients with BDUC have trouble accessing care for bleeding.
- Excessive bruising in BDUC may be interpreted as domestic or child abuse.
- MCAS also increases bleeding due to leaky vessels and heparin release.

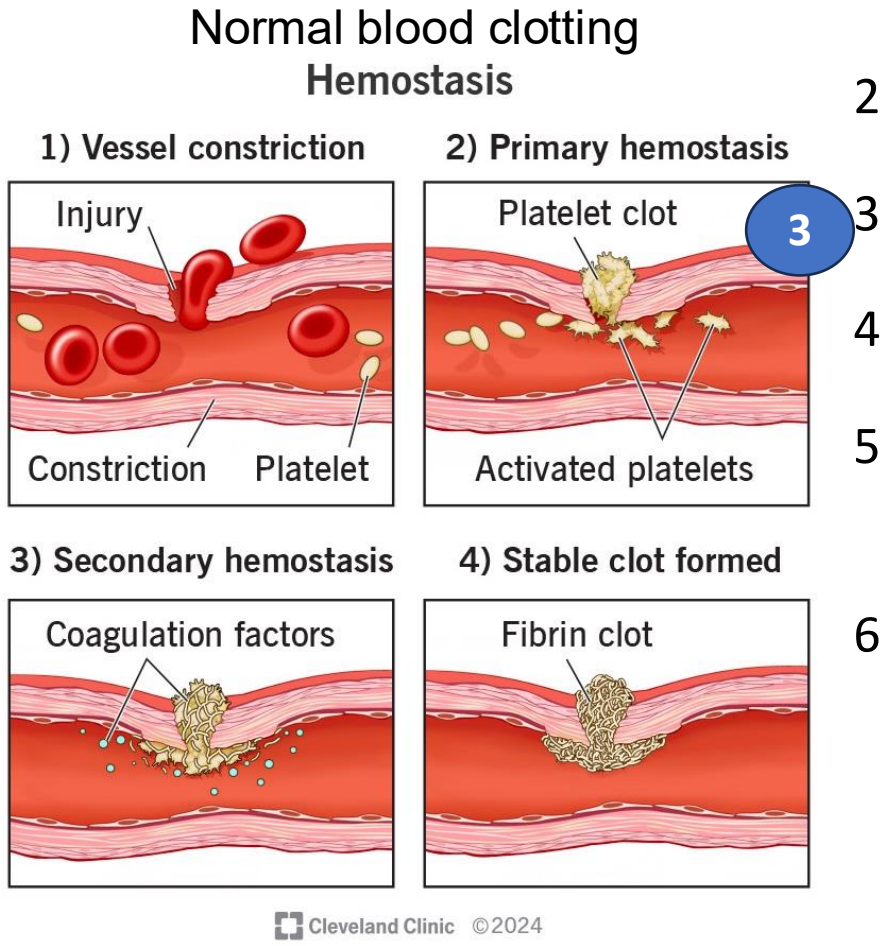
(Wiesmann, 2014; Kumskova, 2023; Wright, 2023; Kendel, 2023; Leinøe, 2024)

ISTH-BAT: International Society of Thrombosis and Haemostasis Bleeding Assessment Tool

MCAS: Mast Cell Activation Syndrome

Why Excessive Bleeding in HSD?

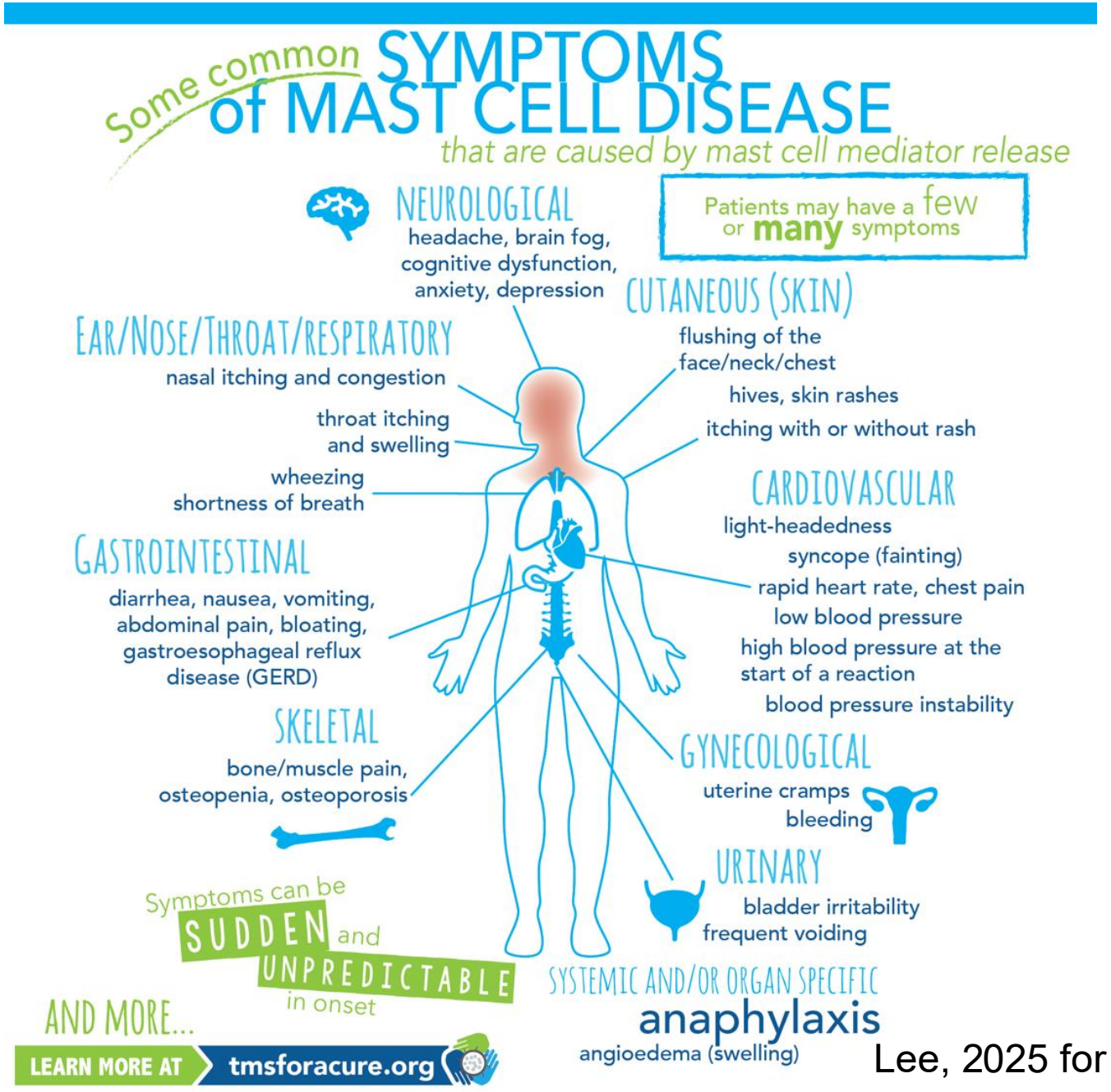




1. Connective tissue around vessels is less protective.
2. Connective tissue in blood vessel walls is fragile and damages more easily. (Artoni, 2018)
3. Abnormal platelet function interferes with clotting to stop bleeding. (Kumskova, 2026)
4. MCAS inflammation may also swell blood vessels (e.g., in uterus).
5. Release of heparin from MC increases bleeding.

(Afrin, 2021)

6. Medications may interfere with clotting.



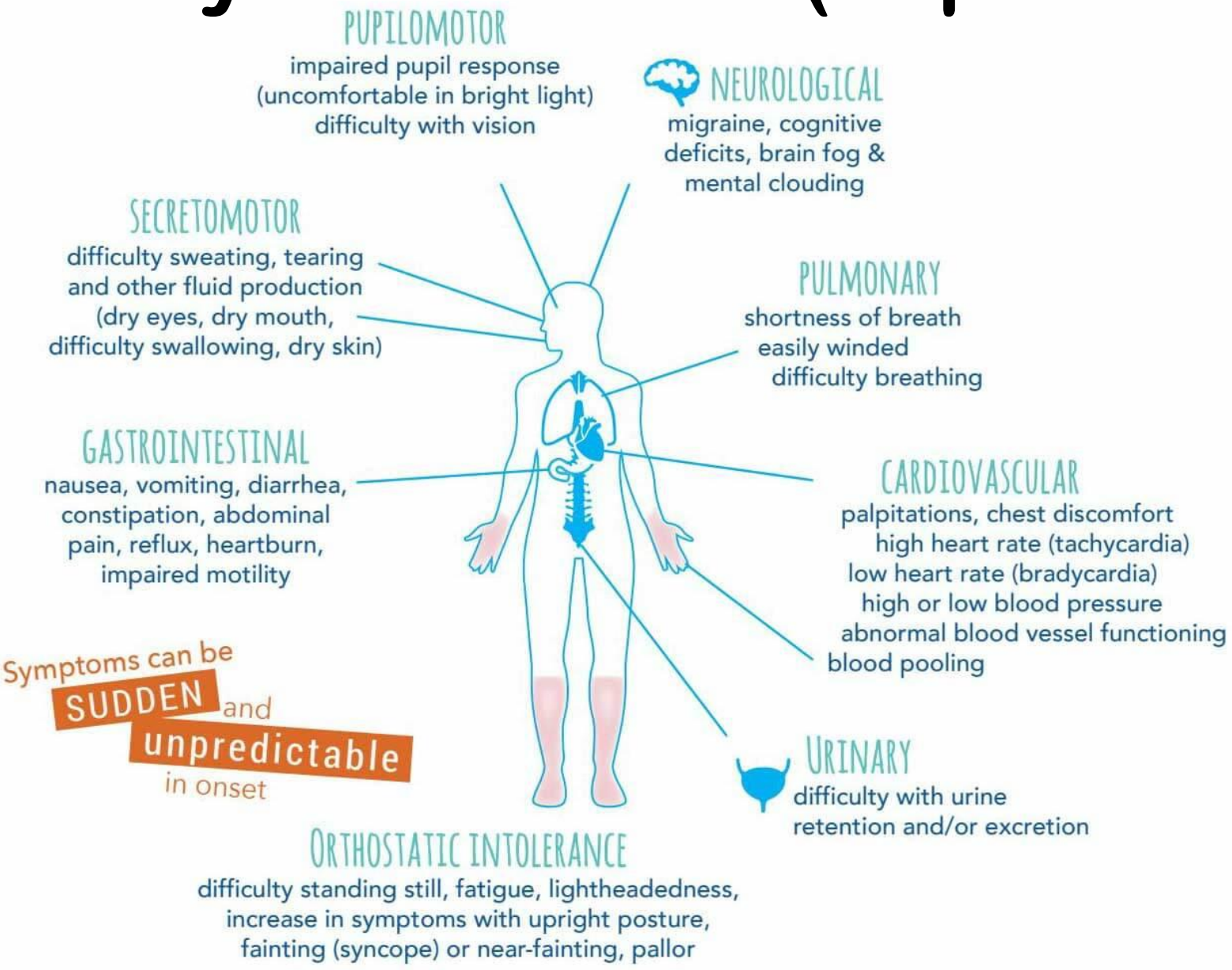
- Mast cells (MC) are non-specific immune cells that contribute to inflammation.
- MC are found in all tissues.
- When MC are ‘activated’ they lead to an excessive inflammatory response.
- MCAS can increase bleeding:
  - Causing swelling in tissues
  - Causing leaky vessels
  - Releasing heparin (blood thinner)

Lee, 2025 for overview of MCAS)

Dysautonomia (especially POTS)

About POTS

HSD 102: POTS & MCAD/MCAS



Also

- POTS is associated with a platelet disorder that contributes to easy bruising and heavy bleeding
- But menstrual bleeding no worse in POTS
- POTS symptoms may be worse during menstrual cycle

Boris, 2025; Gunning, 2022

Why Recognize Bleeding?

- To better recognize when bleeding is excessive and when excessive bleeding is problematic.
- Prevention during dental or surgical procedures, and better management of bleeding.
- Management of bleeding in people with HSD may be different:
  - Managing platelet dysfunction.
  - Managing MCAS, especially in heavy menstrual bleeding.

HMB Consequences

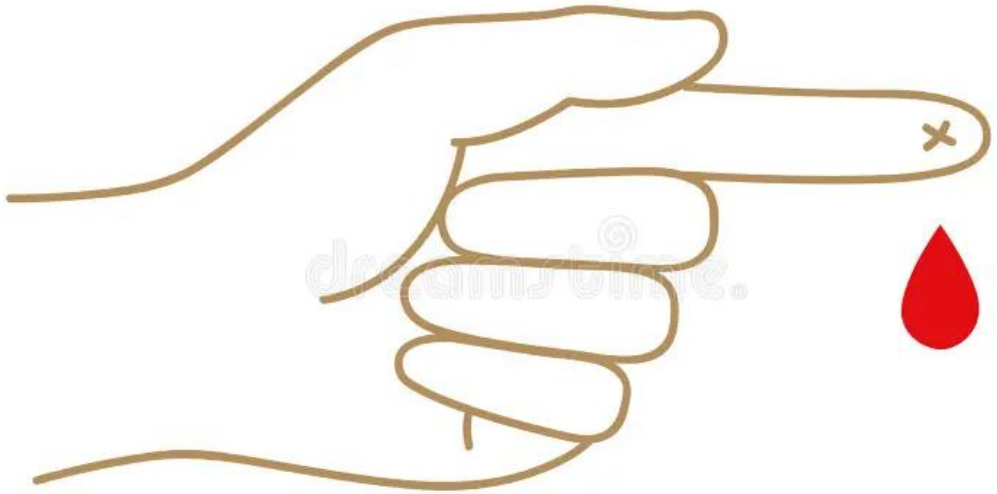
- Iron deficiency anemia.
- Physical discomfort.
- Emotional and social impacts – activities you avoid because of fear that you will leak or stain.
- Associated with lower quality of life, work, school, and sports performance.

(Perelló, 2021)

Testing for Excessive Bleeding

- You complete the Self-BAT (https://letstalkperiod.ca/self-bat/)
  - Score >5 for women, >3 for men indicates possible bleeding disorder
- Your provider completes the BAT.
- Blood test including Complete Blood Count (CBC) and blood smear to look at your red blood cells (RBC).
- Prothrombin Time measures blood clotting due to standard clotting factors.
- Activated Partial Prothromboplastin Time (aPTT) measures blood clotting ability due to different clotting factors.
- Clotting Factor Tests directly measure the amount of clotting factors.
- Platelet Function Assays look at how well platelets clump together.
  - Platelet Aggregation (gold standard) is a specific test to measure platelet clumping

Treatment Related to Bleeding in Patients with HSD



Who Treats Excessive Bleeding?

- For general bleeding issues, talk to your primary care provider.
- For heavy menstrual bleeding, talk to your gynecologist.
- For more severe bleeding, see a hematologist. They may do genetic testing for Von Willebrand Disease (VWD) and hemophilia.
  - While some people believe that only boys and men can get hemophilia, women can present with milder symptoms.

Prophylactic Treatment of Bleeding in HSD

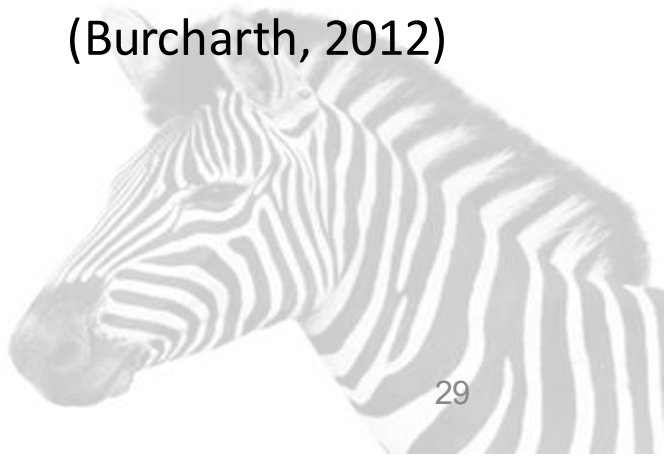
- Recognition that the person has a bleeding disorder.
- Avoidance of activities, such as contact sports, or conditions, such as high blood pressure. (Sidonia, 2023)
- Prophylactic care during surgery, tooth extraction. (Leinøe, 2024; Wiesmann, 2014)
- Avoid tourniquets, as these cause excessive bruising. (Wiesmann, 2014)
- Preparation for excessive bleeding during childbirth. (Pezaro, 2024)
- Vitamin C supplementation. (Leinøe, 2024)
  - (may benefit via impact on mast cells and MCAS)

HSD: Surgery Precautions

"Surgical Precautions"

HSD114: Hospitalization & Surgery

- All surgeries have higher risks of complications for HSD
  - Tissues are more fragile
  - Blood vessel fragility increases bleeding
- Providers should be prepared for excessive bleeding
- Tissue healing is delayed (Ericson, 2017)
- Special procedures for skin sutures: closer together, leave sutures in longer (Burcharth, 2012)

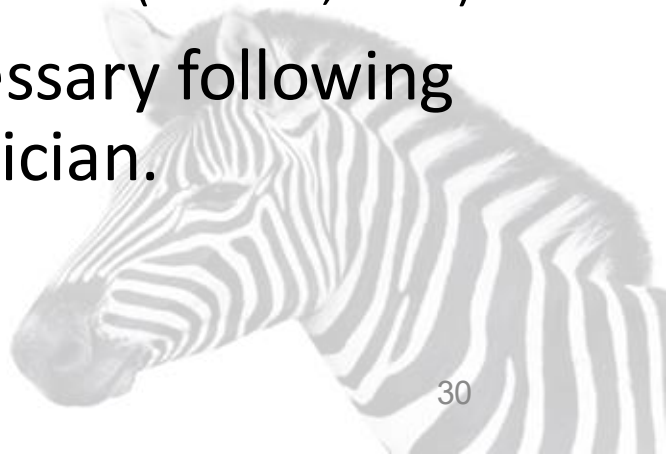


Use Blood Thinners Cautiously

- Medications that further compromise platelet function, such as non-steroidal anti-inflammatories (NSAIDs) and aspirin.
- Some herbal supplements may also increase bleeding:
  - Garlic, hawthorn, cordyceps sinensis, echinacea, and aloe vera (independent of anticoagulants).
  - Ginkgo biloba, chondroitin-glucosamine, melatonin, turmeric, bilberry, chamomile, fenugreek, milk thistle, and peppermint (if taking anticoagulants).

(Hatfield, 2022)

- Note that blood thinners may be appropriate and necessary following surgery, so discuss your bleeding issues with your physician.



Management of (General) Bleeding in HSD

- Prophylactics & treatments used (not necessarily evidence based)

Table 1 Prophylaxis and treatment strategies of respondents for typical patients with significant bleeding history and patients with generalized joint hypermobility

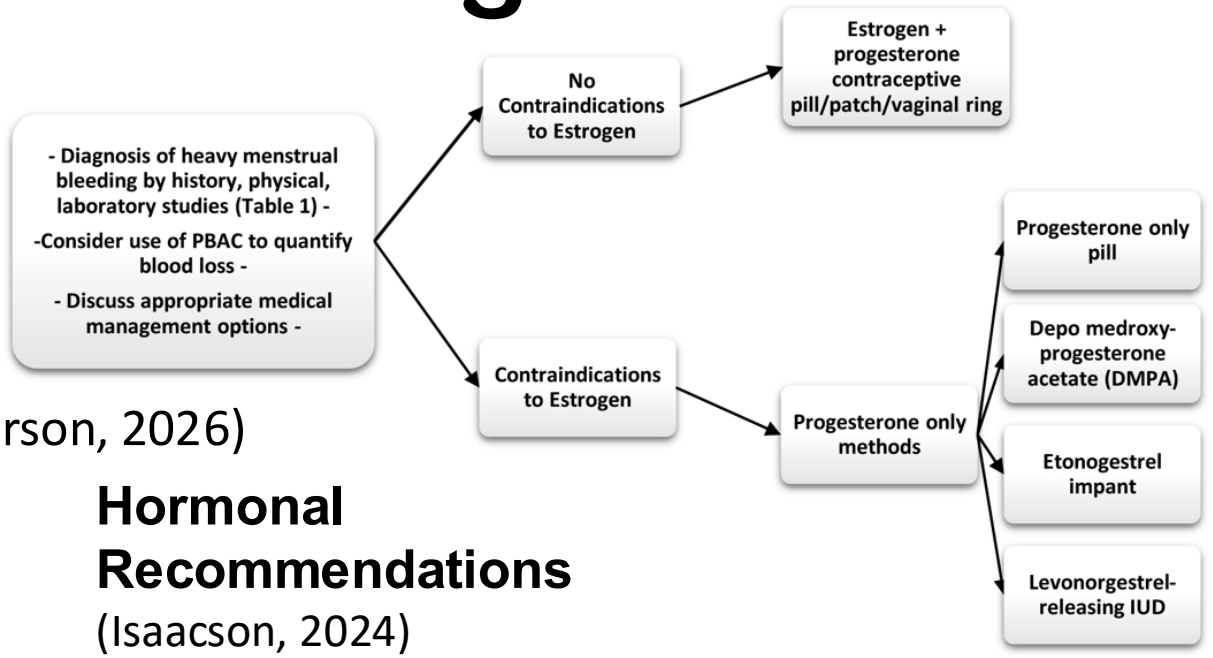
Medication or product	Typical prophylaxis, *n* (%)	Typical treatment, *n* (%)	Hypermobile prophylaxis, *n* (%)	Hypermobile treatment, *n* (%)
Desmopressin acetate	6 (18%)	6 (18%)	7 (21%)	9 (27%)
Antifibrinolytics	10 (30%)	13 (39%)	11 (33%)	14 (42%)
Platelets	3 (9%)	2 (6%)	2 (6%)	2 (6%)
Plasma	1 (3%)	1 (3%)	0 (0%)	0 (0%)

(Kendel, 2021)

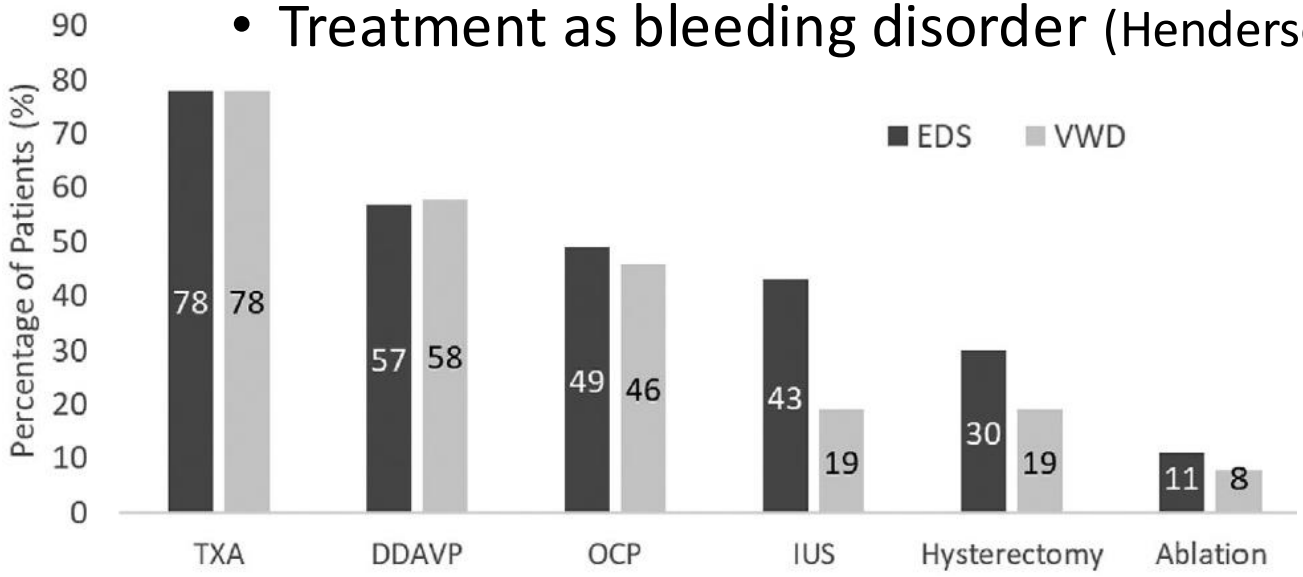
- Desmopressin is recommended during surgery (Wiesmann, 2014)

Rx Heavy Menstrual Bleeding in HSD

- Heavy menstrual bleeding
  - Management of MCAS (Afrin, 2021)
  - Hormonal treatment (Isaacson, 2024)
  - Treatment as bleeding disorder (Henderson, 2026)



Hormonal Recommendations
(Isaacson, 2024)



Comparing EDS (type unknown) and VWD. DDAVP, desmopressin; IUS, intrauterine system; OCP, oral contraceptive pills; TXA, tranexamic acid.

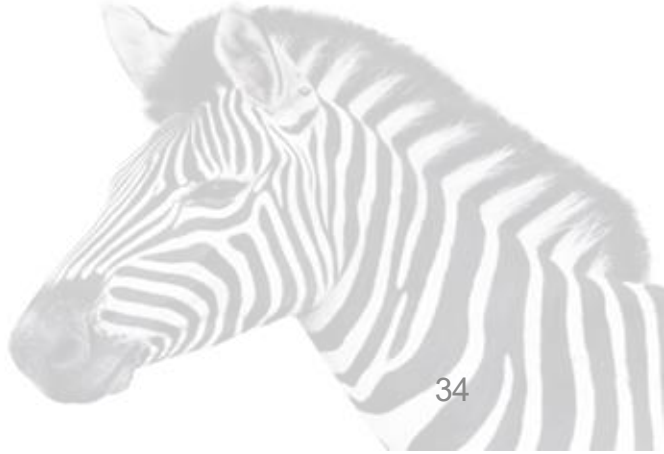
Current practice
(Henderson, 2026)

Difficulty Having Blood Drawn/IV Set

- Many people with EDS and POTS report difficulty when medical providers draw blood or set IV lines.
  - Trouble finding veins.
  - Rolling veins, getting needles into the veins.
  - Severe bruising afterwards.
- This is worse if you are dehydrated because you have been told to stop drinking fluids for a procedure
  - Confirm when you really need to stop drinking water beforehand.
  - If allowed, drink plenty of fluids immediately before the draw.
  - Fluids are better absorbed with electrolytes or a little bit of sugar.
- Warm the arm beforehand.
- Ask for a smaller needle (a 'butterfly needle').
- Ask for the most skilled person.
- Apply compression and elevate the arm afterwards.

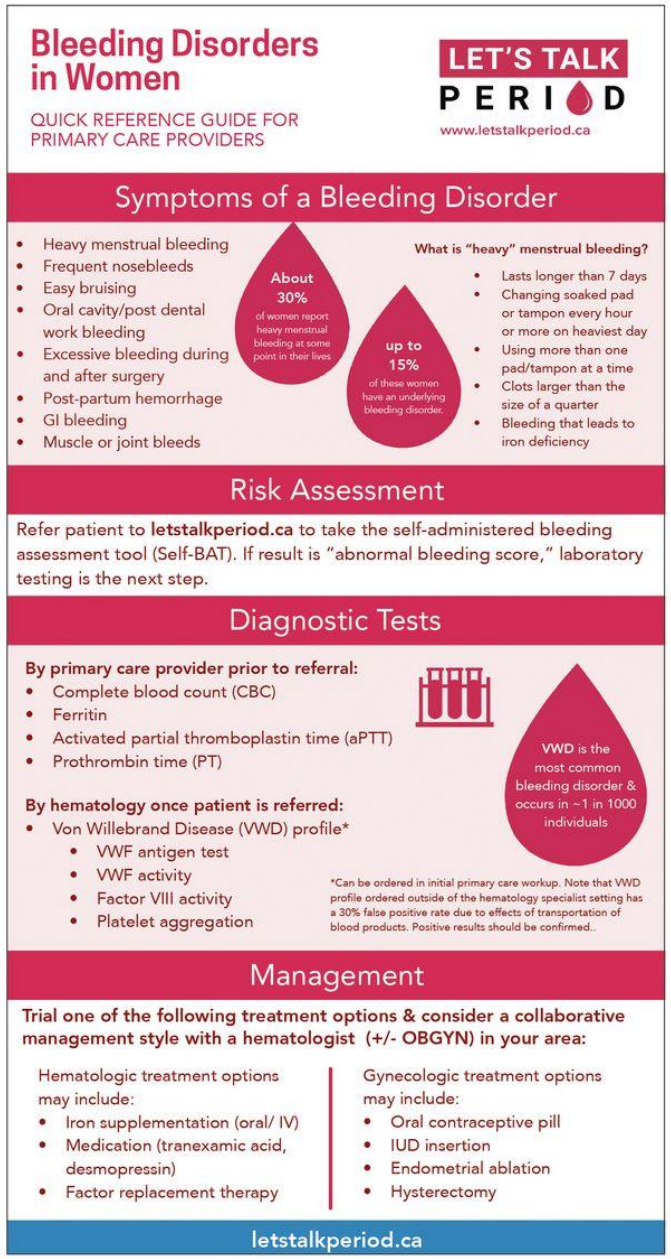
Summary

- Excessive bleeding is common in patients with HSD
  - It is often not recognized or not considered to need treatment
- Research into bleeding tendencies in people with HSD is just beginning, so optimal treatment approaches are not yet known.
  - Preventing excessive bleeding is a good starting point.
  - Medications may be helpful, both meds already used for bleeding and hormones (for heavy menstrual bleeding).
  - Managing MCAS, if present, may also decrease bleeding



MD Management of Excessive Bleeding

Figure 1. Bleeding disorders reference guide for primary care providers



Bleeding Disorders in Women

QUICK REFERENCE GUIDE FOR PRIMARY CARE PROVIDERS

LET'S TALK PERIOD
www.letstalkperiod.ca

Symptoms of a Bleeding Disorder

- Heavy menstrual bleeding
- Frequent nosebleeds
- Easy bruising
- Oral cavity/post dental work bleeding
- Excessive bleeding during and after surgery
- Post-partum hemorrhage
- GI bleeding
- Muscle or joint bleeds

About 30% of women report heavy menstrual bleeding at some point in their lives

up to 15% of these women have an underlying bleeding disorder.

What is "heavy" menstrual bleeding?

- Lasts longer than 7 days
- Changing soaked pad or tampon every hour or more on heaviest day
- Using more than one pad/tampon at a time
- Clots larger than the size of a quarter
- Bleeding that leads to iron deficiency

Risk Assessment

Refer patient to **letstalkperiod.ca** to take the self-administered bleeding assessment tool (Self-BAT). If result is "abnormal bleeding score," laboratory testing is the next step.

Diagnostic Tests

By primary care provider prior to referral:

- Complete blood count (CBC)
- Ferritin
- Activated partial thromboplastin time (aPTT)
- Prothrombin time (PT)

By hematology once patient is referred:

- Von Willebrand Disease (VWD) profile*
  - VWF antigen test
  - VWF activity
  - Factor VIII activity
  - Platelet aggregation

VWD is the most common bleeding disorder & occurs in ~1 in 1000 individuals

*Can be ordered in initial primary care workup. Note that VWD profile ordered outside of the hematology specialist setting has a 30% false positive rate due to effects of transportation of blood products. Positive results should be confirmed..

Management

Trial one of the following treatment options & consider a collaborative management style with a hematologist (+/- OBGYN) in your area:

Hematologic treatment options may include:

- Iron supplementation (oral/ IV)
- Medication (tranexamic acid, desmopressin)
- Factor replacement therapy

Gynecologic treatment options may include:

- Oral contraceptive pill
- IUD insertion
- Endometrial ablation
- Hysterectomy

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IUD—intrauterine device, IV—intravenous, GI—gastrointestinal, OBGYN—obstetrician-gynecologist, VWF—von Willebrand factor.
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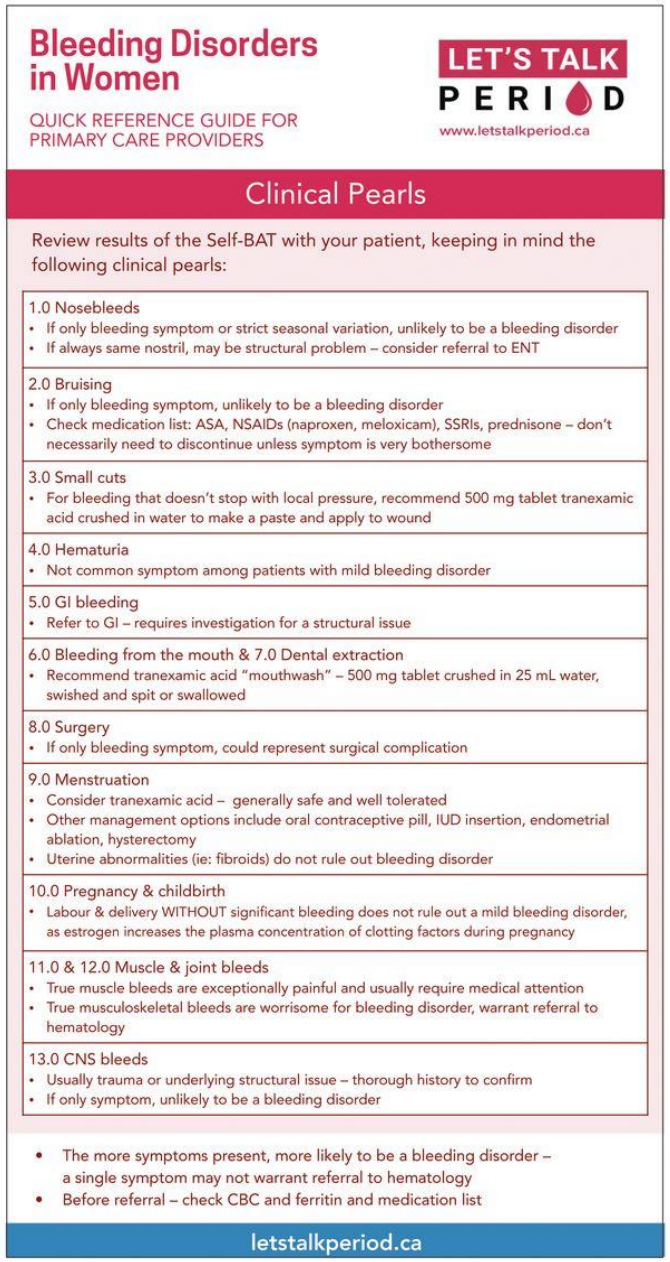
Figure 2. Bleeding disorders clinical pearls



Bleeding Disorders in Women

QUICK REFERENCE GUIDE FOR PRIMARY CARE PROVIDERS

LET'S TALK PERIOD
www.letstalkperiod.ca

Clinical Pearls

Review results of the Self-BAT with your patient, keeping in mind the following clinical pearls:

1.0 Nosebleeds
- If only bleeding symptom or strict seasonal variation, unlikely to be a bleeding disorder
- If always same nostril, may be structural problem – consider referral to ENT

2.0 Bruising
- If only bleeding symptom, unlikely to be a bleeding disorder
- Check medication list: ASA, NSAIDs (naproxen, meloxicam), SSRIs, prednisone – don't necessarily need to discontinue unless symptom is very bothersome

3.0 Small cuts
- For bleeding that doesn't stop with local pressure, recommend 500 mg tablet tranexamic acid crushed in water to make a paste and apply to wound

4.0 Hematuria
- Not common symptom among patients with mild bleeding disorder

5.0 GI bleeding
- Refer to GI – requires investigation for a structural issue

6.0 Bleeding from the mouth & 7.0 Dental extraction
- Recommend tranexamic acid "mouthwash" – 500 mg tablet crushed in 25 mL water, swished and spit or swallowed

8.0 Surgery
- If only bleeding symptom, could represent surgical complication

9.0 Menstruation
- Consider tranexamic acid – generally safe and well tolerated
- Other management options include oral contraceptive pill, IUD insertion, endometrial ablation, hysterectomy
- Uterine abnormalities (ie: fibroids) do not rule out bleeding disorder

10.0 Pregnancy & childbirth
- Labour & delivery WITHOUT significant bleeding does not rule out a mild bleeding disorder, as estrogen increases the plasma concentration of clotting factors during pregnancy

11.0 & 12.0 Muscle & joint bleeds
- True muscle bleeds are exceptionally painful and usually require medical attention
- True musculoskeletal bleeds are worrisome for bleeding disorder, warrant referral to hematology

13.0 CNS bleeds
- Usually trauma or underlying structural issue – thorough history to confirm
- If only symptom, unlikely to be a bleeding disorder

- The more symptoms present, more likely to be a bleeding disorder – a single symptom may not warrant referral to hematology
- Before referral – check CBC and ferritin and medication list

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ASA—acetylsalicylic acid; CBC—complete blood count; CNS—central nervous system; ENT—ears, nose, throat; GI—gastrointestinal; IUD—intrauterine device; NSAID—nonsteroidal anti-inflammatory drug; Self-BAT—self-administered bleeding assessment tool; SSRI—selective serotonin reuptake inhibitor.
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Yeung, 2022

Patient Resources For HSD/hEDS

- "Meeting Your EDS Hospital Stay Needs" (Chronic Pain Partners) – planning for surgery: https://www.chronicpainpartners.com/wp-content/uploads/2023/02/surgery-prep-meeting-your-eds-hospital-stay-needs.pdf
- Surgical prep guide for patients: https://hypermobilityclinic.org/surgical-and-anesthetic-precautions-for-hypermobile-and-eds-patients/
- Leslie's surgical precautions handout (more for providers than patients): https://webspace.clarkson.edu/~lrussek/docs/hypermobility/SurgeryHSD.pdf
- Dental surgery guide to EDS: https://www.rareconnect.org/uploads/documents/eds-dentistry-medical-resource-guide.pdf
- Web site with info to share with your doctors: https://hypermobilityclinic.org/surgical-and-anesthetic-precautions-for-hypermobile-and-eds-patients/

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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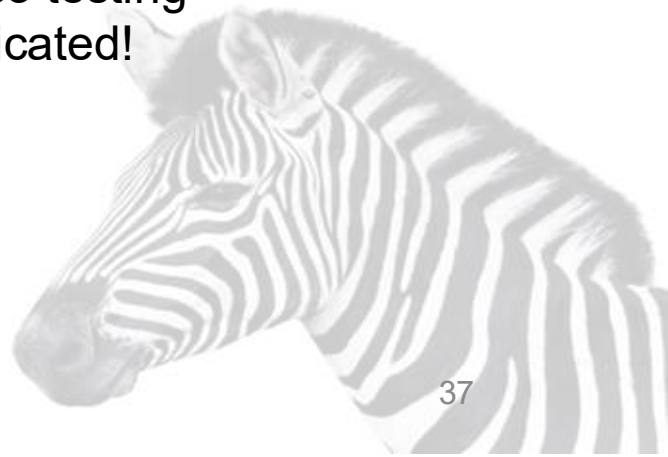
This document is online at *www.AlanSpanosMD.com*. It was updated March 2019.
For more information, see the Ehlers Danlos Society at ehlers-danlos.com.

Handout for patients to share with surgeons and anesthesiologists

This handout and many others to share with your health care providers by Dr. Spanos, MD (retired)
https://rachelleepac.com/spanos-resources/

Also included in my Surgical Precautions handout

But we now know there CAN be clotting disorders, so testing may be indicated!



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

Surgical Resources: POTS & MCAS

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  - https://www.autonomicneuroscience.com/article/S1566-0702(18)30032-8/fulltext
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  - https://tmsforacure.org/perioperative-management/

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Thank You!

Questions?