PRACTICE | FIVE THINGS TO KNOW ABOUT ...

Mild bleeding disorders in adults

Dongmei Sun MD MSc, Chai W. Phua MD

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Patients in primary care frequently report mild bleeding that may warrant further investigation

About 11% of patients in primary care reported bleeding symptoms in a 2010 Canadian study.¹ Mild bleeding disorders should be considered in patients with disproportionate bleeding (i.e., excessive postsurgical bleeding from multiple sites or bleeding that requires blood or iron transfusion).² Mild bleeding disorders can be inherited or acquired; common diagnoses include mild von Willebrand disease, platelet dysfunction and mild-moderate factor deficiencies.² Unlike severe inherited bleeding disorders that are often diagnosed in early life, mild inherited bleeding disorders can present in adulthood.

2 Identifying someone with mild bleeding requires a systematic approach

First steps include a detailed medical history emphasizing bleeding and family history, a physical examination to rule out secondary conditions (e.g., Cushing syndrome or Ehlers–Danlos syndrome) (Appendix 1, available at www.cmaj.ca/ lookup/doi/10.1503/cmaj.201182/tab-related-content) and laboratory-screening tests listed in point 4.² Recognition of an inheritance pattern could help narrow the differential diagnosis (e.g., X-linked recessive pattern in hemophilia A).

3 An assessment tool may be helpful in differentiating clinically relevant from inconsequential bleeding

The ISTH-SSC Bleeding Assessment Tool (ISTH/SSC-BAT) may be used to differentiate the 2 types of bleeding. The tool comprises 14 distinct bleeding manifestations and takes about 20 minutes for the patient to complete (www.isth.org/page/reference_tools).³ A positive score of 4 or more in males and 6 or more in females justifies referral to a specialist. However, the tool may fail to identify people with acquired bleeding disorders and is less sensitive in younger patients who have never undergone surgery.

Initial laboratory screening tests inform decisions about further testing

Initial laboratory screening tests should include complete blood cell count, prothrombin time/international normalized ratio, activated partial thromboplastin time, fibrinogen level and a blood smear. If the ISTH/SSC-BAT score is negative and results for the laboratory screening tests are unremarkable, ongoing clinical observation is sufficient without further testing.⁴ Conversely, a positive ISTH/SSC-BAT score with normal results for laboratory tests will require referral for further evaluation.

An acquired bleeding disorder may develop later in life

Mildly disordered bleeding that arises later in life may indicate the presence of an acquired bleeding disorder, and the patient should be evaluated for underlying systemic disease. The diverse causes for an acquired bleeding disorder are listed in Appendix 1. A structured approach is necessary to ensure pathologies, such as malignant disease associated with acquired bleeding disorders, are ruled out (e.g., acquired hemophilia A, immunoglobulin M hyperviscosity syndrome and light-chain amyloidosis).^{5,6}

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Affiliation: Department of Medicine, Schulich School of Medicine and Dentistry, Western University, London, Ont.

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Correspondence to: Dongmei Sun, Dongmei.Sun@lhsc.on.ca

Bleeding Disorders in Women

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Symptoms of a Bleeding Disorder



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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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