

Bleeding disorders such as von Willebrand disease and rare factor deficiencies result when the patient's blood lacks certain clotting factors. Once diagnosed, if the disorders are not treated properly, it can cause those affected to experience prolonged bleeding after injury, surgery, or physical trauma. It also has the potential to damage organs, tissue and may be life-threatening. Below is a set of recommendations for clinical evaluation and care.



These recommendations are not a substitute for reasonable clinical judgment and decision making and do not exclude other options. Clinical care must be individualized to the specific needs of each patient and interventions must be tailored accordingly.

DISORDER	SIGNS/SYMPTOMS	WHEN TO REFER	COMMENTS
Bleeding disorder	<p>Common symptoms of a bleeding disorder include excessive bleeding when brushing teeth, prolonged bleeding after cut/scrape, bleeding after dental work/extractions and epistaxis</p> <ul style="list-style-type: none"> • In females, menorrhagia is often the first sign • Epistaxis can be due to bleeding disorder – but more commonly due to allergies, colds, picking nose, etc. 	<ul style="list-style-type: none"> • Significant anemia due to menstrual blood loss – hemoglobin <9 • Failed improvement in anemia with oral iron <ul style="list-style-type: none"> - May be candidate for IV iron replacement • Especially in teenagers – common to fail oral iron due to non-compliance • Severe anemia at onset of menses – especially if requiring transfusion <ul style="list-style-type: none"> - May be first sign of a bleeding disorder 	<p>Consider bleeding disorder if any of the following are positive*</p> <ul style="list-style-type: none"> • Duration of menses >7 days and either “flooding” or impairment of daily activities with most periods • A history of treatment of anemia • Family history of a diagnosed bleeding disorder • History of excessive bleeding with tooth extraction, delivery, miscarriage or surgery <p><i>*(Philipp C.S. et al, Am J Ob Gyn 198(2):1-8, 2007)</i></p>

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Case study:

At the start of menses, a 10-year-old girl needed to change pads every 1-2 hours for the first 3-4 days. Her cycle finished around the sixth day. At the start of her second menstrual cycle, she passed multiple clots, and was still bleeding on the 12th day. She was taken to her primary care physician and a CBC demonstrated Hgb 10.1 and platelets 114k. Three days later, she was still bleeding and developed new symptoms of fatigue and lightheadedness. She was taken to the emergency room. Repeat CBC showed a Hgb of 7.1 and platelets 166k. PT and PTT were normal. She was prescribed ferrous sulfate and Ortho-Cyclen (1 tab PO BID x5 days, then 1 tab PO daily). At the time of her hematology consultation three days later, her menses had stopped.

Bleeding history:

- No gingival bleeding, bleeding with cuts/scratches. No epistaxis.
- Post-op hemorrhage after clot came off following T&A – requiring cautery
- No bleeding with surgery (ORIF) for arm fracture at 6 years old

This patient had several red flags for a bleeding disorder; mucosal bleeding, heavy menstrual bleeding with clots, and mucosal bleeding history. Her mucosal bleeding was most suspicious for possible von Willebrand disease or a platelet function defect. Despite anemia and findings of iron deficiency, platelets were lower than expected (110's – 160's). Between 20 – 30% of adolescents with heavy menstrual bleeding have a bleeding disorder.

Diagnostic work-up:

- Normal von Willebrand panel, including multimers
- Platelet aggregation studies revealed an aspirin-like platelet function defect

Treatment:

- Combined hormonal contraception successfully stopped her bleeding, but family requested stopping due to her young age.
- She started tranexamic acid (1300 mg PO TID x5 days starting at Day 1 of each menstrual cycle) with good response.

Cook Children's has a recommended collaborative approach when caring for patients with bleeding disorders:

- OB/GYN
 - Management of hormonal therapy
- Hematology
 - Management of anemia, iron replacement
 - Work-up/evaluation for bleeding disorder Best done after anemia has resolved