



WORLD FEDERATION OF HEMOPHILIA
FÉDÉRATION MONDIALE DE L'HÉMOFILIE
FEDERACIÓN MUNDIAL DE HEMOFILIA

FREQUENTLY ASKED QUESTIONS ON WOMEN AND GIRLS WITH BLEEDING DISORDERS

Q. What is an inherited bleeding disorder?

Injury and bleeding result from damage to blood vessels, from which blood leaks out. Your body's natural response is to try to limit this blood loss by stopping the flow out of the vessel and then later repairing the damage. The first response to blood loss is that your body will try to seal the gap at the bleeding point by making a blood clot at that point. This clotting process uses a combination of blood clotting proteins called clotting factors, and blood cells called platelets that are naturally present in the body. The first factor to arrive to sites of injury is von Willebrand Factor (VWF); it binds to the lining of the damaged blood vessel and attracts platelets to the area to help the blood clot to form. More platelets and other clotting factors are then attracted to the site, making the clot stronger and halting the bleeding. A clot therefore occurs when blood is converted from a liquid form to a solid state (and can no longer flow) and further blood loss is prevented.

An inherited bleeding disorder is a condition where either the platelets are abnormal and non-sticky, or clotting factors are decreased, abnormal or absent. This makes it difficult for a person to stop bleeding and they continue to lose blood.

Q. Who is affected by an inherited bleeding disorder? What are the types of inherited bleeding disorders?

An inherited bleeding disorder is passed down from parent to child and can manifest at any age in men, women, boys, and girls. It can also be a spontaneous mutation, meaning there is no family history. Because of monthly periods or pregnancy, women and girls are more likely to experience heavy menstrual bleeding (HMB), bleeding during pregnancy and childbirth, and bleeding following childbirth (also called post-partum hemorrhage).

Approximately 1 in 10 women with heavy periods have an underlying bleeding disorder. However, heavy periods can also occur due to other gynecological conditions such as uterine fibroids, polyps, endometriosis, or female hormonal imbalance. Non-gynecological causes of heavy periods include thyroid problems, systemic diseases such as severe liver or kidney diseases, or non-inherited (also called acquired) abnormalities of blood coagulation or reduced platelet count.

The types of inherited bleeding disorders include 1) platelet function disorders where the platelets are non-sticky and do not clump (called platelet function defects), and 2) clotting factor deficiencies. The most common inherited bleeding disorders are von Willebrand Disease (VWD), and deficiencies of factor 8 and factor 9 (called hemophilia A and hemophilia B, respectively). Rare bleeding disorders include deficiencies of other factors (factors 1, 2, 5, 5+8, 7, 10, 11, 13, congenital deficiency of vitamin K-dependent factors, plasminogen etc.)

Q. What are the symptoms and signs of bleeding disorders in women and girls?

- Heavy bleeding during menstruation (menstrual period) that can include:
 - Bleeding that lasts longer than 7 days from the time bleeding starts until the time it ends.
 - Flooding or gushing of blood that limits daily activities, such as work, school, exercise, or social activities.
 - Passing clots that are bigger than a grape.
 - Soaking a tampon or pad every hour or more often on the heaviest day(s).
- A diagnosis of “low in iron” or having received treatment for anemia. Anemia can make you look pale and feel tired or weak.
- Symptoms of easy or frequent bleeding may include:
 - Nosebleeds that occur for no apparent reason and last longer than 10 minutes, or that need medical attention.
 - Easy bruising that occurs with no physical injury.
 - Excessive bleeding after a medical procedure or dental procedure.
 - A history of muscle or joint bleeding with no physical injury.

If you have one or more of the bleeding symptoms listed above and a family member with a bleeding disorder (such as von Willebrand disease, hemophilia, or clotting factor deficiencies), you should arrange for laboratory testing to determine whether you have a bleeding disorder. If you have heavy periods as well as other bleeding symptoms or needed iron, admission to hospital, or a blood transfusion in the past, you should also arrange for testing.

Q. What are bleeding symptoms that are distinctive to women and adolescent girls with bleeding disorders?

- Heavy menstrual bleeding as described above.
- After menarche (your first period) or as you reach menopause (your last period), all women may experience heavier or irregular periods; this may be more marked in women and girls with bleeding disorders.
- Pain in the middle of your menstrual cycle (also called mittelschmerz, a German word that means “middle pain”). This pain occurs secondary to bleeding from the ovary at the time of egg release into the peritoneal cavity (the space within the abdomen that contains the abdominal and pelvic organs) which irritates the peritoneum (the membrane that connects and supports the internal organs in the pelvis and abdomen). Please note that this bleeding is not external or visible. In women with severe bleeding disorders, the bleeding may be severe and cause a state of shock or sudden severe pain (called acute abdomen) and require hospitalization. This is rare, but can be life threatening and require urgent medical attention.
- Vaginal bleeding during sex, unscheduled bleeding while taking hormonal therapy, and postmenopausal bleeding.
- Women with bleeding disorders are more likely to suffer symptoms of bleeding and/or pain with common gynecological conditions, such as uterine fibroids and endometriosis.
- Excessive or prolonged bleeding following a gynecological procedure or surgery.

Q. How do bleeding disorders affect pregnancy and childbirth or miscarriage?

- Bleeding can occur during pregnancy or with miscarriages.
- Some types of inherited bleeding disorders can cause an increased risk of miscarriage and loss of baby (fetus) such as severe deficiency of clotting proteins such as fibrinogen or factor 13. It is important to note, however, that for the most common bleeding disorders (carriers of hemophilia, VWD, mild platelet function defects), there is no increased risk of miscarriage.
- Women with bleeding disorders can bleed with procedures, such as spinal anesthesia, Cesarean section, and surgery to remove any remaining products of conception that are still inside the uterus following a miscarriage or termination of pregnancy. Women with bleeding disorders are also at risk of bleeding with any invasive medical intervention such as prenatal diagnostic tests (chorionic villus sampling [CVS] and amniocentesis). For those who are undergoing IVF treatment, the process of ovum collection and embryo transfer can also be associated with bleeding. These bleeding complications can be prevented with prior correction of the disorder.
- Excessive and sometimes dangerous bleeding after childbirth, called post-partum-hemorrhage, can occur. Following birth, the bleeding can be immediate (within 24 hours of birth) or delayed (24 hours to 6 weeks after delivery). Iron deficiency anemia can occur due to blood loss.
- Since bleeding disorders typically run in families, your baby may also be affected and at risk for bleeding. Therefore, you should avoid certain procedures during labor, such as vacuum or forceps delivery, or invasive fetal monitoring (e.g. application of clip to baby's head used to monitor baby's heart rate to prevent head bleeds [brain hemorrhage] in the baby). A doctor specializing in high-risk pregnancies should be consulted.
- Undiagnosed women with inherited bleeding disorders may be at risk for exposure to blood and blood products, and may experience complications such as transfusion reactions and exposure to blood borne infections.
- Excessive bleeding after childbirth (postpartum hemorrhage). Although this may occur at the time of delivery, women with bleeding disorders are also at risk of bleeding days after delivery (secondary postpartum hemorrhage). It is normal after childbirth to have vaginal bleeding similar to a period; this is called a lochia and is usually red in colour for the first 1-2 weeks, gradually changing to dark red and lightening before completely tapering off. For some women, their lochia will decrease by 2-4 weeks after childbirth. However, it is normal for lochia to last up to 6 weeks. Women with inherited bleeding disorders are at risk of bleeding for 2-3 weeks after delivery or experiencing a prolonged period of bleeding (>6 weeks). This is because clotting factors may increase during pregnancy and then return to their low baseline levels by 2-3 weeks after delivery. For women with inherited bleeding disorders, this return to low clotting factor levels may be accompanied by increased vaginal bleeding. If this occurs, medications such as tranexamic acid may be of benefit. If you have bleeding of concern, you should contact your obstetrician and hemophilia centre.

Q. How are bleeding disorders diagnosed?

Your doctor may do the following:

- Obtain a detailed personal history and family history, and perform a physical examination to check for bruises and bleeding sites.
- You may be asked several questions regarding the history and severity of past bleeding symptoms to generate a “bleeding score”. This is called ISTH BAT (International Society on Thrombosis & Hemostasis Bleeding Assessment Tool). The score varies with age and sex, and the higher the value/score, the more likely it is that you have a bleeding disorder.
- In menstruating individuals, menstrual blood loss can be quantified or measured using a pictorial blood assessment chart (PBAC). Loss of more than 80 ml of blood per menstrual cycle is indicative of heavy menstrual bleeding. For those using a menstrual cup, the blood loss can be measured directly.
- Another screening tool called the Philipp tool asks a series of questions to identify which women and girls should be tested and further evaluated for bleeding disorders.
- The doctor may order blood tests that measure blood counts and iron levels, screening tests to see if blood is clotting properly, and tests that measure levels of specific clotting factors. Sometimes tests are repeated as the results may change with age or pregnancy. In some countries, specialized laboratories may offer genetic diagnosis for bleeding disorders, such as hemophilia, von Willebrand disease, rare bleeding, and platelet disorders.

Q. How are bleeding disorders treated?

There is no cure for inherited bleeding disorders. However, treatment can control symptoms or prevent bleeding.

- Hormone therapies. Hormone therapies are medications that contain female hormones, such as estrogens and progesterone; they may come in the form of a pill, patch, injection, vaginal ring, and intrauterine device (IUD). In women and girls, hormonal therapies can treat heavy menstrual bleeding or other gynecological bleeding.
- Other medications. These include desmopressin and antifibrinolytics. Desmopressin (DDAVP) is given intravenously, subcutaneously, or intranasally for hemophilia A and VWD. It increases the levels of clotting factors. Currently, some formulations of desmopressin are not available. Antifibrinolytics, such as tranexamic acid or aminocaproic acid, stop bleeding by preventing the breakdown of clots.
- Clotting factor concentrates. Clotting factor concentrates replace missing or deficient clotting factors. They are given intravenously and are used to either prevent or treat bleeding episodes. Different clotting factors treat different kinds of bleeding disorders.
- Iron supplements. Iron supplements are best taken as one tablet every other day rather than daily to boost iron absorption. If your iron levels fail to improve, you may need an intravenous infusion of iron; these are given in a hospital clinic as there is a risk of an allergic reaction with intravenous iron.

Q. What are some of the consequences and issues of untreated bleeding disorders in women and girls?

- Poor quality of life and restrictions in work, school, sports, and social activities, due to heavy menstruations every month.
- Anemia which leads to fatigue, and further negatively impacts quality of life.
- Need for blood transfusion.
- Bleeding in other parts of the body, such as joints, head, ovaries.
- Hysterectomy and other surgery to control heavy periods.

Other issues include:

- Fear of stigma associated with having an inherited disorder is often a barrier to testing.
- Financial burden of health care and laboratory tests, which may result in diagnostic delays.
- Period poverty due to inadequate access to and cost of sanitary products. It may negatively impact education, causing girls to miss or drop out of school.

Q. What to do if you suspect you or someone you know may have an inherited bleeding disorder?

- The first thing to do is to seek medical care, either from your own doctor or an obstetrician gynecologist, and request referral to a hematologist (a doctor who specializes in blood diseases).
- A variety of treatment options are available, not only for controlling heavy menstrual bleeding, but also as contraception, such as the pill and placement of a hormonal IUD. Note that hormonal therapies while often used for contraception, have a valuable role in controlling excessive menstrual bleeding. These may be recommended by your health care professional, to reduce menstrual blood loss, even if you are not sexually active.
- Your treatment center may have access to a genetic counselor to help with antenatal counselling or prenatal diagnostic testing in the event you are pregnant or planning on becoming pregnant.
- Geographic accessibility, financial accessibility, and availability of care may be a challenge in some countries. The doctor or specialist can either see you in person, or in an outreach clinic or by telemedicine (virtual or video visit) if you live far away. Please see the World Federation of Hemophilia Treatment Center directory at <http://www.wfh.org> to locate a center close to you.
- If you have a bleeding disorder and need to attend the emergency department, make sure to bring with you a letter or card with details of your bleeding disorder from your Hemophilia Treatment Center (HTC) or specialist. Not all doctors are familiar with the specific needs of people with bleeding disorders, so do not be afraid to advocate for yourself. Inform your HTC that you are attending the hospital so they can communicate with local doctors and provide advice.
- Many doctors and health care providers may not be familiar with bleeding disorders. It is often intimidating to let medical professionals know that you suspect that you have a bleeding disorder. In that case, you may want to seek support from your national/local HTC or health center.

- If you have access to a computer or smart phone, you can download information and share it with the health care team.
- Laboratory tests may not be available in your village or town, and you may have to travel to a city to access a laboratory.
- Treatments such as tranexamic acid, aminocaproic acid, and hormonal therapies may be used to stop bleeding without waiting for a lab test. However, treatment with factor concentrates may require knowing which factor is missing.

Additional Resources

1. Centers for Disease Control and Prevention (CDC), HHS
www.cdc.gov/ncbddd/blooddisorders/index.html
2. European Haemophilia Consortium
<https://www.ehc.eu/bleeding-disorders/women-with-bleeding-disorders/>
3. Foundation for Women and girls with blood disorders.
www.fwgbd.org
4. Hemophilia Federation of America
<https://www.hemophiliafed.org/home/for-patient-families/resources/toolkits/women-bleed-too-toolkit/>
5. National Hemophilia Foundation
<https://www.hemophilia.org/bleeding-disorders-a-z/overview/women-and-bleeding-disorders>
6. Office on Women's Health
<https://www.womenshealth.gov/a-z-topics/bleeding-disorders>
7. World Federation of Hemophilia
<https://elearning.wfh.org/elearning-centres/carriers-and-women-with-hemophilia/>

Authors

Rezan Abdul-Kadir, MD, U.K.

Roshni Kulkarni, MD, U.S.A.

Michelle Lavin, MD, Ireland

Special thanks to the WFH Women with Inherited Bleeding Disorders Committee for their feedback on this document.

Published by the World Federation of Hemophilia (WFH)
© World Federation of Hemophilia, 2021

The World Federation of Hemophilia (WFH) does not engage in the practice of medicine and under no circumstances recommends particular treatment for specific individuals. The WFH makes no representation, express or implied, that drug doses or other treatment recommendations in this publication are correct. For these reasons it is strongly recommended that individual seek the advice of a medical advisor and/or consult printed instructions provided referred to in this publication. The World Federation of Hemophilia does not endorse particular treatment products or manufacturers; and reference to a product name is not an endorsement by the WFH.

For further information please contact the World Federation of Hemophilia
Tel: +1 (514) 875-7944 ▪ Fax: +1 (514) 875-8916 ▪ E-mail: wfh@wfh.org ▪ www.wfh.org