Health Facts for You

UWHealth

Hemophilia

Hemophilia occurs when a protein in the blood, which is needed to form a blood clot, is missing or reduced. This certain type of protein is called a clotting factor. When there is not enough of this clotting factor in your body, you do not stop bleeding as quickly as you should.

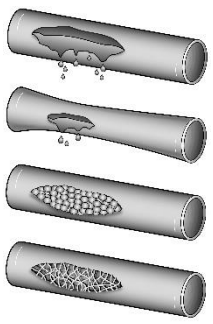
Hemophilia A is when there is a lack of the clotting protein called factor VIII (eight).

Hemophilia B is when there is a lack of the clotting protein called factor IX (nine).

Bleeding and Clotting

1. Bleeding starts when a capillary is injured and blood leaks out.
2. The capillary tightens up to help slow the bleeding.
3. Then blood cells called platelets make a plug to patch the hole.
4. Next, many clotting factors in plasma (part of the blood) work together to form a clot over the plug. This makes the plug stronger and stops the bleeding.

In hemophilia, the missing clotting factor makes it hard to form a clot. Your bleeding will last longer than usual, not faster.



Levels of Hemophilia
How severe it is depends on how much clotting factor is missing from your blood. Normal clotting factor levels are 50-150%.

Mild: A factor level of 5-50%. Bleeding occurs only after a major injury, surgery or dental work.

Moderate: A factor level of 1-5%. Bleeding occurs after the above and after smaller injuries.

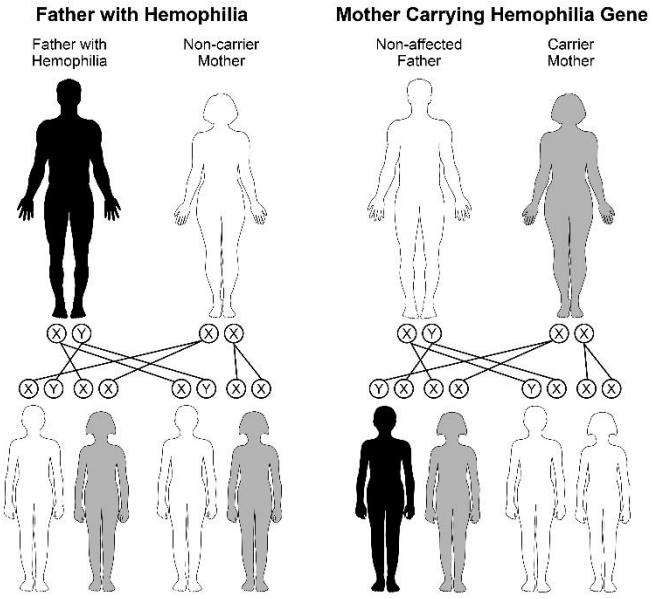
Severe: A factor level of less than 1%. Bleeding can occur after the above but can also occur for no clear reason.

Your level of hemophilia **does not** change over time. The level of hemophilia is the same among family members. If your family has a mild level, then all members who have hemophilia will also have a mild level. But family members can have different patterns of bleeding.

Genetics
Hemophilia is passed down through the family.

- When the father has hemophilia, none of the sons will have hemophilia but all the daughters will be carriers.
- Women who have the hemophilia gene are called carriers. They may show signs of hemophilia, and they can pass it on to their children. For each child, there is a 50% chance that a son will have hemophilia and a 50% chance that a daughter will be a carrier.

- Women can only have hemophilia if their father has hemophilia, and their mother is a carrier. This is very rare.
- Sometimes hemophilia can occur when there is no family history. This occurs in about 30% of new cases.



Types of Bleeds and Symptoms

- **Head** – symptoms may include headache, neck ache, irritable, dizzy or trouble walking, sleepy, sensitive to light, nausea, vomiting, or loss of consciousness.
- **Eye**- symptoms may include pain around the eye, swelling, change in skin color around the eye, or change in vision.
- **Neck and throat** – symptoms may include having trouble swallowing or breathing, altered speech or crying, skin color changes around the neck, or tongue swelling.
- **Abdominal/Gastrointestinal (GI)** – symptoms may include blood in vomit or vomit that looks like coffee grounds, black- or tar-colored stools, nausea or stomach pain.

Joint and Muscle Bleeds

Bleeding occurs most often inside joints and muscles. When a bleed occurs, blood builds up in the joint or muscle space. This buildup of blood causes swelling and pain. It takes time for the body to reabsorb the blood. The enzymes that are released to reabsorb the blood also can cause breakdown of healthy tissue inside joints. Getting treatment for a joint or muscle bleed as soon as you can, will limit the amount of blood that builds up and decrease the risk of long-term problems.

- **Early signs**- a hard hit to the area, a bubbling or tingling feeling inside the joint, a warm feeling inside the muscle or joint, or a baby who is upset or crying for no reason (not because of hunger, thirst, or a need to be held).
- **Late signs**- the skin over the muscle or joint feels warmer than other skin, swelling, pain, stiffness, signs of poor blood flow around the joint or muscle such as cool, numb, or pale skin, or a young child who won't straighten or use an arm or leg.

Other Bleeds

- Mouth and nose
- Cuts and scrapes

For severe or life-threatening bleeds –call 911 or go to the ER.

Treatments

There are treatments used to prevent bleeding and others that stop bleeding that has already started. Some medicines can do both.

Preventing Bleeds:

- Inject clotting factor into a vein on a regular schedule (prophylaxis) or before surgery or procedures (eg dental extractions)

- Inject emicizumab (Hemlibra®) under the skin every 1, 2 or 4 weeks (hemophilia A only)
- Desmopressin – DDAVP (high concentration) A stronger form of DDAVP can be given into a vein or inhaled through the nose to raise factor levels in mild hemophilia A.
- Antifibrinolytic Medicines such as aminocaproic acid or tranexamic acid) are taken by mouth to slow the breakdown of clots.

Managing Bleeds:

- First aid for minor cuts, bruises, and scrapes.
- Rest, ice, compression, elevation (RICE)
- Acetaminophen (Tylenol®) to treat pain. Avoid aspirin products, ibuprofen (Motrin®, Advil®), and naproxen (Aleve®).
- All of the above medicines, except for emicizumab (hemlibra®), can also be used to treat bleeding. How often you give them depends upon how severe the bleed is.

Inhibitors

Inhibitors occur when the body's immune system reacts to the clotting factor that is infused. The immune system sees the factor as a foreign substance and forms inhibitors, or antibodies, to destroy the factor. Most inhibitors develop in the first 75 clotting factor treatments with the greatest risk in the first 10-20 treatments. Inhibitors are most common in patients with severe Hemophilia A.

The most common sign of an inhibitor is bleeding that is not controlled with a standard dose of factor. Inhibitors are diagnosed by doing a simple blood draw. They are most often treated with a high dose of the clotting factor or bypassing agents.

Activity Guidelines

Everyone with a bleeding disorder should stay active. Being active improves joint and muscle function and can improve your overall well-being. The amount of activity will depend on your skill level. We urge you to do sports such as swimming, dancing and walking.

Avoid contact sports with a high risk of injury such as football, hockey and boxing. For your safety, learn the rules. Wear correct gear such as helmets, pads and guards.

Who to Call

UW Health Comprehensive Program for Bleeding Disorders: **608-890-9493**