

Hemophilia

Hemophilia is a bleeding disorder. It occurs when the body does not make enough of the proteins called clotting factor. Clotting factors help stop bleeding.

There are two main types:

Hemophilia A

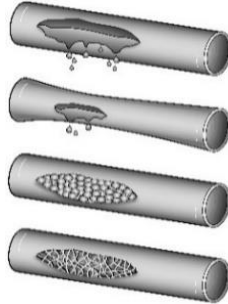
Low or missing factor VIII (8).

Hemophilia B

Low or missing factor IX (9).

How Blood Clots

When a blood vessel is injured or damaged, you start to bleed. The blood vessel tightens to help slow the bleeding. Platelets clump together to form a plug. Clotting factors build a strong clot on top of the plug to stop the bleeding.



When clotting factor is missing or reduced, the clot cannot form well. Bleeding continues longer, not faster.

Hemophilia Severity

Severity describes how serious hemophilia is. The severity depends on how much clotting factor is missing from your blood. Males and females with factor levels less than normal can have hemophilia. Hemophilia tends to have the same severity in families, but bleeding patterns can differ among members.

Signs of Hemophilia

People with hemophilia A or B usually have the same symptoms. These may include:

- Bruising easily.
- Bleeding into joints or muscles, especially in the knees, elbows, or ankles.
- Sudden bleeding inside the body for no clear reason.
- Bleeding that takes a long time to stop after cuts, dental work, or surgery.
- Bleeding that lasts a long time after an injury, especially a head injury.

Bleeding That Occurs with Hemophilia

People with hemophilia can bleed inside or outside the body. The most common bleeding occurs inside, into the muscle or joint. This may happen for no clear reason. It is also common to bleed for a long time after a surgery, removing a tooth or getting a cut.

<u>Normal</u> 50%-150% Normal Clotting.	<u>Mild</u> 5% - 40% Might bleed for a long time after surgery, dental extraction, or a bad injury. Rarely bleeds unless injured.	<u>Moderate</u> 1%-5% Might bleed for a long time after surgery, a bad injury, or dental work. Might bleed for no obvious reason.	<u>Severe</u> less than 1% Bleed often into the joints and sometimes the muscles. Can bleed for no obvious reason.
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Hemophilia and Family History

The severity of hemophilia is consistent among family members, but they may show different patterns of bleeding. Hemophilia is usually passed down from parent to child (inherited). The genes for hemophilia A and B are on the X chromosome.

- A male with hemophilia will pass the gene to all his daughters (who become carriers). The sons will not have hemophilia.
- Sometimes hemophilia can occur when there is no family history.
- Females who have the hemophilia gene can pass it to their children. For each child, there is a 50% chance they will have the hemophilia gene.
- Females who carry the gene may have low factor levels and bleeding symptoms. Female carriers can have

normal factor levels but still have bleeding symptoms.

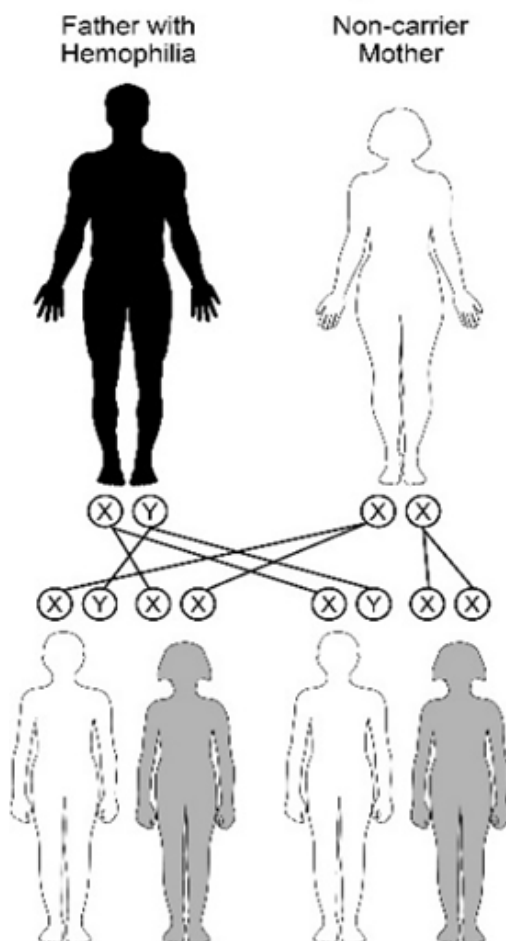
- **Symptomatic carriers:**
Factor levels greater than 40% **with** bleeding symptoms.
- **Asymptomatic carriers:**
Factor levels greater than 40% **without** bleeding symptoms.

Kinds of Bleeds

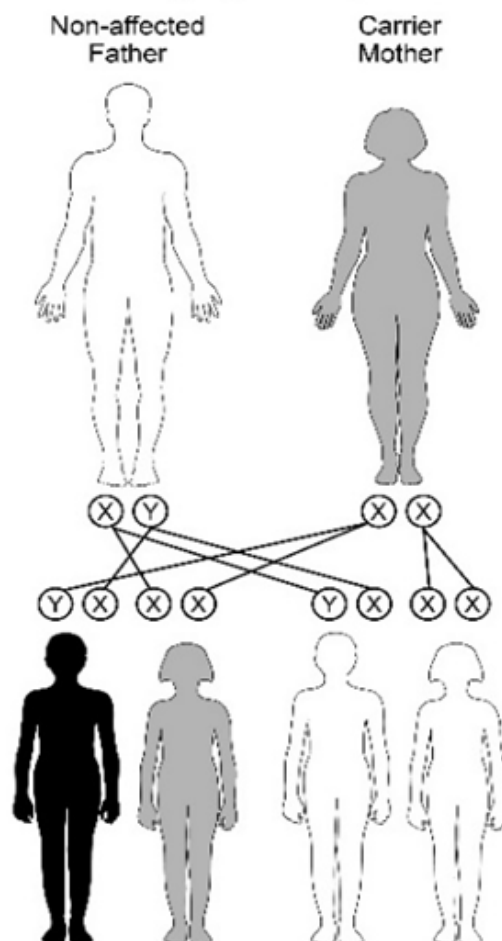
Joint and Muscle Bleeds

Sometimes joint bleeds do not show any visible signs such as bruising. Joint bleeds usually happen in the elbows, knees, or ankles. Get treatment for a joint or muscle bleed as soon as possible.

Father with Hemophilia



Mother Carrying Hemophilia Gene



Bleeding into a joint or muscle causes:

- An ache or “funny feeling”.
- Swelling and warmth.
- Pain and stiffness.
- Difficulty using a joint or muscle.
- Favoring the other limb to avoid using the affected one.

Minor Bleeds

- Mouth and nose
- Cuts and scrapes
- Bruising

Major Bleeds (Severe and Life-Threatening)

Head

Headache, dizziness, sleepiness, nausea, vomiting, and confusion.

- Signs of a head injury may be found right after the injury, **or** they may develop anytime within 2 weeks.

Eye

Pain, swelling, changes in vision.

Neck and throat

Trouble breathing, swallowing, or speaking.

Abdominal/Gut (GI)

Blood in vomit, black or tar-like stools, and stomach pain.

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

Common Bleeds in Young Children

- Muscle bleeding.
- Bleeding after a surgery (heel prick or circumcision).
- Nose, mouth or head bleeds.

Heavy Menstrual (Period) Bleeding

- Bleeding 7+ days.
- Sudden gushes of blood.
- Clots larger than a quarter.
- Soaking through pads or tampons in under 2 hours.
- Feeling tired or dizzy.
- Bleeding that affects daily activities.

Treatments for Bleeding

For minor bleeds, you can use standard first aid measures and medication if needed.

If you have bleeding in your mouth, you can use a wet black tea bag. Place it on the bleeding area and bite down with light pressure. The tea helps stop the bleeding.

You can put ice on a bleed. Wrap the ice in a thin cloth before placing it on the skin. Only use ice for 10 to 20 minutes at a time.

Do not apply heat during a bleed.

For pain management, first use acetaminophen (Tylenol®). Celecoxib is another option to discuss with your provider.

Do not use aspirin products, ibuprofen (Motrin®, Advil®), and naproxen (Aleve®).

After a joint or muscle bleed, follow **RICE**:

- **Rest** the injured area.
- **Ice** to reduce pain and swelling.
- **Compression** with a bandage if needed.
- **Elevation** helps blood flow back toward the heart.

Make sure the joint or muscle feels strong and moves normally before returning to activities, or bleeding may start again.

Ways to Prevent and Control Bleeds

Treatment for hemophilia focuses on preventing bleeds and controlling them when they happen.

There are two main ways to do this:

- **Prophylaxis (prophy):** Regularly scheduled treatments to prevent bleeding. People with moderate or severe hemophilia A or B often prophyl.
- **On-Demand:** Given after a bleed starts, or before and after a procedure to prevent bleeding.

Treatment Options for Hemophilia

There are several treatments that help prevent and control bleeding in hemophilia. Patients, caregivers, and the care team decide on treatment together based on the patient's goals, type and severity of hemophilia, and where bleeding occurs.

Some treatment options your care team might talk to you about are:

- Standard and Extended Half-Life Clotting Factor Concentrates
- Bispecific Antibody
- Desmopressin (DDAVP)
- Antifibrinolytic Medicines
- Hormone Therapy
- New and Investigational Therapies

Inhibitor

An inhibitor is when the body thinks the clotting factor medicine is something harmful and tries to attack it. If you are not responding to your treatments, it can be a sign of an inhibitor. Inhibitors are diagnosed with a blood test and would change your treatments.

Safe Activities

Regular exercise keeps your joints healthy, your muscles stronger, and supports overall health. Choose activities that are safe for you and match what you can do.

- Learn the rules of your activity.
- Use safety gear, like helmets, pads, and guards.
- Talk to your doctor if you are not sure if a sport is safe for you.

Activity Recommendations	Activities to Avoid
• Swimming • Golfing • Biking with a helmet • Baseball • Running/Walking	• Football • Hockey • Wrestling • Rugby • Boxing

Follow-Up Care

UW Health has a team of specialists trained to care for children and adults with hemophilia. We offer one-on-one support to help you manage your condition and stay healthy.

Your care plan will include visits with a provider, nurse, social worker, and/or physical therapist. How often you come to clinic depends on your needs. Most patients have a yearly check-in and may have blood work done at that visit.

Who to Call

(608) 890-9493 (direct clinic line)
Monday – Friday, 8:00 am – 4:30 pm.

After hours, weekends, or holidays, please contact the pediatric or adult hematologist (doctor of blood) on call at **(608) 262-0486** or **1-800-323-8942**.