



Understanding and Treating Hemophilia

SANFORD[®]
HEALTH

Emergency Contacts:

Health Care Providers:

Others:

Welcome

Learning about bleeding disorders can be overwhelming.
Sorting through all the information can be hard.

This handbook will help explain:

- **Hemophilia**4
- **Bleeds**7
 - Soft tissue bleeding or bruising7
 - Joint bleeds7
 - Muscle bleeds8
 - Nosebleeds8
 - Heavy menstrual bleeding9
 - Early home treatment9
- **Emergency Care**11
 - Head11
 - Eye12
 - Throat, neck, and mouth12
 - Belly and bowel12
 - Urine13
 - Deep cuts and broken bones13
- **Medications**14
- **Inhibitors**15
- **Living With Hemophilia**16
- **Hemophilia Treatment Centers**18
- **Online Resources**20

Hemophilia

Hemophilia (heem-oh-feel-ee-ah) is a bleeding disorder that causes you to bleed longer than normal when cut, injured, or sometimes without cause. It is an inherited disease.

People with hemophilia do not bleed more than others bleed; they just bleed longer.

There are two types of hemophilia:

- Hemophilia A – low levels of factor VIII (8).
 - About 1 in 5,000 males is born with hemophilia A each year.
- Hemophilia B – low levels of factor IX (9).
 - About 1 in 25,000 males is born with hemophilia B each year.

It is important to know what type of hemophilia you or your child has. The medicine used to treat hemophilia A is different from the medicine used to treat hemophilia B.



Setting the Record Straight

- Hemophilia has no cure at this time.
- There are treatments to help you lead a healthy and active life.
- Hemophilia is not contagious (you cannot “catch it”).
- People with hemophilia do not bleed more – they just bleed longer.
- Hemophilia does not only affect males. Some females can have low factor levels consistent with mild hemophilia.
- Herbal supplements, vitamins and diet do not treat hemophilia.
- Usually baseline factor levels do not change over time.

How Will Hemophilia Affect Me?

Hemophilia affects people differently. The amount of clotting factor (either factor VIII or factor IX) in your body tells us whether you have mild, moderate, or severe hemophilia.

Do you know what your level of clotting factor is? Are you mild, moderate or severe?

Level of Clotting Factor

Severity of Disease

5-40 percent	Mild hemophilia
1-4 percent	Moderate hemophilia
Less than 1 percent	Severe hemophilia

Inheritance – It Is in the Genes

- Chromosomes are threads inside a cell that carry instructions for how a person's body looks and works.
- Women have 2 X chromosomes (XX). Men have 1 X and 1 Y chromosome (XY). The hemophilia gene is carried on the X chromosome (X-linked recessive).
- Men with hemophilia do not pass the gene on to their sons. Their sons will not have hemophilia.
- All daughters of men with hemophilia will be carriers.
- Carriers have a 50 percent (1 out of 2) chance of passing the gene on to each of their children.
- Each son of a carrier has a 50 percent (1 out of 2) chance of having hemophilia. Each daughter has a 50 percent (1 out of 2) chance of being a carrier.
- Carriers will have varying factor levels. Some will have low levels like mild hemophilia.
- 3/4 of all cases are inherited. 1/4 are thought to be spontaneous gene mutations.

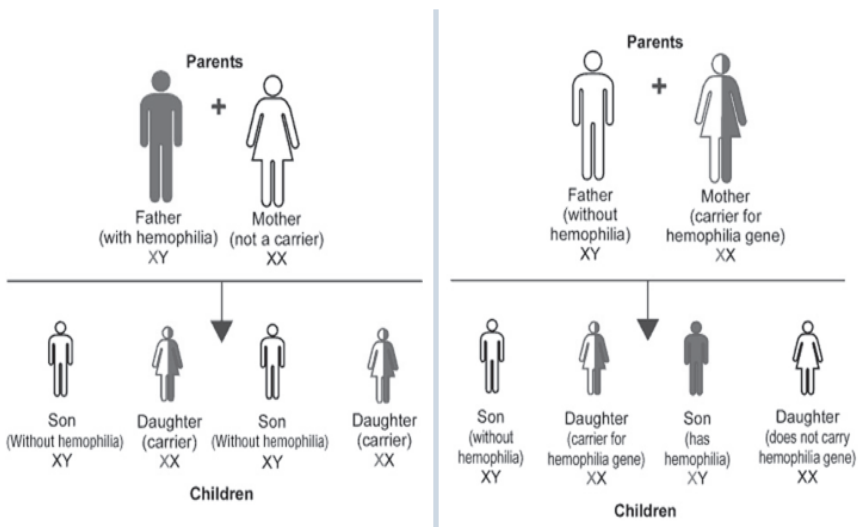


Image used with permission from Indiana Hemophilia and Thrombosis Center

Bleeds

Bleeding happens when a blood vessel is broken. We call this a bleed. This can happen for no known reason or from an injury. Bleeds do not always show bruising, making them harder to spot.

You need to:

- Listen to your body for cues that may indicate a bleed.
- Know the different types of bleeds.
- Know how to treat them.
- Record your bleeds. It is very important to keep a detailed record of your bleeding episodes. Be sure to include how many factor doses were required to treat the bleed.
- Tell your hemophilia provider about your bleeds.

Early Home Treatment

Early treatment of bleeding is especially important for people with hemophilia. Follow these steps when you have a bleed:

- Take factor as soon as you see or feel any signs of bleeding. If you suspect a bleed, always factor first. The signs of bleeding may be:
 - Tingling
 - Warmth
 - Bubbling
 - Pain
 - Difficulty moving the affected area
 - Swelling
- R.I.C.E. (Rest, Ice, Compress, Elevate)
 - Rest: Do not use the affected area until the bleed completely resolves.
 - Ice: Wrap ice in a towel and apply to the site of injury for no more than 20 minutes at a time.
 - Compress: Wrap the affected area with an elastic bandage to limit bleeding and support the joint.
 - Elevate: Raise the affected area above the level of the heart to decrease pain or swelling.
- Call your doctor and/or go to the nearest emergency department.

Soft Tissue Bleeding or Bruising

Soft tissue bleeds or bruises are very common in those with hemophilia. The bruises are caused by bleeding underneath the skin near the surface. This bleeding may cause a raised area, called a hematoma.

Mild bruises and soft tissue bleeds rarely need treatment with factor concentrate.

Joint Bleeds

Joint bleeds occur in the space where two bones meet. They happen most often in the elbows, knees, and ankles. Joints in the shoulders, wrists, fingers, hips, and toes can bleed also. When the joint fills up with blood, the pressure causes pain.

It is important to treat these bleeds immediately in order to prevent permanent damage.

Signs of a Joint Bleed

- Swelling and/or pain
- Bubbling, tingling or a warm feeling

Your child may not be able to tell you if he is having a joint bleed. The following symptoms may indicate a problem:

- More irritable than normal
- Cannot be calmed even with basic cares and needs being met
- Limping, favoring a particular arm or leg, or limiting use of a joint

Muscle Bleeds

You can also have bleeding into the muscles. These occur most often in the arms, legs, and thighs. This can happen because of an injury or for no reason at all. As the blood builds up in the muscle, it can cause pressure on the nerves and blood vessels that may be very painful.

Signs of a Muscle Bleed

- Trouble moving your arm or leg
- Pain in the muscle
- Muscle feels tight, swollen, and warm to the touch
- Numbness or tingling
- Changes in skin color (red, blue or white)

Your child may not be able to express to you when there is a muscle bleed. Signs to watch for:

- Unable or refuses to move arm or leg
- May crawl instead of walk
- Older child may complain of a pulled muscle

Nose Bleeds

Causes:

- Injury to the nose or face
- Breathing very dry air
- Nose picking
- Sneezing and strong nose blowing
- Strep throat
- Cigarette smoking

What to do if you have a nose bleed:

1. Pinch bridge of nose, lean slightly forward, and hold pressure for a minimum of 5-10 minutes. You may apply ice (covered) to the nose.
2. If you have bleeding longer than 30 minutes, seek urgent medical attention.
3. If nose bleeds become frequent, your doctor may consider medicines to reduce their frequency or refer you to an ear, nose, and throat specialist.

Heavy Menstrual Periods

Hemophilia carriers (women) commonly have issues with heavy periods. Contact your doctor for treatment options.

Signs you may notice:

- Saturating a pad or tampon every 1 to 2 hours
- Periods lasting longer than 7 days
- Passing of blood clots
- Anemia (low hemoglobin count)



Emergencies

Seek emergency care or call 911 right away if you suspect the following bleeds.

You Are the Expert About Your Care

- Tell the Emergency Department (ED) staff about your disorder and explain your specific needs. To the ED staff, hemophilia is a rare condition. To you, it is your everyday life. In some cases, you will know more about your bleeding disorder than the ED staff.
- If possible, bring your own factor. If the ED supplies your factor, have them show you the box and mix it in front of you to verify it is the right factor for you. Keep supplies in a bag that is easy to grab in case of an emergency.
- Many invasive procedures require pretreatment with factor concentrate. Exceptions are an IV start or a blood draw.

Items to Take With You to the ED

- Factor concentrate.
- Hemophilia treatment letter and medication list.
- Special medical supplies such as port needles.
- Your doctor's phone number or contact information.

Head Bleed

Bleeding in or around the brain may cause permanent damage or even death. It is important to watch for signs of a head bleed if you have been hit in the head. Rarely, head bleeds can occur even without a known injury. This is a true emergency!

Signs of a Head Bleed

- Headache (often described as the worst headache of your life)
- Confusion, trouble paying attention
- Slurred speech
- Drowsiness, sluggishness
- Irritability

- Loss of consciousness (passing out)
- Vision changes
- Sudden, forceful vomiting
- Numbness, tingling or weakness
- Dizziness
- Clumsiness, stumbling, falling, difficulty walking
- Seizures (convulsions)

Eye Bleed

This can occur from injury to the eye or to the surrounding area. Treat promptly and seek urgent medical attention to avoid permanent vision loss.

Signs of an Eye Bleed

- Pain and swelling in and around the eye.
- Change in vision, including double or blurred vision.
- White of the eye becomes red.

Throat, Neck, and Mouth Bleeds

Bleeding can occur in the throat, neck, tongue, or cheek from an injury or for no reason at all. If left untreated, the bleeding and swelling may make it difficult to breathe.

Treat this type of bleed immediately!

Signs of a Throat, Neck, or Mouth Bleed

- Face, jaw, tongue or neck swelling
- Choking or difficulty swallowing
- Trouble breathing
- Throwing up blood

Belly or Bowel Bleed

Bleeding from the stomach or the bowels can lead to serious complications. Bleeding in the belly or bowels can begin after surgery, after an injury, or sometimes it can happen for no reason at all. Prompt treatment is crucial.

Signs That You May Have Belly or Bowel Bleed

- Red or dark colored vomit (may look like coffee grounds)
- Red, maroon, black, or tar-colored stools
- Unexplained belly swelling or pain

Urinary Bleeding

Bloody urine is never normal. You may see pink, red, or cola-colored urine.

Contact your doctor immediately if you notice blood in your urine.

Deep Cuts and Broken Bones

With these types of injuries, you can have prolonged bleeding and delayed healing. It is important to treat promptly and contact your doctor.

- Apply direct pressure to the cut.
- Do not move the area if you suspect a broken bone.
- Raise the affected area above the level of the heart.
- Apply ice as needed.
- If there is a foreign object lodged anywhere in your body, do not remove it.



Medications

Factor Replacement

The clotting factor is injected into the vein and goes directly into the blood to help make a clot to stop the bleeding.

This treatment will temporarily replace the missing or low levels of factor VIII (8) or factor IX (9) that you or your child needs.

There are many different factor products available for both hemophilia A and B. The Hemophilia Treatment Center staff will help you decide which product is right for you.

- **Plasma-derived** – Factor made from the blood of other people. Companies use several methods to get rid of viruses and unwanted proteins in factor made from plasma. Read the package insert or ask your hemophilia treatment center to find out how the factor you use is treated.
- **Recombinant** – Factor that is created in the lab without using human blood. Animal cells are used to produce these factor proteins.

Treatment Options

- **On demand** – Giving factor only with bleeding episodes.
- **Scheduled medication to prevent bleeding** – Regularly infusing factor to prevent bleeding.
- **Non-Factor Hemophilia Treatment** – Efficizumab (Hemlibra (R)) - Monoclonal antibody that mimics factor VIII to prevent bleedings. This treatment is used only for Hemophilia A with or without inhibitors.

Inhibitors

Inhibitors are antibodies that develop against the clotting factor. If a person with an inhibitor has a bleed, the regular dose or type of factor will probably not stop the bleed.

- As many as 33 out of 100 people with hemophilia A and 6 out of 100 people with hemophilia B may develop an inhibitor in their lifetime.
- You should suspect an inhibitor if bleeding does not stop after several infusions of your normal factor concentrate treatment. Special treatment is needed.

Other Helpful Medications

Acetaminophen (Tylenol) – Acceptable to use as it does not increase bleeding risk.

Aminocaproic Acid (Amicar) – Prevents a clot from dissolving too quickly. This medication is most commonly used to treat bleeding in your mouth because your saliva (spit) can break down a clot.

- Useful after dental procedures and certain surgical procedures.

Desmopressin Acetate (DDAVP) – Causes the body to release factor VIII. DDAVP can be given intravenously (IV), subcutaneously (a shot), or intranasally (as a nasal spray). The nasal spray form of this medication is called Stimate. Possible side effects of DDAVP include facial flushing, increased heart rate, red conjunctiva, and headache.

- May be useful in treating people with mild hemophilia A.

Tranexamic Acid (Lysteda) - Antifibrinolytics - These help prevent a clot from dissolving too quickly. They can be used to treat bleeding in the mouth, nose, heavy menstrual periods, after dental procedures or surgeries affecting the mucous membranes.

Medicines to Avoid

- Do not use aspirin. This can cause prolonged bleeding.
- Non-steroidal Anti-Inflammatories (NSAIDS) – Ibuprofen, Aleve, Motrin, naproxen and others should be avoided as they can cause prolonged bleeding as well. Talk with your hemophilia treatment center prior to taking any of these medications.

Living With Hemophilia

Babysitters and Daycare

You want to be comfortable when leaving your child in the care of others. Communication and education are the keys.

- Your hemophilia team has many resources for you. They will provide education to the caregivers at your request.
- Ask your daycare provider to call you right away with any concerns about the health and safety of your child.

Schools

The school should be aware of your child's bleeding disorder so they know what to do in case of an injury.

Consider a 504 Accommodation Plan through your school. This is a form that outlines the specific needs of your child. It lists any accommodations that your child may need to participate fully in school. The hemophilia team will offer to meet with your child's school to provide education if desired.

Medical Identification

People with hemophilia should wear visible medical identification (ID) at all times. The ID should list your name and condition (Hemophilia A or Hemophilia B).

Healthy Weight and Nutrition

Extra weight places strain on the joints and muscles and may lead to bleeds. Focusing on nutrition and weight management can have a positive impact on your quality of life.

- Ask your nurse or doctor if you would like a visit with a dietitian.
- You may work with a physical therapist to develop an exercise program.

Surgery

Contact your hemophilia team at least 2 weeks before any planned surgery. The team will develop a plan of care to prevent any bleeding complications with surgery.

Dental Care

It is important to take good care of your teeth.

- Do regular brushing and flossing.
- Visit your dentist twice a year for regular checkups.
- Contact your doctor before any dental procedures.

Immunizations

You or your child should receive all immunizations.

- When possible, subcutaneous immunizations are preferred over intramuscular immunizations.
- Apply direct pressure to the site for 3-5 minutes.
- Then apply ice for 3-5 minutes.



Hemophilia Treatment Centers

Hemophilia is a complex disorder. Good quality medical care from doctors and nurses who know a lot about the disorder can help prevent some serious problems. According to the Centers for Disease Control (CDC) “Often the best choice is a comprehensive hemophilia treatment center (HTC)”.

HTCs are specialized health care centers that bring together a team of doctors, nurses, and other health professionals experienced in treating people with hemophilia.

Hemophilia Treatment Centers are located in Fargo and Sioux Falls. They offer coordinated care and the following services:

- Home infusion training and education
- School and daycare education
- Social services
- Physical Therapy
- Pharmacy services
- Research
- Genetic Counseling

The Health Care Team Revolves Around You

The most important member of the team is you. You are the expert about your health. We will work together to help you learn the skills you need to take care of yourself. We will work with you to make a plan of care that is right for you.



