Understanding and Treating Hemophilia
Emergency Contacts:

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Health Care Providers:

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Others:

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Welcome

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

This handbook will help explain:

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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.
Basic Hemophilia

About bleeding disorders

• Bleeding disorders affect blood clotting.
• Without effective blood clotting, cuts and other injuries may not heal the right way.
• Bleeding into the joints can cause pain and swelling. If not treated, this can lead to permanent damage.

Hemophilia

Hemophilia (say 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.

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.
- The only way to treat or prevent a bleed is by replacing the missing factor protein in the blood with factor concentrate.
- 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 female carriers have low enough factor levels that they also can have bleeding problems.
- Herbal supplements, vitamins and diet do not treat hemophilia.
- The factor levels do not change over time.

Interesting Fact:

- There are currently around 400,000 people with hemophilia in the world.
- There are about the same amount of people with hemophilia in the world as there are elephants.

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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?

<table>
<thead>
<tr>
<th>Level of clotting factor</th>
<th>Severity of disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-40 percent</td>
<td>Mild hemophilia</td>
</tr>
<tr>
<td>1-4 percent</td>
<td>Moderate hemophilia</td>
</tr>
<tr>
<td>Less than 1 percent</td>
<td>Severe hemophilia</td>
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</table>
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.

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.
Emergency Care

Deciding where to get treatment

• Dial 911 for emergency medical assistance.

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.

• Many invasive procedures require pre-treatment 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.

Keep supplies in a bag that is easy to grab in case of an emergency.
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
  - Bubbling
  - Warmth
  - Pain
  - Swelling
  - Difficulty moving the affected area

• **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.
Emergencies

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

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 fractures

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 fracture (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.
Home treatment

Depending on the severity, the following are bleeds that you may be able to treat at home.

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)
Medications

Treating hemophilia

• If you have hemophilia A or B, you are missing a specific factor protein in your blood.

• The best treatment is to replace the missing factor protein.

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.

Factor concentrate

• 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.

• All of the current factor products are thoroughly tested. They are effective and safe.

• All factor products are freeze-dried into a powder and placed in small glass bottles. The powder is called bold factor concentrate and factor.

• The dose and expiration date will be clearly marked on the bottle. Refer to the package insert for specific instructions on how to mix the sterile water and factor (powder).

• After the powder has dissolved in the sterile water, it is drawn into a syringe to be given.
• The medication is then injected into a vein. Many people who use factor concentrate are able to give the medication on their own, without needing to be seen at a healthcare facility for each injection.

Types of factor products
There are two types of factor products available.

• **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. The goal of this method of treatment is to stop the bleeding quickly to minimize damage.

• **Prophylaxis (prophy)** – Regularly infusing factor to prevent bleeding. The goal of this treatment is to keep the levels of factor VIII (8) or IX (9) in the blood high enough to help prevent bleeding.

Potential serious side effects of factor concentrate

Allergic reaction

• Hives
• Tightness of the chest
• Itching
• Swelling
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
Premarin Cream – Estrogen cream may help thicken and strengthen the lining of the vessels and walls inside the nostrils.

• Over time, it may help prevent recurrent nose bleeds, but will not stop active bleeding.

Medication Administration

Intravenous (IV)

• A small tube placed into a vein with a needle to give medication into the vein.

• **Do not** start an IV in the same arm as a bleed or injury.

• You or your family member may learn how to place an IV to give factor at home.

Port-a-Cath (port)

• A port is a small, round disk connected to a plastic tube that is surgically placed under the skin.

• The tube is placed in a large vein to give IV medicines.

• The port is used by placing a needle through the skin into the small, round disk.

• The port can be left in place for many years.

• Ask your doctor if a port may be an option for you.

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.

Everyday Living with Bleeding Disorders

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

• Your doctor has many resources to help teach caregivers.

• We are willing to meet directly with them to make sure they know how to care for your child.

• Ask your daycare provider to call you right away with any concerns about the health and safety of your child.

Schools
Hemophilia does not affect your child’s ability to learn. The school should be aware of your child’s bleeding disorder so they know what to do in case of an injury.

• Consider developing a 504 Accommodation Plan through your school. A 504 Accommodation Plan 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.
Medical identification
People with hemophilia should wear visible medical identification (ID) at all times.

• The ID should list your name and type of illness (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.

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.

Surgery
Contact your doctor before any planned surgery.

• Your doctor will work with your surgical team to make sure you have the right treatment to prevent any complications.

• You may need to receive factor before and for many days after surgery.
Immunizations

You or your child should receive all immunizations.

- When possible, subcutaneous immunizations are preferred over intramuscular immunizations.

- To prevent too much bleeding after the immunization:
  - Apply direct pressure to the site for 3-5 minutes.
  - Then apply ice for 3-5 minutes.
Hemophilia Treatment Centers

Ask your doctor about the nearest hemophilia treatment center, which may offer coordinated care and the following services:

• Home infusion training and education
• School and daycare education
• Social services
• Physical Therapy
• Pharmacy services
• Research
Online resources

The National Hemophilia Foundation (NHF)
http://www.hemophilia.org/

Hemophilia Federation of America
http://www.hemophiliafed.org/

World Federation of Hemophilia
http://www.wfh.org/

Medline Plus
http://www.medlineplus.gov

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