Hemophilia

What is Hemophilia?\textsuperscript{1,2}

Hemophilia is a bleeding disorder. People with hemophilia are born without or do not have enough clotting factors. Clotting factors are proteins found in the blood that prevent bleeding and make the blood clot. It takes longer for people with hemophilia to stop bleeding. Bruises and small cuts do not cause serious problems for people with hemophilia. But, bleeding inside the body, such as bleeding in the joints and muscles, may lead to problems. Bleeding can also happen during a dental procedure or surgery. A person with hemophilia can also bleed for no reason.

Who gets hemophilia?\textsuperscript{1,2}

Hemophilia is a genetic disorder. Parents can pass it down to their children. Hemophilia almost always occurs in males. Most of the time, it is passed from the mother to the son. In the United States, one in 5,000 boys are born with hemophilia each year. Sometimes people can get hemophilia even if they are not born with it. This can happen from a random change in the person’s genes.

Are there different types of hemophilia?\textsuperscript{1,2}

There are two main types known as hemophilia A and hemophilia B. It is important to know what type of hemophilia you or your child has in order to get the correct treatment.

Table 1. Types of Hemophilia

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemophilia A (Classic Hemophilia)</td>
<td>• More common</td>
</tr>
<tr>
<td></td>
<td>• Missing Factor VIII (eight)</td>
</tr>
<tr>
<td>Hemophilia B (Christmas Disease)</td>
<td>• Less common</td>
</tr>
<tr>
<td></td>
<td>• Missing Factor IX (nine)</td>
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</tbody>
</table>
**How is hemophilia classified?**

Hemophilia can be mild, moderate, or severe. Your hemophilia level does not change over time. Your level depends on the amount of clotting factor in the blood. The clotting factor level tells you what percent of clotting factors are working compared to average levels. Remember that clotting factors are proteins that stop a bleed. Knowing you or your child’s clotting factor level helps you know what to expect during a bleed.

**Where do bleeds happen and how will I know?**

Common bleeds include bruises, bleeding in the joints, bleeding from the nose, bleeding in the mouth, and deep muscle bleeding. Serious bleeds that are emergency situations are head bleeds, neck or throat bleeds, stomach bleeds, kidney or bladder bleeds, and eye bleeds. Below are symptoms of bleeds. When in doubt, seek treatment or call your hemophilia treatment center or doctor. Do not hesitate to call your doctor.

**Table 2. Symptoms of Bleeds**

<table>
<thead>
<tr>
<th>Bleeding Site</th>
<th>Symptoms of Bleeds</th>
</tr>
</thead>
<tbody>
<tr>
<td>External bleeding (bleeding on the body’s surface)</td>
<td>• Bleeding in the mouth from a cut, a bite, or losing a tooth</td>
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<tr>
<td></td>
<td>• Nosebleeds for no reason</td>
</tr>
<tr>
<td></td>
<td>• Heavy bleeding from a small cut</td>
</tr>
<tr>
<td></td>
<td>• Bleeding from a cut that starts after stopping for a short time</td>
</tr>
<tr>
<td>Kidneys or bladder</td>
<td>• Blood in the urine</td>
</tr>
<tr>
<td>Intestines or stomach</td>
<td>• Blood in the stool</td>
</tr>
<tr>
<td>Joints (knees, elbows, or other joints)</td>
<td>• Joint swelling</td>
</tr>
<tr>
<td></td>
<td>• Joint stiffness</td>
</tr>
<tr>
<td></td>
<td>• Joint pain</td>
</tr>
<tr>
<td></td>
<td>• Joint is warm to the touch</td>
</tr>
<tr>
<td>Muscles:</td>
<td>• Muscle tightness</td>
</tr>
<tr>
<td>• Upper arms</td>
<td>• Muscle pain</td>
</tr>
<tr>
<td>• Forearms</td>
<td>• Muscle feels warm</td>
</tr>
<tr>
<td>• Calf</td>
<td>• Muscle swelling</td>
</tr>
<tr>
<td>• Thigh</td>
<td>• Tingling or numbness</td>
</tr>
<tr>
<td>• Buttocks</td>
<td>• Unable to straighten arm or leg</td>
</tr>
<tr>
<td>• Groin</td>
<td></td>
</tr>
<tr>
<td>Bleeding Site</td>
<td>Symptoms of Bleeds</td>
</tr>
<tr>
<td>---------------</td>
<td>-------------------</td>
</tr>
</tbody>
</table>
| Brain         | • Repeated vomiting  
|               | • Long-lasting headaches or neck pain or stiffness  
|               | • Sleepiness or changes in behavior  
|               | • Double vision  
|               | • Weakness or clumsiness  
|               | • Convulsions or seizures |

These symptoms may not show up until hours or days later. All head injuries are a Medical Emergency. Please go to the Emergency Room or contact your doctor right away.

What can I do to prevent bleeds?¹

For your child with hemophilia:
- Choose soft toys without sharp corners.
- Pad clothing if bruising occurs often when your baby is learning to walk.
- Use protective gear such as helmets and kneepads for activities.
- Use safety belts and straps in highchairs, car seats, and strollers to help protect your child.
- Immunization shots should be given under the skin and not into the muscles. Apply pressure for five minutes after the shot is given.
- Give your child’s teacher and others who care for your child information about hemophilia and how to handle a bleed. Ask your OptumRx pharmacist for handouts.

For everybody with hemophilia:
- Use good dental hygiene. Talk to your doctor before all dental procedures. Tell the dentist that you or your child has hemophilia.
- Avoid aspirin or medications that contain aspirin.
- Carefully use Non-Steroidal Anti-Inflammatory Drugs (NSAIDs), such as ibuprofen (Advil®, Motrin®) and naproxen sodium (Aleve®). They can increase the risk of bleeds. The use of acetaminophen (Tylenol®) for pain is okay.
What do I do if there is a bleed?¹

- Treat bleeds early with factor replacement therapy as directed by your doctor.
- Contact your doctor.
- Keep a record of bleeds - include the name of the factor used, how much was used, the lot number, and the expiration date. This will help your doctor track your therapy to create the best treatment plan for you. Your OptumRx pharmacist can provide you a log to help you track your bleeds and infusions.
- For joint bleeds, try RICE. It can reduce swelling, pain and joint damage when used together with factor replacement therapy. RICE stands for:
  - Rest – it’s important to rest the joint that is bleeding.
  - Ice – use ice packs wrapped in a thin towel on the bleeding joint. Put the ice pack on for 15 minutes and remove for 45 minutes, then repeat.
  - Compression – wrap an ACE bandage around the bleeding joint. If you feel throbbing or if the wrap feels too tight, remove it and rewrap the joint so it’s looser.
  - Elevation – raise the bleeding joint so that it’s higher than your heart.

What is the main treatment for hemophilia?¹

Replacement therapy is the main treatment for hemophilia. Clotting factor is replaced by slowly dripping or injecting clotting factor into a vein. Clotting factors made from human blood are called plasma-derived factor products. Clotting factors that are not made from human blood are called recombinant factor products. Replacement therapy can be used regularly to help prevent bleeding. Or it can be used only when needed to stop a bleed.

Can I use factor products at home?

Many people use factor products at home for themselves or for their children. Special instructions are needed before using factor products at home. Your doctor or nurse will show you how.

How do you store factor products?

Some factor products may need to be refrigerated or stored in a certain way. You can look for storage instructions in the medication guide that came with your factor product. Or you can contact your OptumRx pharmacist if you have any questions about factor product storage.
**What do I do if I run out of factor product when treating a bleed?**

Always make sure you have enough factor product in case you have a bleed. Use an infusion log to write down how much factor you have used and how much is left. This way you’ll know if you’re running low. Be sure to refill your factor product before you run out.

If you run out of factor product when you have a bleed, go to the nearest Emergency Room or Hemophilia Treatment Center. They should be able to provide you with additional factor products.

**Do I need to be concerned about getting infections from factor products?¹,⁴**

The chance of infection has decreased because blood donors are checked. Donated blood products are tested and treated to kill viruses. There is a smaller chance of getting an infection from recombinant factor products than plasma-derived products. Register for the Patient Notification System free of charge. This program tells people about recalls on any of the factor products.

**Patient Notification System**

Phone: 1-888-UPDATE-U (1-888-873-2838)

Web site: www.patientnotificationsystem.org

**What are Hemophilia Treatment Centers (HTC)?¹**

Hemophilia treatment centers (HTC) are resources for families and people with hemophilia. Medical experts at HTCs provide treatment, education and support. They teach people how to do home treatments and can also provide information to your doctor. People who get care at HTCs are:

- Less likely to have bleeding problems.
- Less likely to have to go to the hospital.
- More likely to have a better quality of life.

How often a person visits a HTC will vary. Some individuals are followed closely by their doctors and only go to a HTC once or twice a year for check-ups, while others may go more often. For a listing of HTCs near you, please visit the Centers for Disease Control and Prevention at: www.cdc.gov/ncbdd/blooddisorders/index.html and click on Treatment Centers.
Where can I go to find more information about hemophilia?

OptumRx can give you more information about hemophilia. Some of the topics include:

- Children’s books on hemophilia
- Dental care
- Hemophilia resource guide
- Infusion log
- Nosebleeds
- Prevention
- Raising a child with hemophilia
- Teaching your child about hemophilia

For more information on hemophilia, please contact the following resources:

**OptumRx**
Phone: 1-888-702-8423
Website: www.optumrx.com

**National Hemophilia Foundation Information Resource Center**
Phone: 1-800-42-HAND1 (1-800-424-2634)
Website: www.hemophilia.org

**World Federation of Hemophilia**
Website: www.wfh.org

**References:**


The information in this handout doesn’t replace the medical advice, diagnosis, or treatment provided by your doctor. Always ask your doctor or a qualified health provider if you have any questions about your medical condition.