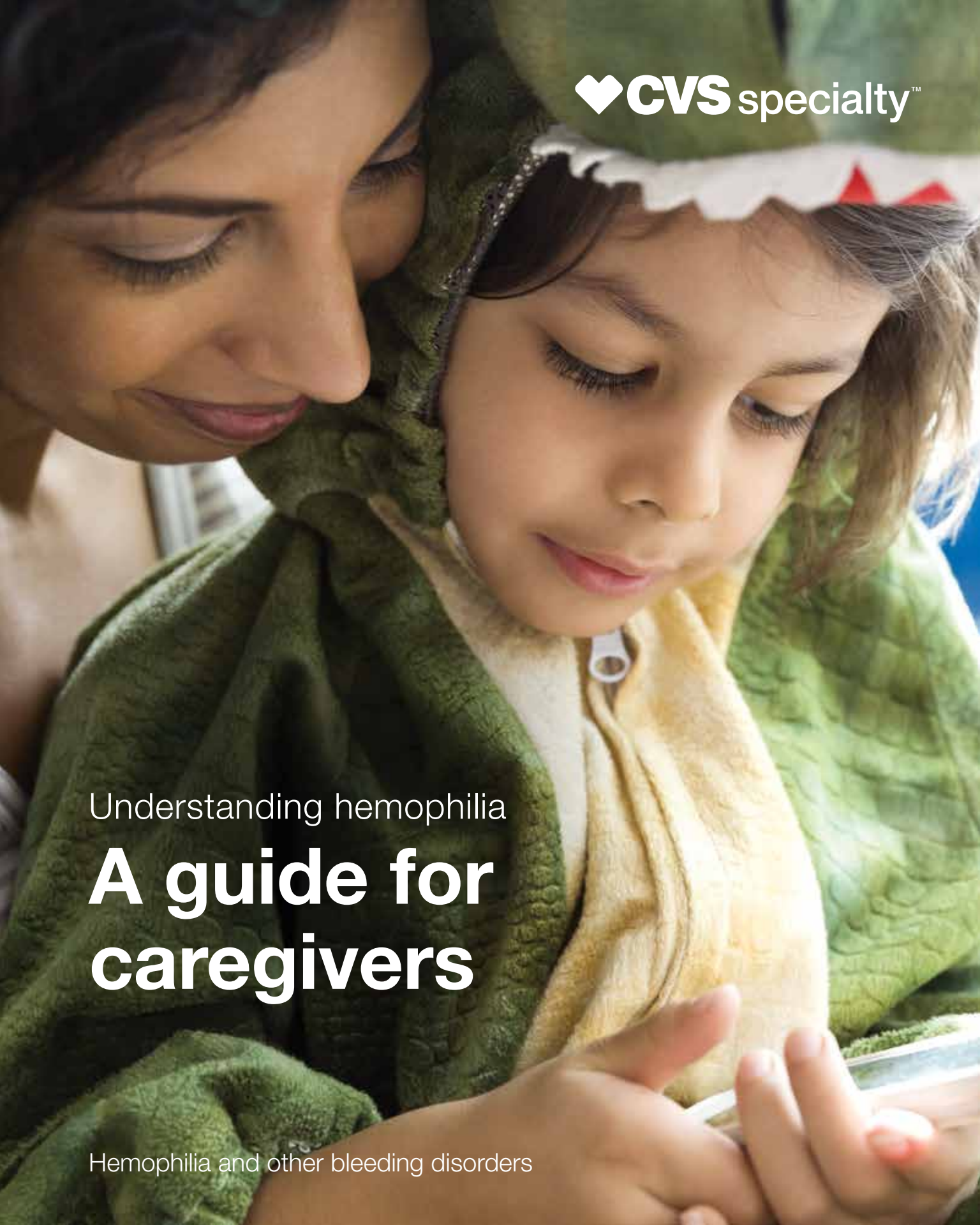
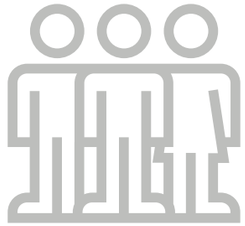


Understanding hemophilia

A guide for caregivers

Hemophilia and other bleeding disorders



As a teacher, coach, babysitter, daycare worker or someone who cares for children and adults, it is important to understand the basics of hemophilia.

Hemophilia affects 1 in 5,000 male births.[1]

Hemophilia is a rare bleeding disorder that causes blood to take a long time to clot. It happens when the blood is missing or does not have enough of a protein called clotting factor. A person with hemophilia will not bleed any faster than a person without hemophilia, but he or she will bleed slower and longer. This type of bleeding disorder most often affects males and is usually passed down through families.

Hemophilia A or B?

There are two types of hemophilia: hemophilia A and hemophilia B. The missing clotting factor determines the type of hemophilia. A person with hemophilia A is missing or does not have enough factor VIII. Type A, also known as classic hemophilia, is four times as common as hemophilia B.[2] If a person is missing or doesn't have the necessary amount of factor IX, he or she has hemophilia B, also called Christmas disease.

How is hemophilia diagnosed?

Hemophilia can be mild, moderate or severe, depending on how much clotting factor is in the blood. Severe hemophilia is usually diagnosed during infancy but the mild form may go undetected until adulthood. A blood test is used to measure the amount of factor VIII or factor IX levels. People without hemophilia normally have a factor level around 100 percent. However, a level between 50 and 150 percent is considered to be normal as well.

Many women who carry the gene for hemophilia have lower than normal levels of clotting factor. Depending on the severity of her condition, a woman may have abnormal clotting. For example, if her mom was a carrier and her father had hemophilia, she may experience abnormal clotting as well. However, if she inherits a milder form she may only experience symptoms during her monthly cycle or not at all.

[1] cdc.gov
[2] hemophilia.org

What causes hemophilia?

Since hemophilia is an inherited disease the gene is passed from parent to child. About 80 percent of people (mostly men) with hemophilia have type A, while the other 20 percent have hemophilia B.[2] If a female carries the gene that causes hemophilia, she can pass it on to her children. The type and severity of the condition is determined by the affected gene, which is passed along to the child.

What are the signs?

Symptoms are different for everyone but the most common are:

- Abnormal bleeding and bruising easily
- Pain and swelling in muscles and joints
- Blood in the urine and stool
- Nosebleeds with no cause
- Severe bruising after receiving immunizations as an infant
- Unusual infant bruising on face, back or stomach
- Delayed healing after an injury

Family history? Test early. If there is a strong family history of hemophilia, a newborn should be tested as soon as possible for the disorder.

The clotting factor gene is found only on the X chromosome.

How is hemophilia treated?

It's important to manage hemophilia by taking steps to stop abnormal bleeding as soon as it starts. The most common treatment is to infuse the missing factor VIII or factor IX into a person's veins to help create a clot. This is known as factor replacement therapy.

Another type of factor replacement therapy is prophylaxis, which means a person receives an infusion of clotting factor on a regular basis to increase the level of factor VIII or factor IX in his or her blood. It is generally used for people with severe hemophilia. Prophylaxis infusions are generally administered on a set schedule and the infusions are most often given in the morning.

By maintaining a higher level of clotting factor, people with hemophilia experience fewer bleeding episodes. While there is no cure for hemophilia, proper treatment helps ensure that most people will lead full, active lives.

Always check with your doctor or pharmacist before taking any medicines.

What is a bleeding episode? Does it hurt?

A bleeding episode occurs when a person with hemophilia has an injury to the tissues in the body. Without the correct amount of clotting factor the clot falls apart, causing re-bleeding in the injured area. Minor scrapes usually don't cause pain or problems, but many bleeding episodes can result in considerable pain if they're not treated promptly. Immediate treatment is the best way to prevent pain and injury. If you are unsure of a bleed, you should infuse.

How serious is the bleeding episode?

Minor — Any bleeding that is caught just as it starts. This type heals quickly and sometimes without much treatment. Most minor bleeds can be treated with basic first aid. Keep in mind that some children may not want to admit they are bleeding so take notice if a child is favoring one limb over the other. Heavy back packs can also lead to minor bleeds.

Major — Any bleeding caused by an injury that involves swelling and pain, or any bleeding that cannot be stopped on its own. A major bleeding episode includes muscles and joints. This type should always be treated promptly to prevent damage to joints or other complications.

Severe — A potentially life-threatening injury or bleeding that can cause permanent damage or death. A severe bleeding episode may include bleeding from surgery or dental procedures, deep muscle bleeding, bleeding inside the head, abdominal bleeding, or bleeding around the throat or neck. Consult a doctor or visit the ER if severe bleeding occurs.

Are there any other medications that can help stop or prevent bleeding?

Before surgery or dental work, people with mild hemophilia are sometimes prescribed a drug called desmopressin acetate (commonly known as DDAVP®). This synthetic hormone stimulates the release of existing factor VIII. It can be given through a vein or as a nasal spray (Stimate®).

Your doctor may also prescribe a medication known as an anti-fibrinolytic agent. This drug, commonly called Amicar® (aminocaprioic acid), slows the normal breakdown of blood clots and is helpful for treating oral bleeding.

People with hemophilia should avoid drugs that can exacerbate bleeding problems, such as aspirin, heparin, warfarin and nonsteroidal anti-inflammatory drugs (NSAIDs), including such drugs like Motrin, Ibuprofen, and Advil.

When is it time to go to the hospital?

Having hemophilia used to mean relying on hospitals and clinics each time you needed clotting factor. These days frequent absences from school and work, and long hours spent in emergency rooms or clinics can be avoided. With the support of hematologists, hemophilia treatment centers, local and national hemophilia foundations, and specialized home care providers, like CVS Specialty, people with hemophilia and their families can learn to recognize bleeding episodes early and infuse factor at home. Decreasing the amount of time families have to spend in emergency rooms and clinics has helped to improve quality of life. Today, children with hemophilia have fewer hospitalizations, spend more time in school, are better integrated with peers and experience less joint damage when compared to previous generations with hemophilia.

Living with hemophilia

Today, new and improved treatments allow most people with hemophilia to lead normal, active lives. Research has shown that being physically active is extremely important for people with hemophilia. Exercise can actually reduce or prevent bleeding episodes by strengthening the muscles surrounding vulnerable joints. The National Hemophilia Foundation strongly encourages regular exercise for people with hemophilia of all ages in order to preserve joint function.

Many sports and activities are safe for a person with hemophilia.

High-risk, full-contact activities to avoid:

- Boxing
- Rugby
- Football
- Wrestling
- Other sports that are considered high risk for head or severe injury

Safe activities to try:

- Swimming
- Walking
- Golfing
- Bicycling

Patients should talk to their CVS Specialty Hemophilia CareTeam about the best activities in which to participate, so they can maintain a healthy and active lifestyle.

Von Willebrand disease (vWD) and hemophilia

vWD is the most common inherited bleeding disorder. It is caused by a deficiency or defect of von Willebrand factor (vWF). Unlike hemophilia, von Willebrand disease affects women and men equally. It is named after Eric von Willebrand, the doctor who first described the bleeding disorder.

The signs and symptoms of vWD depend on which type of vWD the person has and how serious it is. Many people have such mild symptoms that they don't know they have vWD. People who have type 3 vWD may have all of the symptoms listed in the next column and severe bleeding episodes for no reason. These bleeding episodes can be fatal if not treated right away.

The most common symptoms of vWD are:

- Bruising easily
- Increased or prolonged menstrual bleeding
- Frequent and prolonged nose bleeds
- Blood in stool or urine
- Prolonged bleeding after dental work, childbirth, surgery or injury

Early diagnosis of von Willebrand disease is important to make sure that the person is treated and can live a healthy, active life. A simple blood test will allow a doctor to see how much von Willebrand factor is present. Sometimes people have to be tested a couple of times because the von Willebrand factor levels in the blood can go higher (for a day or two) because of exercise, surgery, certain medicines, illness or stress.

Want to know more?

If you have any questions about hemophilia or von Willebrand disease (vWD), talk to your doctor or contact the CVS Specialty Hemophilia Care Program toll-free at **1-866-RxCare-1 (1-866-792-2731)**. One of our hemophilia CareTeam representatives will be happy to answer your questions.

Your name: _____

Your diagnosis: _____

Your doctor's name(s): **Telephone number(s):**

Your nurse's name(s): **Telephone number(s):**

Emergency number(s): _____

Your treatment center name: **Telephone number(s):**

Other team members: **Telephone number(s):**

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