



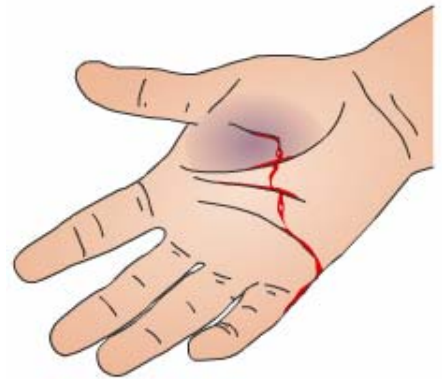
Hemophilia

Introduction

Hemophilia is a rare bleeding disorder in which the blood does not clot normally. About 1 in 10,000 people are born with hemophilia.

Hemophilia can be mild, moderate, or severe. Thanks to advances in medicine, people with hemophilia are likely to live a normal lifespan.

This reference summary will help you understand hemophilia, including its symptoms and causes. It also discusses how hemophilia is diagnosed and treated.



Blood

Blood is made of blood cells floating in plasma. Plasma is mostly made of water with chemicals in it.

There are three basic types of blood cells: red blood cells, white blood cells, and platelets.

Red blood cells are also called erythrocytes. They make up almost half of blood. They help the body by moving oxygen from the lungs to different cells in the body.



Red Blood Cells

Red blood cells are filled with hemoglobin. Hemoglobin is a protein that picks up oxygen in the lungs and delivers it to cells all around the body. It also picks up carbon dioxide, or CO₂, from the cells to be breathed out from the lungs.

White blood cells are also called leukocytes. They fight disease and infection by attacking and killing germs that get into the body.



White Blood Cells

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There are several kinds of white blood cells. Each kind fights different kinds of germs in different ways.

Platelets are also called thrombocytes. They are small pieces of cell that help blood clot and stop bleeding.



Platelets

Special proteins are also needed for blood to clot normally. These proteins are called clotting factors. They are found in the plasma. There are several types of clotting factors.

Clotting factors work with platelets to help blood clot. When blood vessels are injured, clotting factors help platelets stick together to plug cuts in the vessels. This stops bleeding.

Symptoms

The main symptoms of hemophilia are bleeding too much and easy bruising.

How much a person bleeds depends on the severity of the hemophilia. Mild hemophilia may not cause excessive bleeding until a dental procedure, an accident, or surgery.

Bleeding can happen inside the body or can be seen on the outside. Bleeding that can be seen on the outside is known as external bleeding. If the bleeding occurs inside the body and is not obvious on the outside it is known as internal bleeding.

Symptoms of external bleeding include:

- Bleeding from a cut that starts again after stopping for a short time
- Bleeding in the mouth from a cut or bite or from losing a tooth
- Heavy bleeding from a small cut
- Nosebleeds for no obvious reason



Symptoms of internal bleeding include:

- Blood in the stool
- Blood in the urine
- Blood in the brain
- Large bruises

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Bleeding in the knees, elbows, or other joints is another common type of internal bleeding in people with hemophilia. This bleeding can happen without obvious injury. Bleeding first causes the joint to feel tight. The joint then becomes swollen, hot to touch, and painful to bend. Joint bleeding that isn't treated quickly can damage the joint.

Internal bleeding can also happen in the brain after a bump on the head or a more serious injury. This requires immediate medical attention.

Symptoms of bleeding in the brain include:

- Confusion
- Long-lasting, painful headaches
- Neck pain or stiffness
- Repeated vomiting
- Sleepiness
- Convulsions or seizures
- Double vision
- Problems walking
- Sudden weakness

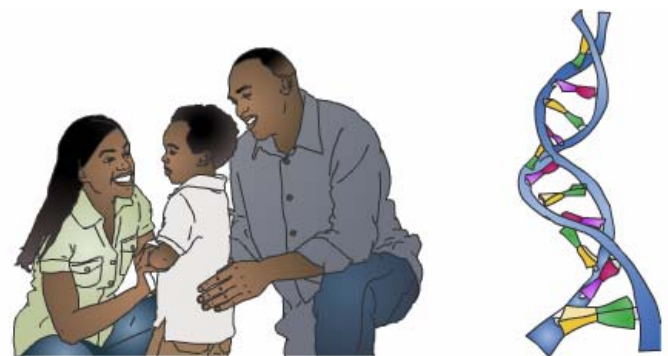


Causes

Hemophilia is often inherited. "Inherited" means that the disorder is passed from parents to children. Hemophilia is most often found in males. Females may also inherit the disorder in very rare cases.

People born with hemophilia have little or no clotting factor. There are two main types of hemophilia: type A and type B.

Different clotting factors are low or missing in each type of hemophilia. About 90% of people with hemophilia have type A.



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Rarely, hemophilia can be acquired. “Acquired” means you are not born with the disorder. Instead, you develop it during your lifetime.

Hemophilia can be acquired if your body forms antibodies that attack the clotting factors in your bloodstream. The antibodies can prevent the clotting factors from working.

Diagnosis

If you have symptoms of hemophilia, your healthcare provider will try to find out if hemophilia is the cause of your symptoms or if there is another cause.

Your doctor will ask you about your family medical history as well as your own. This will reveal whether you or your family members have bleeding problems.

Your healthcare provider will perform a physical exam. Blood tests are also used to diagnose hemophilia.



Blood tests for hemophilia are used to find out:

- How long it takes for your blood to clot
- Whether your blood has low levels of any clotting factors
- Whether any clotting factors are completely missing from your blood

The test results will show:

- Whether you have hemophilia
- What type of hemophilia you have
- How severe it is

Hemophilia A and B are classified as mild, moderate, or severe. This depends on how much clotting factor is in the blood.

People with mild hemophilia have 5 to 30% of normal clotting factor. People with 1 to 5% of normal clotting factor have moderate hemophilia. People with less than 1% of normal clotting factor have severe hemophilia.

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Treatment

The type of treatment for hemophilia depends on:

- The severity of hemophilia
- The activities you will be doing
- The dental or medical procedures you will be having

The main treatment for hemophilia is replacement therapy. Replacement therapy gives you the clotting factor you are missing. Concentrates of clotting factor are given through an IV drip or an injection into a vein.

Clotting factor concentrates can be made from human blood. The blood is treated to prevent the spread of diseases. The risk of getting a disease from human clotting factors is very small with the current methods of screening and treating donated blood.



There are also clotting factor concentrates that aren't made from human blood. These are called recombinant clotting factors.

Some people are treated with replacement therapy on a regular basis. This is called preventative therapy. Others receive treatment only when they need to stop either active or expected bleeding. This is known as demand therapy.

Many people receive their treatment for hemophilia at home. Talk to your healthcare provider about your options for home treatment.

There are also other treatments available for hemophilia. Desmopressin is a hormone that may be used to treat mild or moderate hemophilia A. Desmopressin increases the level of the missing clotting factor in the blood. It is usually used before dental work or before playing certain sports to reduce the risk of bleeding.

Antifibrinolytic medicines are sometimes used with replacement therapy to treat hemophilia. They help keep blood clots from breaking down. Often these medicines are used before dental work or to treat mild bleeding. Clots in the body are broken down with the help of special chemicals. This process is known as fibrinolysis. Medications that prevent the breakdown of such clots are known as antifibrinolytics.

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Pain medicines, steroids, and physical therapy are other treatments for hemophilia. These are often used to treat a specific area of the body that is bleeding.

Talk to your healthcare provider about which type of treatment is best for you. Your healthcare provider will be able to tell you which treatment options may be considered for your type of hemophilia.

For all types of hemophilia, getting quick treatment for bleeding is important. Quick treatment can limit damage to your body.

Summary

Hemophilia is a rare bleeding disorder in which the blood does not clot normally. People born with hemophilia have little or no clotting factor. Hemophilia usually is inherited.

The main symptoms of hemophilia are excessive bleeding and easy bruising. Thanks to improvements in treatment, people with hemophilia today are likely to live a normal lifespan.



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