



What is Hemophilia?

Hemophilia is an inherited bleeding disorder caused by missing or reduced blood clotting factor

Factor VIII clotting protein deficiency = hemophilia A

Factor IX clotting protein deficiency = hemophilia B

1. About 1 in 5,000 males is born with hemophilia
2. Approximately 400,000 people around the world have hemophilia
3. People of all races and economic groups are affected by hemophilia
4. Factor VIII clotting protein deficiency is about 4 times as common as Factor IX clotting protein deficiency
5. Current available treatments allow people with hemophilia to have a life expectancy similar to people without hemophilia

Severity of hemophilia

1. The severity (bleeding) of hemophilia is usually related to the clotting factor level

Severity	Clotting Factor Level % Activity	Bleeding Episodes
Severe	Less than 1%	Spontaneous bleeding, usually in joints and muscles
Moderate	1%-5%	Occasional spontaneous bleeding; severe bleeding with trauma or surgery
Mild	6%-24%	Bleeding with major trauma or surgery

[World Federation of Hemophilia. Guidelines for the management of hemophilia. 2005.]

[Medical Encyclopedia: Hemophilia A. Available at: www.nlm.nih.gov/medlineplus/print/ency/article/00538.htm.]

[Medical Encyclopedia: Hemophilia B. Available at: www.nlm.nih.gov/medlineplus/print/ency/article/00539.htm.]

Symptoms of hemophilia

1. The severity of symptoms in people with hemophilia depends on the severity of the disease
2. Severe hemophilia is usually recognized early—sometimes the first bleeding episode occurs when a boy is circumcised
3. Other bleeding episodes can occur when the infant starts crawling and/or walking
4. Mild cases may go unnoticed until later in life

Common symptoms of hemophilia include the following:

- Pain and swelling
- Bruising
- Spontaneous bleeding
- Joint bleeds (knee, ankle, elbow, hip, shoulder)
- Muscle bleeds (calf, forearm, bicep, iliopsoas)
- Nosebleeds
- Blood in urine/stool, or vomiting blood
- Prolonged bleeding from cuts, pulled teeth, or surgery
- Excess bleeding following circumcision
- Limited movement or loss of range of motion

Complications of hemophilia

1. The major complications of hemophilia are the development of inhibitors and damage to joints:
 - disease and or infection
 - An inhibitor is a protein called an antibody that some people make against factor VIII or factor IX. This inhibitor inactivates the factor so that it cannot work in the body to form a clot and stop bleeding. Although the development of inhibitors complicates hemophilia treatment, there are ways to successfully manage bleeding.
 - Joint damage is caused by repeated bleeding in and around the joint. Hemarthrosis is another name for joint bleed. Although permanent damage can be caused by one serious bleed, normally the damage occurs over years. The severity of the damage depends on the number of bleeds and the extent of the bleeding. Joint damage in people with hemophilia is called hemophilic arthropathy.

[The complications of hemophilia. Canadian Hemophilia Society. Available at: www.hemophilia.ca/en/2.1.9.php. Accessed: January 24, 2008.]

[National Hemophilia Foundation. Facts about inhibitors. October, 2007.]

[Medical Encyclopedia: Hemophilia B. Available at: www.nlm.nih.gov/medlineplus/print/ency/article/00539.htm.]