

Hemophilia Fact Sheet

What It Is

Hemophilia is a hereditary bleeding disorder.¹ In people with hemophilia, blood does not clot normally causing the individual to bleed for an unusually lengthy period of time. The primary concern for people with hemophilia is internal bleeding, mainly into muscles and joints.²

There are two types of hemophilia: hemophilia A and hemophilia B. Both are caused by a low level or absence of one of the proteins in the blood (called factors) that control bleeding. Hemophilia A is caused by a deficiency of factor VIII, and hemophilia B is caused by a deficiency of factor IX.^{1,2}

Causes and Symptoms

Hemophilia is a genetic disorder which is passed down from a parent to a child. It cannot be caught or transmitted. About one third of new cases are caused by a new mutation of the gene in the mother or the child. In these cases, there is no previous history of hemophilia in the family.

Symptoms of hemophilia include large bruises, bleeding into muscles and joints (especially the knees, elbow and ankles), spontaneous or internal bleeding into vital organs, and bleeding for a long time after getting a cut, removing a tooth, or having surgery. Some bleeds can be life-threatening and require immediate treatment. These include bleeds in the head, throat, gut or iliopsoas muscles.

Hemophilia is classified as severe, moderate or mild based on the level of factor VIII or IX in the blood. Severe hemophiliacs with less than 1% of the normal level of factor VIII or IX in the blood have hemorrhages several times a month, often the result of a minor bump or twist, or with no obvious cause. Moderate hemophiliacs bleed less often, usually due to minor trauma, such as a sports injury. Mild hemophiliacs have even fewer hemorrhages and usually bleed only as a result of surgery or major injury. Women with mild hemophilia may bleed more during menstruation.²

Incidence and Prevalence

Hemophilia A affects fewer than 1 in 10,000 people, or about 2,500 Canadians. Hemophilia B is even less common, affecting approximately 1 in 50,000 people, or about 600 Canadians.¹

Hemophilia affects people of all races, colors and ethnic origins.² The most severe forms of hemophilia occur only in males. In extremely rare cases, females can be seriously affected if the father has hemophilia and the mother is a carrier. However, many women who are carriers have symptoms of mild hemophilia.¹

Living with Hemophilia

Without proper treatment, hemophilia is crippling and often fatal. With modern treatment, most people with hemophilia can lead full, active lives.¹

While exercise and sports may be thought to cause an increased bleeding risk for people with hemophilia, exercise can actually help prevent bleeds by increasing muscle strength which decreases spontaneous bleeds and joint damage. However, some exercise programs or sports are riskier than others, and the benefits must be weighed against the risks. The severity of a person's hemophilia should also be considered before beginning any physical activity.²

Diagnosis and Treatment

Hemophilia is diagnosed by taking a blood sample and measuring the level of factor coagulation activity in the blood. These tests can be done at a hemophilia treatment center. Given the hereditary nature of the condition, hemophilia is often diagnosed in the first year of life.¹ A comprehensive family history can also allow for prenatal diagnosis.

Hemophilia is treated by replacing the missing clotting factor in the blood. This is done by injecting a product that contains the needed factor into a vein. Bleeding stops when enough clotting factor reaches the bleeding site. It is very important that treatment is given as quickly as possible to prevent long-term damage.²

Factor concentrates: As the treatment of choice for hemophilia, factor concentrates can be made from human blood (called plasma-derived products), such as Humate-P and or manufactured using genetically engineered cells that carry a human factor gene (called recombinant products), such as HELIXATE[®] FS. All commercially prepared factor concentrates are treated to remove or inactivate blood-borne viruses. Additionally, people with mild hemophilia A sometimes use desmopressin (also called DDAVP), a synthetic hormone that stimulates the release of factor VIII.

Cryoprecipitate: For hemophilia A patients with joint and muscle bleeds, cryoprecipitate contains increased amounts of factor VIII but is less safe from viral contamination than concentrates and is more difficult to store and administer.

References

¹ Canadian Hemophilia Society. Hemophilia A and B. Accessed 4/28/09: <http://www.hemophilia.ca/en/bleeding-disorders/hemophilia-a-and-b/>

² World Federation of Hemophilia. Frequently Asked Questions About Hemophilia. Accessed 4/28/09: <http://www.wfh.org/index.asp?lang=EN>