

Have a Bleeding Disorder? Get tested regularly for an inhibitor.

The sooner it's discovered, the quicker you can get treatment.

Inhibitors — a potentially dangerous health problem — affect people with bleeding disorders, such as hemophilia and von Willebrand disease (VWD). If you have a bleeding disorder, finding an inhibitor early and receiving treatment can lower your risk for developing serious health problems.

Blood contains many proteins called clotting factors that help to stop bleeding. People with hemophilia are missing or have low levels of either clotting factor VIII (factor 8) or factor IX (factor 9). People with the most severe form of von Willebrand disease (VWD), which is called Type 3 VWD, are missing or have low levels of factor VIII. People with these conditions may experience bleeding episodes in their joints, muscles, and internal organs. A bleeding episode is a bleed which lasts longer than several minutes. The bleed can occur in response to little or no physical trauma. Medicines (called clotting factor concentrates, or simply “factor”) rich in the missing proteins are commonly used to treat these conditions. These medicines are infused or injected into a vein to replace the missing proteins in the blood. The infused clotting factor concentrates help to improve the blood's ability to clot in order to stop bleeding episodes and help prevent future ones.

What are inhibitors?

In some people with hemophilia or VWD, their immune systems do not accept the clotting factor concentrates, and will make inhibitors (antibodies to factor) to destroy the clotting factor concentrates. Inhibitors stop clotting factor concentrate treatment from working which makes it harder to stop or prevent bleeding episodes.

People with hemophilia or VWD who develop an inhibitor do not respond as well to treatment with clotting factor concentrates. They are **twice as likely to be hospitalized** for a bleeding episode.

Who develops inhibitors?

If you have a bleeding disorder, you may be at risk for developing an inhibitor. This is true regardless of your age or the severity of your condition. It is estimated that **1 out of every 5 people** with hemophilia will develop an inhibitor in their lifetime. For some people the inhibitor is transient, which means that it may last only a short time and may not require treatment. For other people, however, the inhibitor can have severe health consequences and require special medical treatment.

It is not yet known exactly what causes inhibitors. Certain characteristics may increase your risk for developing an inhibitor, including:

- Number of lifetime exposures to clotting factor concentrates
- Race and ethnicity
- Family history of inhibitors
- Frequency, amount, and type of clotting factor concentrate treatment
- The type of bleeding disorder you have — **hemophilia A, hemophilia B or VWD Type 3** (People with VWD Types 1 and 2 are not prone to inhibitors)
- The specific changes in your genes that caused your bleeding disorder (called a genotype). Genes are the genetic instructions in all living things. The genes that a person inherits from his or her parents can determine many things, like what a person will look like and whether the person might have certain diseases.



“The complications of long-term damage to his joints, the unrelenting pain, just general quality of life, that’s been by far the biggest challenge. Another big challenge for us as a family and, for my husband in particular, is the high cost of hemophilia in general and treating an inhibitor in particular.”

— Jane Cavanaugh Smith

Should I be tested every year for an inhibitor?

Yes. A person with an inhibitor may not show any obvious physical signs or symptoms (such as increased bleeding or requiring more clotting factor concentrates to treat a bleeding episode than in the past). Therefore a laboratory test is necessary to diagnose an inhibitor. It is important to find inhibitors early. Studies have found that the sooner an inhibitor is detected, the more likely a person is to respond to treatment designed to eliminate the inhibitor. In some cases, inhibitor treatment can last several years.

The best way to find inhibitors early is through routine testing. People with hemophilia and VWD should be **tested at least once a year**. The Nijmegen-Bethesda Assay is the best laboratory test to diagnose an inhibitor. For more information on when you should be tested, see the recommendations by the Medical and Scientific Advisory Council of the National Hemophilia Foundation, described [here: http://bit.ly/1RRubUm](http://bit.ly/1RRubUm).

What can I do to help identify a potential problem early?

- **Enroll in Community Counts** (<http://1.usa.gov/1Tt2TXz>) at your local hemophilia treatment center (HTC) and take advantage of the free inhibitor testing provided as part of this project. Community Counts aims to gather and share information about common health issues, health and treatment-related problems, and causes of death that affect people with bleeding disorders who receive their care at HTCs. Participation in Community Counts also helps CDC gather information about inhibitors in the bleeding disorders community. Community Counts is a collaborative effort between CDC, the American Thrombosis and Hemostasis Network (ATHN) and the United States HTC network.
- **Keep records of your infusions** so that your doctor can tell if the prescribed clotting factor concentrate treatment is working well for you.
- **Be alert for changes in how your bleeding episodes respond to clotting factor concentrate treatment** and notify your doctor right away if you notice changes.
- **Get a laboratory test** to determine what changes in your genes caused you to have a bleeding disorder and **ask your doctor about your risk for inhibitors**. People with certain genotypes have a higher risk for inhibitors.
- **Participate in research studies and/or clinical trials** to help researchers learn what increases the risk for inhibitors. Once it is known who has a higher risk of developing inhibitors, it may be possible to create strategies to prevent them from occurring. Data from many people are needed in order to identify what causes inhibitors.

For more information on inhibitors, please visit the CDC website <http://www.cdc.gov/ncbddd/hemophilia/inhibitors.html>.

Get free inhibitor testing today! Community Counts is a CDC-funded public health monitoring program that provides inhibitor testing to more than 135 HTCs across the country.

For more information, visit the Community Counts website today! <http://1.usa.gov/1Tt2TXz>