

# What is von Willebrand disease?



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# What is von Willebrand disease (VWD)?

von Willebrand disease (VWD) is a bleeding disorder. People with VWD have a problem with a protein in their blood that helps control bleeding. They do not have enough of the protein or it does not work the way it should. It takes longer for blood to clot and for bleeding to stop.

There are different types of VWD. All are caused by a problem with the von Willebrand factor (VWF) protein. When a blood vessel is injured and bleeding occurs, VWF helps cells in the blood, called platelets, mesh together and form a clot to stop the bleeding.

VWD is the most common bleeding disorder that people have. It affects both males and females. VWD is generally less severe than other bleeding disorders. Many people with VWD may not know that they have the disorder because their bleeding symptoms are very mild. For most people with VWD, the disorder causes little or no disruption to their lives except when there is a serious injury or need for surgery. However, with all forms of VWD, there can be bleeding problems.

## How do people get VWD?

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VWD is usually inherited. It is passed down through the genes from either parent to a child of either sex. Sometimes there is evidence of a family history of bleeding problems. However, bleeding symptoms can vary a lot within a family. Sometimes there is no family history and VWD occurs due to a spontaneous change in the VWD gene before the baby is born.

# What are the symptoms of VWD?

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The main symptoms of VWD are:

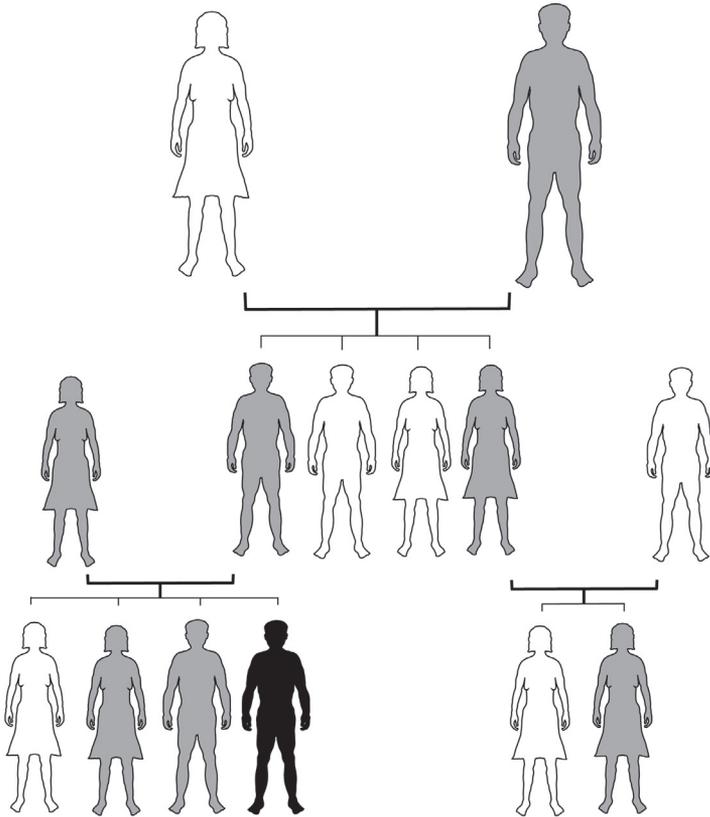
- easy bruising
- frequent or prolonged nose bleeds
- bleeding from gums
- prolonged bleeding from minor cuts
- heavy or prolonged menstrual bleeding
- bleeding in the upper and lower gastrointestinal tract
- prolonged bleeding following injury, surgery, dental work, or childbirth

Many people with VWD have few or no symptoms. People with more serious VWD may have more bleeding problems. Symptoms can also change over time. Sometimes VWD is discovered only when there is heavy bleeding after a serious accident or a dental or surgical procedure.

More women than men show symptoms of VWD. Women with VWD often bleed more or longer than normal with menstruation and following childbirth. Some women with VWD have a lot of menstrual pain or irregular menstruation.

Blood type can play a role. People with Type O blood often have lower levels of VWF than people with Types A, B, or AB. This means people with VWD and Type O blood may have more problems with bleeding.

# Inheritance of von Willebrand disease



## Inheritance of von Willebrand disease



NONE



MILD



SEVERE

## How is VWD diagnosed?

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VWD is not easy to diagnose. People who think they have a bleeding problem should see a hematologist who specializes in bleeding disorders. Proper tests can be done at a bleeding disorders treatment centre. Since the VWF protein has more than one function, more than one lab test should be used to diagnose VWD.

Laboratory testing for VWD is also difficult. VWD cannot be diagnosed with routine blood tests. Testing involves measuring a person's level and activity of VWF, and that of another blood clotting protein, factor VIII (FVIII). Testing is often repeated because a person's VWF and FVIII levels can vary at different times.

## Different types of VWD

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There are three main types of VWD. Within each type of VWD, the disorder can be mild, moderate, or severe. Bleeding symptoms can be quite variable within each type depending in part on the VWF activity. It is important to know which type of VWD a person has, because treatment is different for each type.

**Type 1 VWD** is the most common form. People with Type 1 VWD have lower than normal levels of VWF. Symptoms are usually very mild. Still, it is possible for someone with Type 1 VWD to have serious bleeding.

**Type 2 VWD** involves a defect in the VWF structure. The VWF protein does not work properly, causing lower than normal VWF activity. There are different Type 2 VWD defects. Symptoms are usually moderate.

**Type 3 VWD** is usually the most serious form. People with Type 3 VWD have very little or no VWF. Symptoms are more severe. People with Type 3 VWD can have bleeding into muscles and joints, sometimes without injury.

## How is VWD treated?

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VWD can be treated with a synthetic drug called desmopressin, a clotting factor concentrate that contains VWF, or other drugs that help control bleeding. The type of treatment depends in part on the type of VWD a person has. People with mild forms of VWD often do not require treatment for the disorder except for surgery or dental work.

**Desmopressin** is generally effective for treating Type 1 VWD, and helps prevent or treat bleeding in some forms of Type 2 VWD. It is used to control bleeding in an emergency or during surgery. It can be injected or taken by nasal spray, and raises VWF and FVIII levels to help blood clot. Desmopressin does not work for everyone. A doctor needs to do tests to find out if an individual responds to the drug. Ideally, tests should be done before treatment is needed.

**Factor concentrates** are used when desmopressin is not effective or when there is a high risk of major bleeding. Factor concentrates contain VWF and FVIII. This is the preferred treatment for Type 3 VWD, most forms of Type 2 VWD, and for serious bleeding or major surgery in all types of VWD.

Bleeding in mucous membranes (inside the nose, mouth, intestines, or womb) can be controlled by drugs such as **tranexamic acid** (Cyklokapron), **aminocaproic acid** (Amicar), or by **fibrin glue**. However, these products are used to maintain a clot and do not actually help form a clot.

Hormone treatment, such as oral contraceptives, helps increase VWF and FVIII levels and control menstrual bleeding. If hormone treatment is not prescribed, antifibrinolytic agents may be effective for treating heavy menstruation.

These treatments may have side effects, so people with VWD should talk to their physician about possible side effects of treatment.

## Issues for girls and women with VWD

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Women with VWD tend to have more symptoms than men because of menstruation and childbirth. Girls may have especially heavy bleeding when they begin to menstruate. Women with VWD often have heavier and/or longer menstrual flow. This heavier menstrual flow can cause anemia (lower iron in red blood cells, resulting in weakness and fatigue). Women with VWD should be checked regularly for anemia.

A woman with VWD should see an obstetrician as soon as she suspects she is pregnant. The obstetrician should work with a bleeding disorders treatment centre to provide the best care during the pregnancy and childbirth. During pregnancy, women experience an increase in VWF and FVIII levels. This provides better protection from bleeding during delivery. However, after delivery, these clotting levels decrease and women with VWD may then develop bleeding.

Women with VWD entering menopause (end of menstruation, usually between the ages of 45 and 50) are at increased risk of unpredictable and heavy bleeding. It is important for a woman with VWD to maintain a strong relationship with her gynecologist as she approaches menopause.

## Important points for people living with VWD

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- Carry information about your disorder, the treatment that has been prescribed, and the name and telephone number of your physician or treatment centre. In emergencies, a medical bracelet or other identification notifies healthcare personnel of your bleeding disorder.
- Give schools information about VWD and how to handle situations that may arise. The most common problem encountered at school is a nose bleed.
- Register at a centre that specializes in diagnosis and treatment of bleeding disorders, as they are likely to offer the best standards of care and information.
- Check all medications with your doctor. Some over-the-counter medications should be avoided because they interfere with clotting.
- Exercise regularly to keep joints and muscles strong and stay in good health.
- When travelling, find the addresses and telephone numbers of the bleeding disorders treatment centres at your destination(s) and bring the information with you.

## Selected resources

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### **World Federation of Hemophilia**

**[www.wfh.org](http://www.wfh.org)**

- The Basic Science, Diagnosis, and Clinical Management of von Willebrand Disease
- Pregnancy in Women with Inherited Bleeding Disorders
- Gynecological Complications in Women with Bleeding Disorders
- Desmopressin (DDAVP) in the Treatment of Bleeding Disorders: The First 20 Years

### **Canadian Hemophilia Society**

**[www.hemophilia.ca](http://www.hemophilia.ca)**

- All About von Willebrand Disease
- Amicar and Cyklokapron, A Guide for Patients and Caregivers
- Desmopressin, A Guide for Patients and Caregivers

### **U.S. National Hemophilia Foundation**

**[www.hemophilia.org/resources/handi\\_pubs.htm](http://www.hemophilia.org/resources/handi_pubs.htm)**

- von Willebrand Disease: Just the FAQs
- A Guide for Women and Girls with Bleeding Disorders
- Project Red Flag – [www.projectredflag.org](http://www.projectredflag.org)

### **Association française des hémophiles**

**[www.afh.asso.fr](http://www.afh.asso.fr)**

- La maladie de Willebrand
- [www.orpha.net/data/patho/Pub/fr/Willebrand-FRfrPub3497.pdf](http://www.orpha.net/data/patho/Pub/fr/Willebrand-FRfrPub3497.pdf)

### **Schweizerische Hämophilie-Gesellschaft**

**[www.shg.ch](http://www.shg.ch)**

- Formes particulières d'hémophilie, la maladie de von Willebrand
- Informationen zur Hämophilie, Von Willebrand Krankheit

## Selected resources

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### **Haemophilia Foundation Australia**

**[www.haemophilia.org.au](http://www.haemophilia.org.au)**

- A Guide for People Living with von Willebrand Disorder
- Meeting von Willebrand Disorder for the First Time: A Guide for Parents
- Understanding von Willebrand Disorder: A Guide for Teachers

### **Angelo Bianchi Bonomi Haemophilia Thrombosis Centre**

- von Willebrand Disease: A Complex, Not Complicated Disorder (If Known)

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