





# Von Willebrand Disease...

the most common bleeding disorder

## Your Questions Answered

<p>Why do I bleed so much at the dentist?</p>		<p>How can I control my heavy periods?</p>
		
<p>Why do I bruise so easily?</p>		<p>Why do I get so many nosebleeds?</p>



This booklet has been adapted from the Canadian Hemophilia Society (CHS) publication, *All about von Willebrand Disease... for People with von Willebrand Disease and their Families* written by David Page.

The CHS would like to thank Dr. David Lillicrap, *Medical Director, Kingston/Belleville Regional Hemophilia Program, Kingston, ON*, for reviewing this document.

*The CHS consults qualified medical professionals before distributing any medical information. However, the CHS does not practice medicine and in no circumstances recommends particular treatments for specific individuals. In all cases, it is recommended that individuals consult a physician before pursuing any course of treatment.*

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The CHS would like to thank Aventis Behring for providing the funding for the publication of this booklet.



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published by the Canadian Hemophilia Society, October 2001

ISBN 0-920967-38-8

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## What is von Willebrand Disease?

Von Willebrand Disease (vWD) is the most common bleeding disorder that people have.

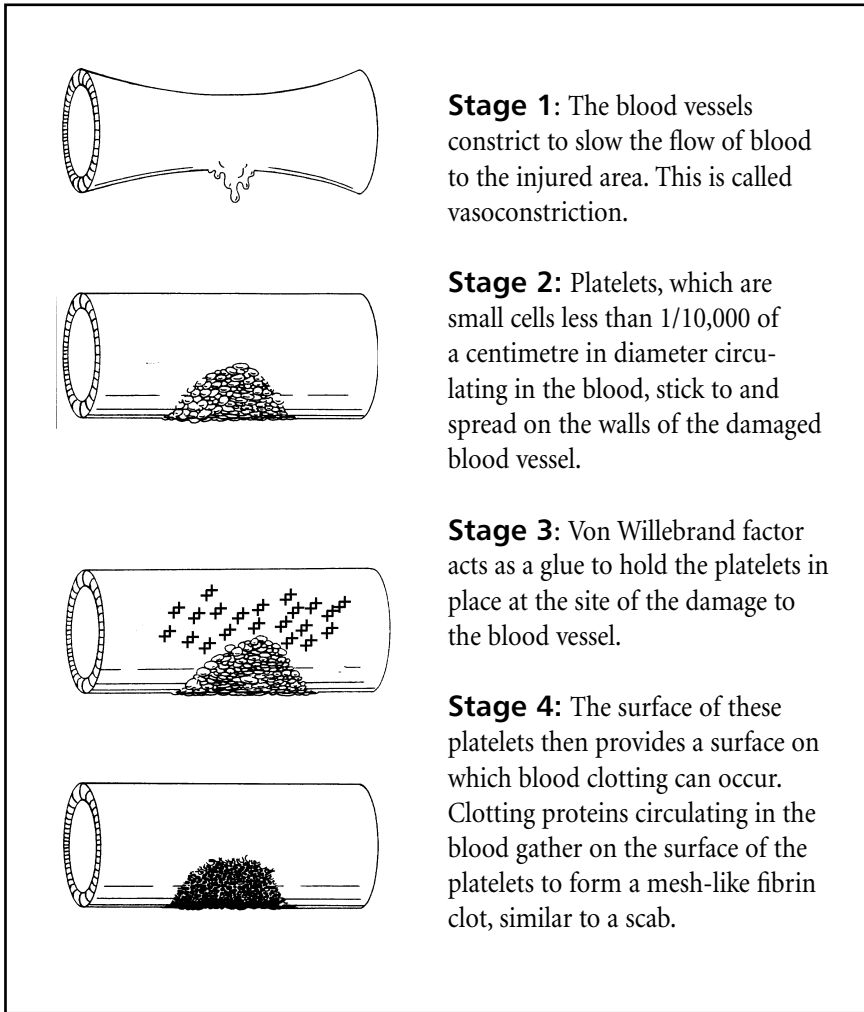
There are various types of vWD. (Type 1, Type 2, Type 3). All the different types are caused by a problem with the von Willebrand factor (vWF). This is a protein in blood which is necessary for proper blood coagulation, or clotting.

When there is not enough vWF in the blood, or when it does not work the way it should, the blood takes longer to clot.

## How does blood clot normally?

Blood is carried throughout the body within a network of blood vessels. When tissues are injured, damage to a blood vessel may result in leakage of blood through holes in the vessel wall. The vessels can break near the surface, as in a cut. Or they can break deep inside the body, making a bruise or an internal hemorrhage.

When a blood vessel is damaged, there are four stages in the normal formation of a clot. See Figure 1.



Normal von Willebrand factor (vWF)

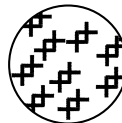


Figure 1



## How does vWD affect the normal clotting of blood?

Von Willebrand Disease affects the last three stages in the blood clotting process. See Figure 2.

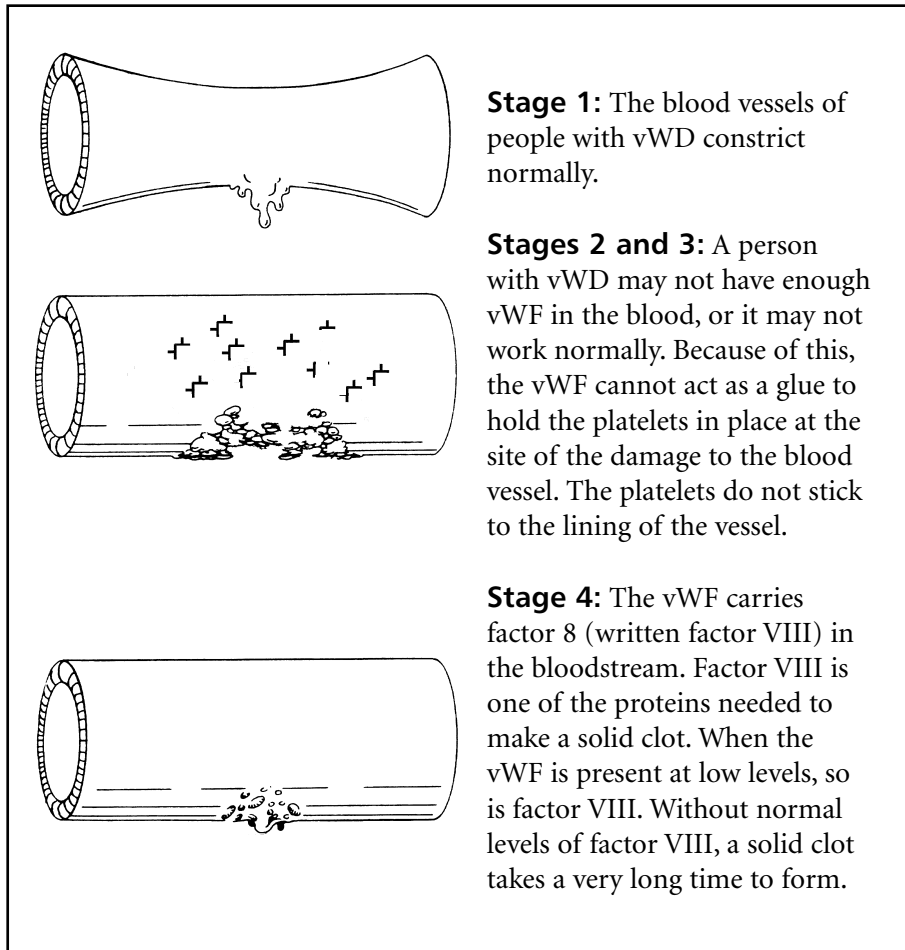
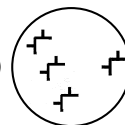


Figure 2

Abnormal von Willebrand factor (vWF)





## How common is von Willebrand Disease?

Doctors now think that vWD could affect as many as 1 in 100 people, or 300,000 Canadians. Because many of these people have only very mild symptoms, only a small number of them know they have the disease. Research has shown that as many as 99 out of 100 people with von Willebrand Disease have not been diagnosed.

A study reported that 1 woman out of 5 who went to see their doctors because of heavy, prolonged bleeding during their periods (menorrhagia) actually has a bleeding disorder.



## Who can have von Willebrand Disease?

Von Willebrand Disease affects both men and women. However, because vWD can cause heavy menstrual bleeding and prolonged bleeding after childbirth, more women than men have noticeable symptoms.

Children, too, can have vWD. They are born with it. This is because vWD is a hereditary disorder.



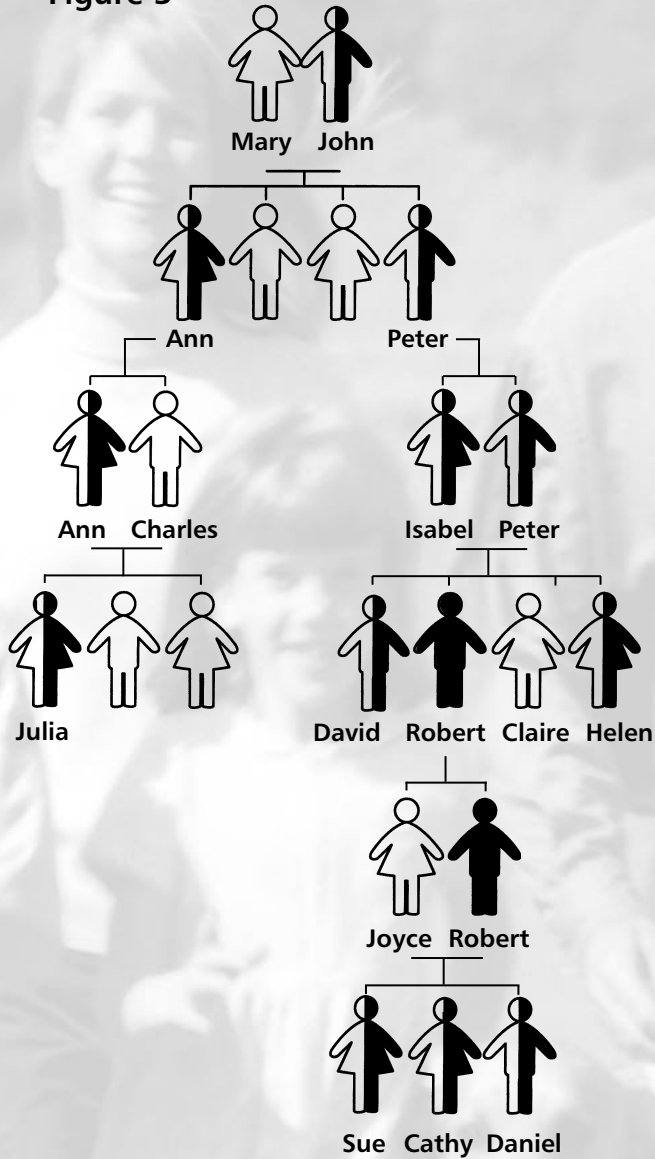
## How does a person get von Willebrand Disease?

vWD is a hereditary disorder. There are two ways of getting the hereditary form of vWD. See **Figure 3**.

- It can be passed from a parent who has the defective gene (even if this person has no symptoms of vWD) to a child at the time of conception. The parent is called a *carrier*.
- One of the baby's genes can undergo a change. This is called a genetic mutation. The baby's parents do not carry this defective gene, and the parents' other children would not inherit it.

Male and female children have an equal chance of inheriting vWD.

Figure 3



normal

mild von Willebrand disease

severe von Willebrand disease

## Is there always a history of bleeding in the family?

Not always. There are several reasons why.

- The level of vWF is not the same from person to person even in the same family. As a result, one person may bleed more than another.
- Blood type can play a role. People with Type O blood often have lower levels of vWF than people with Types A, B and AB. So people with Type O blood may have more problems with bleeding.
- Doctors believe there are other factors that also affect how severe symptoms are, but they are not yet well known.
- There may be no history of the disease because no one else in the family has it. The baby could have gotten the abnormal gene through a new mutation. In this case, chromosome 12 changes at conception or soon after. That baby's parents are not carriers. However, the baby, after growing up, could pass on vWD to its children.

## How serious is von Willebrand Disease?

It depends on the type of vWD. Most people have such mild cases that they never know they have the disease. Others only realize they have a bleeding problem after a serious accident or surgery. Some people with vWD bleed quite frequently.

## Is there a cure for von Willebrand Disease?

No, there isn't. It is a lifelong, most often mild, condition. Fortunately, there are safe, effective treatments for all types of vWD.



## What are the symptoms of von Willebrand Disease?

**Common symptoms of vWD are:**

- easy bruising
- heavy menstrual periods
- frequent or prolonged nosebleeds
- prolonged bleeding after injury, surgery, childbirth, or dental work.



## What other signs of a bleeding disorder are seen in women?

Heavy, prolonged menstrual bleeding is the most common symptom for women with vWD. Some women have heavy bleeding throughout the normal menstrual period. This is called menorrhagia.

Other women bleed unpredictably throughout the month. This is called metrorrhagia.

The following guidelines should alert a woman to a potential problem:

- flow which lasts longer than 7 days
- flow soaking one maxipad in less than 2 hours
- unpredictable bleeding
- menstrual bleeding which affects quality of life
- periods heavy enough to cause anemia.

In a woman with vWD, comparison to other family members can be misleading as they, too, may be affected by vWD.



## Are symptoms of von Willebrand Disease the same for everybody?

No. The symptoms of vWD vary greatly from person to person. Even members of the same family will have different symptoms.

Most people have such mild cases that they never know they have the disease. Others only realize they have a bleeding problem after a serious accident or surgery. Some people with vWD bleed quite frequently.

The type of vWD affects the severity of the symptoms.

- Type 1 vWD is the most common form, accounting for 75% of all cases of vWD. In Type 1 vWD, the von Willebrand factor (vWF) works normally, but there is not enough of it.
- Type 2 vWD is less common than Type 1. It represents 20-25% of all cases. In Type 2 vWD, the amount of vWF in people's blood is often normal. The problem is that the vWF does not work properly.
- Type 3 vWD is very rare. However, it is the most severe type of vWD. People with Type 3 vWD have very little vWF in their blood. As a result, bleeding can happen often and, if untreated, can be serious.



## Is diagnosing von Willebrand Disease easy?

No, diagnosing vWD is not easy. Many doctors are not familiar with it. As a result, vWD is often misdiagnosed. Diagnosing vWD is difficult even for an experienced doctor. This is because the results of a person's blood tests can vary from day to day. The test results can be normal, even when the person has vWD. There are a number of factors that cause the level of vWF to rise in the blood and appear to be normal. Some of them are:

- being pregnant
- breastfeeding
- normal hormonal changes during a woman's monthly menstrual cycle
- being on the birth control pill (oral contraceptive)
- having an infection
- having recently undergone surgery
- having recently had a blood transfusion
- doing a lot of exercise.



## Are routine blood tests a good way to diagnose von Willebrand Disease?

No, they are not. Routine blood tests will often give normal results in people who have von Willebrand Disease.

With this condition:

- the prothrombin time is always normal
- the partial thromboplastin time is often normal and
- the bleeding time is often normal.

In fact, the test for bleeding time is the least accurate of all tests used.



## **Where should I go to get tested for von Willebrand Disease?**

A person who thinks he/she has a bleeding problem should see a hematologist who specializes in bleeding disorders. Such a doctor can be found at a Hemophilia/Bleeding Disorder Comprehensive Care Program. Ask your family physician or gynecologist for a referral. See page 13 for more information.

## **Is medical treatment always necessary for bleeds?**

No. Minor bleeding episodes associated with vWD often do not require medical treatment. For example:

- small bruises usually disappear on their own
- larger bruises or minor bleeding into muscles or joints can often be controlled by applying cold (an ice pack wrapped in a towel) and elevating the limb
- bleeding from minor cuts can be stopped by applying pressure.
- nose bleeds may be stopped with simple first aid techniques

However, sometimes medical treatment is necessary.

## **Where is the best place for a person with von Willebrand Disease to get treatment?**

Few doctors are familiar with vWD. Even hematologists, who deal with diseases of the blood, are rarely experts in diagnosing and treating bleeding disorders. Many obstetricians and gynecologists remain unaware of the consequences of vWD for women.

Therefore, the best place for a person with vWD to get treatment is a Hemophilia/Bleeding Disorder Comprehensive Care Program.

Once a proper diagnosis has been made, and a treatment plan has been organized, the doctors at the Hemophilia/Bleeding Disorder Comprehensive Care Program can work with the individual and his/her family physician to provide care.



## What are the treatment options for von Willebrand Disease?

The type of treatment depends in part on the type of vWD a person has:

Treatment options include:

- Oral contraceptives (the Pill) raise the level of von Willebrand factor in the blood.
- Desmopressin acetate is a synthetic drug which is a copy of a natural hormone. Desmopressin can be given by:
  - intravenous infusion or injection under the skin. This medication is DDAVP® Injection or Octostim®. Note that DDAVP® Injection (4ug per 1 ml. ampoule) and Octostim® (15 ug per 1 ml. ampoule) are the same drug in different strengths.
  - nasal spray. This medication is Octostim® Nasal Spray.
- Tranexamic acid (Cyklokapron®) and aminocaproic acid (Amicar®) are drugs that help to hold a clot in place once it has formed.
- Thrombin and fibrin glue are removed from blood and manufactured as natural clotting agents that come in powder form. They can be applied directly to the site of bleeding.
- FVIII/vWF concentrate (Humate P®) is made from pooled human plasma. This concentrate replaces the missing vWF in the blood long enough to allow clotting to take place.

## For more information...

For more information please contact the Canadian Hemophilia Society. We're the largest national consumer organization working to ensure that, for people with bleeding disorders, the bleeding stops.

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# Notes

Canadian Hemophilia Society  
**We're all related by blood.**

