



All About von Willebrand Disease...

...for people with von Willebrand disease and their families



Canadian Hemophilia Society
Help Stop the Bleeding

Second Edition



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Preface

In recent years, our awareness of the challenges, concerns and misunderstandings faced by women, men, girls and boys with von Willebrand disease has grown considerably. In year 2000 the Canadian Hemophilia Society realized there was a need for a comprehensive, easy-to-understand book about living with this common bleeding disorder. With the generous involvement of the bleeding disorder community, both health care providers and consumers, we gathered and organized the most up-to-date information and advice available.

Now in 2007 we present to you—the children, adults, families and health care professionals whose lives are touched by von Willebrand disease—the second, revised edition.

It is our sincere wish that this book will inform and guide you in all aspects of living with von Willebrand disease. With your input, we will continue to learn, and to raise awareness and understanding within the bleeding disorders community and the general population. This book is only the beginning.



Canadian Hemophilia Society
Help Stop the Bleeding

Canadian Hemophilia Society Mission

The Canadian Hemophilia Society strives to improve the health and quality of life for all people with inherited bleeding disorders and to find a cure.

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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An introduction to von Willebrand disease

VWD

What is von Willebrand disease?

Von Willebrand disease (VWD) is the most common bleeding disorder that people have.

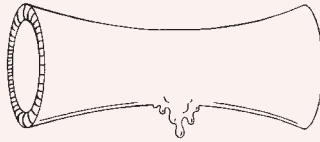
There are various types of VWD. (See “*Types of Von Willebrand Disease*” on page 7.) All the different types are caused by a problem with the von Willebrand factor (VWF). This is a protein in blood which is necessary for proper blood coagulation, or clotting.

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.

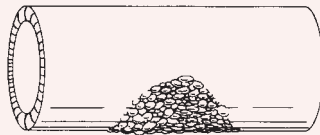
How does blood clot normally?

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.

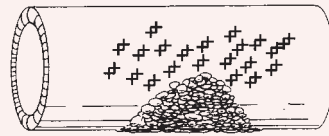
When a blood vessel is damaged, there are four stages in the normal formation of a clot. **See Figure 1.**



Stage 1: The blood vessels constrict to slow the flow of blood to the injured area. This is called vasoconstriction.



Stage 2: Platelets, which are small cells less than 1/10,000 of a centimetre in diameter circulating in the blood, stick to and spread on the walls of the damaged blood vessel.



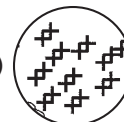
Stage 3: Von Willebrand factor acts as a glue to hold the platelets in place at the site of the damage to the blood vessel.



Stage 4: The surface of these platelets then provides a surface on which blood clotting can occur. Clotting proteins circulating in the blood gather on the surface of the platelets to form a mesh-like fibrin clot, similar to a scab.

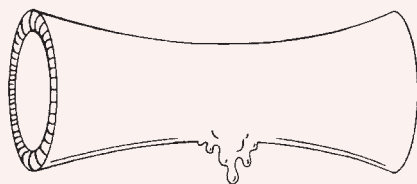
Figure 1.

Normal von Willebrand factor (VWF)

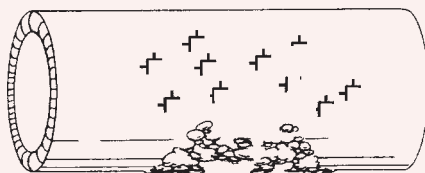


How does VWD affect the normal clotting of blood?

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



Stage 1: The blood vessels of people with VWD constrict normally.



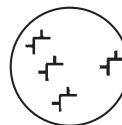
Stages 2 and 3: A person with VWD may not have enough VWF in the blood, or it may not work normally. Because of this, the VWF cannot act as a glue to hold the platelets in place at the site of the damage to the blood vessel. The platelets do not stick to the lining of the vessel.



Stage 4: The VWF carries factor 8 (written factor VIII) in the bloodstream. Factor VIII is one of the proteins needed to make a solid clot. When the VWF is present at low levels, so is factor VIII. Without normal levels of factor VIII, a solid clot takes a very long time to form.

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 1,000 people, or 30,000 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 9 out of 10 people with von Willebrand disease have not been diagnosed.

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.



Can VWD be passed from parents to children?

Yes. If one or both of the parents have VWD, they can pass it on to their children. (See “*Heredity*” on page 11.)

Why is it called von Willebrand disease?

It is named after the Finnish physician, Erik von Willebrand, who first described the condition in 1925. He realized that the disease was different from hemophilia, another important bleeding disorder, which mainly affects males.

How serious is von Willebrand disease?

It depends on the type of disease. 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?

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

Types of von Willebrand disease

What are the different types of von Willebrand disease?

Von Willebrand disease is divided into three categories - Type 1, Type 2 and Type 3. Type 2 VWD is itself divided into several sub-types.

What is Type 1 VWD?

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.

Many people with Type 1 VWD have no symptoms at all until they experience a bad injury or an operation. Then, they could have serious bleeding.

Others have mild symptoms such as:

- minor bruises
- bleeding from the nose and gums
- prolonged bleeding from cuts.

During their periods, some women with Type 1 VWD have heavy, abnormally long bleeding, called menorrhagia. This can have a major effect on health and quality of life.

Injuries and surgery can lead to severe bleeding even in this mild form of VWD. That's why it's important for people who think they might have VWD to get tested.



What is Type 2 von Willebrand disease?

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.

There are several sub-types of Type 2 VWD. It is important to get an exact diagnosis because the sub-types are treated differently.

Type 2A VWD is the most common sub-type. It represents 15-20% of all cases of VWD. In Type 2A VWD, the amount of VWF is often normal. However, because of a defect in the VWF protein, the platelets do not bind together well. The VWF does not act as a glue to hold the platelets in place to plug a hole in a blood vessel.

Type 2B VWD is the next most common. It represents about 5% of all cases of VWD. In Type 2B VWD, the VWF binds to platelets in the bloodstream, instead of binding at the site of the injury to the blood vessel. Then, the body removes these large bundles of platelets from circulation. This causes a shortage of platelets.

Type 2N VWD is much rarer. (The "N" stands for Normandy, France where the sub-type was first identified.) This is what doctors know about it.

- In Type 2N the VWF works normally with platelets. As a result, the grouping of platelets around the injury happens as it should.

- VWF also helps to carry around factor VIII in the blood and stabilize it so it can take part in the formation of a solid clot. In Type 2N the VWF does not transport factor VIII. As a result, factor VIII levels are low.
- Sometimes, because of the low factor VIII levels, Type 2N is mistaken for factor VIII deficiency hemophilia.
- In order for a child to get Type 2N, both parents must pass on the defective gene.

There are several other extremely rare sub-types of Type 2 VWD, including Type 2M. The “M” stands for “Multimer”, a part of the structure of the VWF molecule. In Type 2M, binding of the VWF to platelets is impaired.



What is Type 3 von Willebrand disease?

Type 3 VWD is very rare. It affects about 1 in 500,000 people. However, it is the most severe type of VWD. People with Type 3 VWD have very little VWF in their blood. Because VWF transports factor VIII, they also have very low levels of factor VIII. As a result, bleeding can happen often and, if untreated, can be serious.

Usually, in order for a baby to get Type 3 VWD, both parents must pass on the defective VWD gene. However, in some cases, the disease can result from a combination of one parent passing on the defective gene and a mutation in the child’s gene inherited from the other parent.

How does a person get von Willebrand disease?

Von Willebrand disease is a hereditary disorder. It is caused by an abnormal gene.

Each cell of the body contains structures called chromosomes. A chromosome is a long chain of chemicals known as DNA. This DNA is arranged into about 25,000 units called genes which determine such things as the colour of a person's eyes.

Each cell contains 46 of these chromosomes arranged in 23 pairs. Von Willebrand disease is caused by a defect on chromosome 12. The defect could be on the chromosome 12 from the mother, on the chromosome 12 from the father, or on both.

There are two ways of getting the hereditary form of VWD.

- 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.
- 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.

Because it is a hereditary disorder, VWD often affects several members of the same family. One can often trace VWD through a family tree. **Figure 3** shows three generations of a family with VWD.

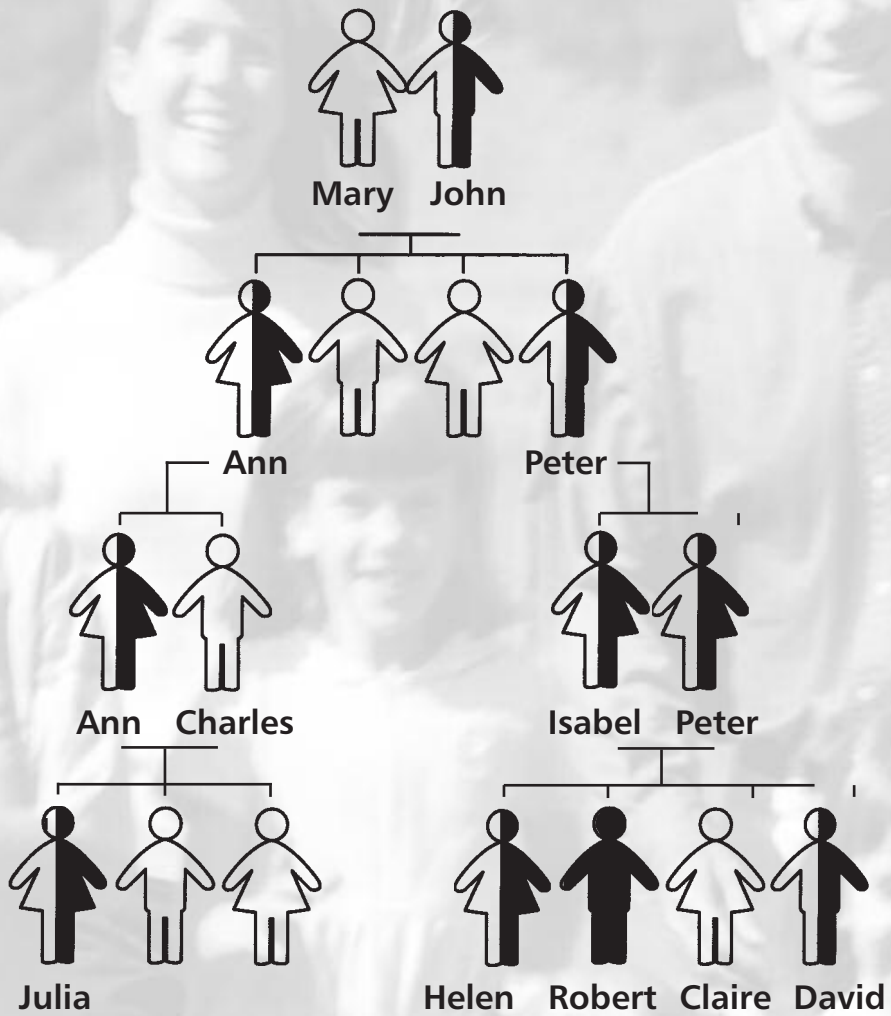
In the first generation: The grandfather (John) has mild Type 1 VWD and the grandmother (Mary) is unaffected by VWD.

In the second generation: There is a 50% chance that each of John and Mary's children will be born with the VWD gene. The diagram shows one of the two daughters (Ann) and one of the two sons (Peter) inheriting the defective gene. They both have mild Type 1 VWD.

In the third and fourth generations: The daughter with VWD (Ann) marries a man who does not have VWD (Charles). Their children have the same 50 % chance of inheriting the disease. In the diagram one daughter (Julia) gets VWD.

The son with VWD (Peter) marries a woman (Isabel) who also carries the abnormal chromosome 12. Their children have a 25% chance of being unaffected (Claire), a 50% chance of inheriting the defective gene from one of the parents and thus having mild Type 1 VWD (Helen and David), and a 25% chance of inheriting the defective gene from both parents (Robert) and getting severe Type 3 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 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.
- In very rare cases a person can develop VWD later in life. This is an acquired form of VWD. Neither of the person's parents carries the VWD gene, nor does the person affected. In acquired VWD, the body's immune system suddenly develops special antibodies, called inhibitors, to the VWF in the blood. In other words, the body sees the VWF as a foreign substance and then destroys it. This can happen as a result of medication a person has taken. Inhibitors can also be caused by other medical conditions, such as rheumatoid arthritis, certain types of kidney disease and certain cancers.

Do the parents of a child with Type 3 von Willebrand disease show signs of the disease?

In most cases, parents of a child with severe Type 3 VWD do not show symptoms. Occasionally one, or rarely both, will have mild bleeding problems.

Is von Willebrand disease always the result of a genetic condition?

No. As explained on page 14, in cases of acquired VWD, there is no defective VWD gene.

Is it normal for a parent to feel guilty about passing on von Willebrand disease to his/her child?

Some parents do feel guilty after their child has been diagnosed with VWD. Such feelings are not unusual. However, VWD is a genetic condition and no one's fault. What's more, children with VWD can live normal, healthy lives and realize their full potential.



□ What are the symptoms of von Willebrand disease?

Common symptoms of VWD are:

- easy bruising
- bleeding from nose and gums
- prolonged bleeding from skin lacerations
- bleeding from the gums when baby teeth fall out or after tooth extractions
- heavy or prolonged bleeding during menstruation, called menorrhagia.

However, many people with VWD do not notice anything is wrong. They realize they have a bleeding problem only when another person in the family is diagnosed with VWD or after they have a serious injury or surgery.

□ When are symptoms of von Willebrand disease first seen in children?

Symptoms of VWD can begin at any age. The signs are:

- bruises from minor bumps
- nose bleeds
- prolonged bleeding from minor cuts.





In cases of severe Type 3 VWD, bleeding can occur in newborns, especially from the umbilical cord and at the time of circumcision. These types of bleeding can also occur with Types 1 and 2 VWD, but only in rare cases.

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.

The type of VWD affects the severity of the symptoms, too.

- The symptoms of Type 1 VWD are usually very mild. However, it is possible for a person with Type 1 VWD to experience serious bleeding episodes.
- The symptoms of Type 2 VWD are moderate.
- The symptoms of Type 3 are more severe. People with Type 3 VWD can have bleeding into muscles and joints, sometimes without injury.

Symptoms of bleeding into a joint or muscle are:

- a feeling of tightness in the joint or muscle
- a puffiness to the touch
- heat at the site of the bleeding
- a limitation in movement of the joint
- increasing pain.

What's more, because of the menstrual cycle and childbirth, women tend to have more symptoms than men do.



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

Menorrhagia, metorrhagia

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 metorrhagia. If women lose enough blood over a long period, they suffer from iron deficiency anemia.

It can be hard for any woman to tell if her bleeding is unusually heavy compared to others. In a woman with VWD, comparison to other family members can be misleading as they, too, may be affected by VWD. Sisters, mothers, aunts and grandmothers often have the same problem. Nobody sees it as special or, if they do, they say, “All the women in our family bleed a lot during their periods.” 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.

Menstrual bleeding can be especially heavy at the time of a girl's first periods. For this reason, when there is a family history of a bleeding disorder, she should be closely followed through puberty. The medical team should include:

- a gynecologist
- a hematologist with experience in treating bleeding disorders and
- a family physician or pediatrician.

Dysmenorrhea and mid-cycle pain

Some women with VWD have pain during their menstrual periods. This is called dysmenorrhea. Dysmenorrhea is usually worse in women with heavy flow such as those with VWD. They can also have pain at mid-cycle at the moment of ovulation. With ovulation slight bleeding of the ovary is common. For women with VWD this bleeding can be significant and cause severe pain.

Some women can have a separate condition called endometriosis. With this condition endometrial tissue forms outside the uterus, for example, around the abdomen. When a woman menstruates, endometrial tissue, wherever it is in the body, bleeds. If these women also have VWD, the bleeding may be heavy. The blood can irritate the abdominal wall, causing pain. Typically, this pain starts a few days before the onset of vaginal bleeding.

How many women with menorrhagia actually have a bleeding disorder?

A study by Kadir et al (*The Lancet*, February 14, 1998) reported that 1 woman out of 5 who went to see doctors because of heavy, prolonged bleeding during their periods (menorrhagia) actually has a bleeding disorder. This means that menorrhagia caused by bleeding disorders is much more common than doctors thought in the past.

The most common bleeding disorder diagnosed in this study was von Willebrand disease.

Careful questioning by doctors showed that many of the women with bleeding disorders could report a family history of bleeding problems.

In the general population less than 10% of women have menorrhagia. However, Kadir et al (*Haemophilia*, May 1998) found that 73% of women with von Willebrand disease suffer from menorrhagia.

How does menopause affect a woman with von Willebrand disease?

After many years of battling heavy periods, one might expect women with VWD to look forward to menopause. In fact, hormone regulation becomes more chaotic as a woman enters menopause and this increases her risk of unpredictable and heavy bleeding. This is particularly true of a woman with VWD where the consequences of poor hormone regulation can be catastrophic. It is important that a woman with VWD maintain a strong relationship with her gynecologist even after childbearing as she enters her forties so as to anticipate the combined issues of menopause and VWD.



How does menorrhagia affect a woman's quality of life?

Menorrhagia seriously affects a woman's quality of life. Here are some examples. Women with menorrhagia caused by VWD may...

- limit the amount of time they work
- change careers as a result of bleeding problems
- be unable to work normally during their menstrual periods
- have lost faith in the medical profession after being told for years that their problems were not real
- suffer constant fatigue from iron deficiency anemia
- suffer from depression as a result of the strain of their bleeding disorder
- suffer pain during their menstrual periods and times of ovulation
- have increased episodes of ovarian cysts, pain, internal bleeding and risk of subsequent surgery with potential loss of the ovaries
- live with the embarrassment of staining due to heavy bleeding
- have had hysterectomies because doctors did not diagnose a bleeding disorder. This meant they had unnecessary surgery and could not have any more children.

Happily, with proper diagnosis and treatment, these problems can be dramatically reduced and even eliminated.



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.

For example:

- a person with a low platelet count because of Type 2B VWD could be misdiagnosed with leukemia
- a woman with heavy, prolonged menstrual bleeding because of VWD, who has not responded to hormone therapy, could be advised to have a hysterectomy.

For this reason, 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 Treatment Centre. There, a diagnosis will be made by a specialist who is familiar with bleeding disorders and who has experience doing the proper blood tests.

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.

VWF levels can change over time and tend to increase with age. In addition, people with blood group O have naturally lower levels of VWF. This, too, can make tests inconclusive.



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 almost always normal and
- the bleeding time is often normal.

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



What tests are reliable in diagnosing von Willebrand disease?

A doctor who suspects VWD will first take a complete family history. He/she will then do a number of blood tests to find out:

- if the person has VWD and
- what type of VWD the person has.

The tests may include ...

• Bleeding time	This measures the length of time it takes for a simple cut to stop bleeding.
• Factor VIII:C	This measures the amount of factor VIII clotting activity.
• VWF: antigen	This measures the amount of von Willebrand factor.
• Ristocetin cofactor activity	This measures how well the VWF works.
• VWF multimers	This examines the structure of the VWF.
• Platelet function tests	These measure how well the platelets work.



How long does it take to get the results?

Some of the tests must be done in a specialized laboratory. Therefore, it may be several weeks before test results come back.



If all the tests are normal the first time, should a person be re-tested?

Sometimes. This is because a person's test results can be normal one day and abnormal a month later.

Even if all the tests are normal the first time, and the doctor does not believe there is a bleeding disorder, a person who believes he or she has a bleeding problem should discuss the need for being re-tested with his/her physician.

If the health care institution does not have experience with von Willebrand disease, the person should ask to be referred to a hematologist experienced with bleeding disorders or to a Hemophilia/Bleeding Disorder Treatment Centre.

Unfortunately, many people in the past have accepted their doctors' diagnoses that all was normal. They had to live with the bleeding problems caused by VWD when they could have received proper treatment.

What reactions do people have when they are diagnosed with von Willebrand disease?

Many people feel relief that the problems they have experienced over many years have finally been diagnosed. With diagnosis comes the possibility of effective treatment.

Others feel worried or scared about being diagnosed with a lifelong condition, especially one with such an impressive name. Over time, however, these fears can change into feelings of empowerment as more is learned about the disease, and the person takes more control over it.

It is important to get support from others at this time. Medical people, friends and family members can help a person through this difficult period. Contact with people who are already living with VWD can be of special comfort in allaying fears. Get in touch with the local chapter of the Canadian Hemophilia Society for this kind of help.

Should a person inform other family members of the diagnosis?

Yes. Other family members may also have symptoms of VWD and may not have been diagnosed.





Treatment options

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 (See “*Nose Bleeds*” on page 50.)

However, sometimes medical treatment is necessary. The type of treatment depends in part on the type of VWD a person has.

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 Treatment Centre. (See “*Comprehensive Care*” on page 45.)

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

Recommended treatments for men and women



What are the treatment options for Type 1 von Willebrand disease?

DESMOPRESSIN

Desmopressin is a synthetic drug which is a copy of a natural hormone. It acts by releasing VWF stored in the lining of the blood vessels. Desmopressin is not made from blood.

Desmopressin is the preferred treatment for Type 1 VWD. It can be taken in three different ways.

- It can be injected into a vein. Most often, the brand name for this kind of desmopressin is DDAVP, Octostim or Stimate.
- It can be injected subcutaneously (under the skin). The brand name for this kind of desmopressin is also DDAVP, Octostim or Stimate.
- It can be taken by nasal spray. The brand name of the nasal spray is often Octostim Spray or Stimate Spray.

Desmopressin is effective for many people with Type 1 VWD. However, different people respond to desmopressin in different ways. Therefore, a doctor needs to do tests to find out each individual's response to the drug. Ideally, these tests are done before any urgent need for the drug, such as surgery.

Since desmopressin acts by releasing VWF stored in the body, a person cannot take it repeatedly over a short period. A sufficient amount of time, usually 24 hours, must elapse between doses of desmopressin to allow the body to rebuild its stores.

In major surgery, desmopressin alone may not be enough to control bleeding. In such a case, a person should also receive a concentrate of VWF and factor VIII. (See *Factor VIII/von Willebrand Factor Concentrates* on page 35.)

Desmopressin can sometimes have some mild side effects. These are:

- facial flushing
- mild headache
- nausea and abdominal cramps.

Desmopressin is an anti-diuretic; that is, it can make the body retain water. Therefore, doctors recommend that after receiving desmopressin people drink only enough fluid to satisfy thirst.

In very rare cases, desmopressin can cause more serious side effects. If a person has a very bad headache or has not been able to urinate 24 hours after taking desmopressin, he/she should go to the Hemophilia/Bleeding Disorder Treatment Centre or emergency room for help. In infants, fluid overload caused by desmopressin can cause seizures.

In the elderly and in people with cardiovascular disease, desmopressin can cause more serious side effects and may not be recommended.

CYKLOKAPRON AND AMICAR

Cyklokapron (tranexamic acid) and Amicar (aminocaproic acid) are drugs that help to hold a clot in place once it has formed. They act by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

They do not help to actually form a clot. This means they can not be used instead of desmopressin or factor VIII/VWF concentrate.

They can be used to hold a clot in place in mucous membranes such as:

- the inside of the mouth
- the inside of the nose
- inside the intestines (the gut)
- inside the uterus (the womb).

Cyklokapron and Amicar have proven very useful for people with VWD. They are used:

- before dental work
- when a person has mouth, nose and minor intestinal bleeding
- for women with heavy, prolonged menstrual bleeding.

Cyklokapron comes in tablet form. Amicar comes in tablet, liquid and injectable form. They can sometimes have some mild side effects. These are:

- feeling sick to the stomach (nausea)
- feeling tired or sleepy
- feeling dizzy
- having loose bowel movements (diarrhea)
- having pain in the stomach.

These mild side effects go away when:

- a person stops taking the drugs
- the doctor reduces the dosage.

A person with urinary tract bleeding (blood in the urine) should not take these drugs.

FIBRIN GLUE

In the coagulation process, the final clot is made up of fibrin. A product known as fibrin glue is removed from blood and manufactured as a natural clotting agent. It can be applied directly to the site of bleeding. It is especially useful in tooth extractions and surgery. Over a period of 2 to 4 weeks, as healing progresses, the fibrin is absorbed by the body.



What are the treatment options for Types 2 and 3 von Willebrand disease?

FACTOR VIII/VON WILLEBRAND FACTOR CONCENTRATE

Factor VIII/von Willebrand factor concentrate (FVIII/VWF concentrate) is the preferred treatment for:

- Type 3 VWD
- most forms of Type 2 VWD
- for serious bleeding or major surgery in all types of VWD.

This concentrate replaces the missing VWF in the blood long enough to allow clotting to take place.

FVIII/VWF concentrate is made from pooled human plasma. Plasma is a pale yellow liquid contained in blood. The plasma has been fractionated to extract the factor VIII and von Willebrand factor. This means that the different parts of the blood have been separated from each other so that each person can receive only that part which he/she needs.

The plasma used for FVIII/VWF concentrate is screened for blood-borne viruses such as HIV and hepatitis B and C. Any plasma found to contain these viruses is not used. Products made from the remaining plasma are then treated to destroy any viruses that may still be present. Factor concentrates used today have an excellent safety record.

Humate P is the brand name most commonly used in Canada. Alphanate is another factor VIII concentrate containing VWF. However, the amount of VWF in this product is variable.

FVIII/VWF concentrate is injected into a vein. It can be administered at a clinic, doctor's office or emergency room. Many people learn to inject it themselves at home. (See "*What is home care?*" on page 47.)

Other factor VIII concentrates such as monoclonal factor VIII and recombinant factor VIII contain no von Willebrand factor. Therefore, they are of no value in treating VWD.

CYKLOKAPRON AND AMICAR

Bleeding in mucous membranes can sometimes be treated with Fibrin, Cyklokapron and Amicar. (See the treatments for Type 1 VWD.)

DESMOPRESSIN

Desmopressin is not often recommended for Type 2 VWD. In most forms of Type 2 VWD, the von Willebrand factor does not work properly. As desmopressin works by releasing stored-up von Willebrand factor, it would only succeed in releasing a larger amount of a protein that does not work.

With Type 2B VWD, desmopressin could make the clumping together of platelets even worse than it already is. This would lower the platelet count or even cause unwanted clots in the bloodstream. Formation of an effective clot would be even more difficult.

However, some individuals with Type 2A VWD have a good response to desmopressin. Therefore, it is recommended that, at the time of diagnosis, tests be done to measure a person's response to desmopressin.

Nor is desmopressin recommended in the treatment of Type 3 VWD. With this type of VWD, very little VWF is stored in the blood vessel linings so desmopressin would not be effective in releasing stored-up VWF.



Medical Options for People with von Willebrand Disease

DISORDER	PREFERRED TREATMENT	ALTERNATIVE
Type 1	Desmopressin	FVIII/VWF conc.
Type 2A	Desmopressin, if individual responds	FVIII/VWF conc.
Type 2B	FVIII/VWF concentrate	
Type 3	FVIII/VWF concentrate	

N.B. Oral contraceptives, other hormone therapy and antifibrinolytic drugs may also be useful for women with menorrhagia, in particular circumstances.

Not recommended treatments

CRYOPRECIPITATE

Cryoprecipitate is a blood component made from plasma, containing VWF and factor VIII. It was commonly used to treat VWD in the past. However, because there is no method to kill viruses in cryoprecipitate, it is no longer recommended.

Currently, techniques are being developed to treat cryoprecipitate for viruses. When they are proven effective, cryoprecipitate may once again be used as a treatment for VWD.

Recommended treatments for women with gynecological complications



What are the medical treatment options for girls and women with gynecological complications?

HORMONE THERAPY

Oral contraceptives (the Pill) raise the level of von Willebrand factor in the blood for women with Type 1 VWD. For many women with VWD who suffer from menorrhagia, this hormone therapy alone is effective in reducing bleeding to normal.

This hormone therapy will not improve factor levels for women with:

- Type 2 VWD, because this is a problem in the way the VWF works, rather than the quantity
- Type 3 VWD, because there is very little VWF in the body to start with.

However, oral contraceptives can be helpful even for these women. They regulate the menstrual periods and reduce the flow of blood.

Other hormone therapies may be prescribed when oral contraceptives do not work well. These include a GnRH analogue to shut down the hormones of the ovary at the level of the brain. High dose progesterone derivatives can also be used in certain situations but their effectiveness in controlling uterine blood loss is somewhat unpredictable.

The use of the Pill for adolescent girls raises some difficult issues.



- Some parents may not want their daughter to take the Pill, fearing it will lead to early sexual activity. However, many studies have shown that therapeutic use of the Pill is not linked to early onset of sexual activity. This is a very effective means of controlling bleeding from VWD and should not be dismissed for such unfounded reasons.
- The adolescent girl may hesitate to go against her parents' wishes, even when the Pill may be the only effective way to control her menorrhagia.
- Parents and adolescents also fear use of the Pill may lead to cancer, infertility, blood clots or stroke. These concerns can be lessened with accurate information.

The personnel at the Hemophilia/Bleeding Disorder Treatment Centre can help families work through these issues.

ANTI-FIBRINOLYTIC AGENTS (Cyklokapron and Amicar)

For women with VWD and menorrhagia, Cyklokapron and Amicar can be started on the first day of menstrual bleeding and taken for 5 days in a row. They can even be combined with the use of oral contraceptives for women who do not respond to desmopressin.



What are the surgical treatment options for menorrhagia?

For some women, medical treatments alone will not work. Heavy, prolonged bleeding during the menstrual cycle will continue. For these women, surgery is an option. However, this is a big step. Women should have all the important information before making their decisions.

These are some of the options.

ENDOMETRIAL ABLATION (uterine ablation)

The purpose of this operation is to destroy the lining of the uterus. This is the endometrial tissue that bleeds so much during menstruation. The uterine lining is burned away. Hormone therapy is given for two months before the operation to reduce endometrial growth.

Advantages

- The operation is done through the vagina so no surgical cutting is needed. There is much less chance of bleeding than with a hysterectomy.
- The operation can be done in a doctor's clinic. Therefore, the woman does not have to stay in hospital.
- The recovery time is much shorter than with a hysterectomy.
- The success rates are high.

Disadvantages

- Unlike with medical treatment, the woman can no longer have children.
- The operation may have to be repeated.
- In a small percentage of women, this operation does not reduce bleeding.

HYSTERECTOMY (removal of the uterus)

The purpose of this operation is to remove the uterus so that menstrual bleeding stops once and for all. Sometimes, the ovaries and the fallopian tubes are removed as well.

Unfortunately, this operation is sometimes recommended to women with menorrhagia even before testing for von Willebrand disease or other bleeding disorders. This means that some women lose the ability to have children, when their bleeding could be successfully treated.

Advantages

- Hysterectomy stops menstrual bleeding once and for all.
- It may be the only option for women who do not respond to medical treatment, and for whom endometrial ablation is not effective.

Disadvantages

- Unlike with medical treatment, the woman can no longer have children.
- A hysterectomy is a major operation. In women with bleeding disorders, there is increased risk of bleeding both during and after the operation. This can be managed with FVIII/VWF concentrates.
- The operation requires a stay in hospital.
- The recovery time is much longer than with an endometrial ablation.
- When a woman undergoes a full hysterectomy (removal of the uterus and the ovaries), long-term hormone therapy is required.

LAPAROSCOPIC SURGERY (to remove endometrial tissue outside the uterus)

The purpose of this operation is to remove endometrial tissue that has formed outside the uterus. This tissue bleeds during menstruation. The bleeding can cause pain in the pelvis and abdomen. Two small incisions are cut in the abdomen. Two tubes are inserted – one for a tiny camera, the other for the instruments to cut out the endometrial tissue.

Advantages

- This operation can reduce pain and bleeding in the woman who does not respond to hormone therapy or other medical treatment.

Disadvantages

- While not a major operation, a woman with a bleeding disorder will need appropriate preparation with FVIII/VWF concentrates.

OOPHORECTOMY (Removal of the ovaries)

The purpose of this operation is to stop bleeding from the ovaries. This bleeding may happen even though:

- a woman is having hormone therapy to reduce menorrhagia
- an endometrial ablation has been done or
- a partial hysterectomy has been done.

Advantages

- It can reduce bleeding and pain.

Disadvantages

- An oophorectomy is a major operation.
- Women can no longer have children.
- Women need to take hormones.

DILATION AND CURETTAGE (D&C)

The purpose of this operation is to scrape and clean the lining of the uterus. This may need to be done to diagnose another problem; however, for women with menorrhagia, it will not be effective in reducing bleeding. In fact, the opposite is probably true. The D&C will remove any existing platelet plugs and fibrin clots and make the bleeding worse.

Living with von Willebrand disease

Comprehensive care

What is comprehensive care?

Comprehensive care is: all of the medical services needed by a person with VWD and his/her family for the treatment of VWD and related conditions.

This care is provided at a Hemophilia/Bleeding Disorder Treatment Centre.

This is a place where a person with VWD can receive all the care he/she needs at one time. It is called comprehensive care because it offers a complete range of services. The following people work there:

• **the Medical Director. He/she is often a hematologist who specializes in the area of blood clotting. He/she:**

- prescribes the lab tests to find out the exact bleeding problem
- prescribes the proper treatment to control and prevent bleeding
- monitors the overall health of the person with VWD.



- **the Nurse Coordinator. He/she is the front-line person in the clinic. He/she:**
 - helps families deal with the day-to-day problems related to VWD
 - answers families' questions over the phone or at the clinic
 - provides out-patient care at the clinic
 - teaches families how to do home therapy (See “*What is home care?*” page 47)
 - organizes the delivery of blood products for home use
 - coordinates appointments with other members of the comprehensive care team.

- **the Obstetrician/Gynecologist. He/she:**
 - works with the hematologist to prevent and control menorrhagia
 - helps the woman with VWD to avoid complications during pregnancy and childbirth.

- **the Dentist. He/she:**
 - provides dental care
 - works closely with the hematologist to prevent bleeding during dental work.

- **the Physiotherapist (especially for people with Type 3 VWD). He/she:**
 - checks the person's joints and muscles to make sure joint movement is not lost and that muscles remain strong
 - helps the person to regain lost joint function or to rebuild muscles through exercise
 - helps the person to find a sports and exercise program to keep in top shape.

The comprehensive care team will add other individuals as needed. These include, among others:

- a social worker (to help families deal with the stresses of a bleeding disorder)
- a genetic counselor (to give information to carriers)
- an orthopedic specialist (for joint problems and joint surgery, especially for people with Type 3 VWD).



What is home care?

Home care is the infusion of factor concentrates or desmopressin at home. This is most helpful for people who need treatment often. A decision to go on home care will be made in consultation with the comprehensive care program team. Questions that need to be asked are:

- Is treatment needed often enough to justify home care?
- Is the person with VWD or his/her caregiver willing to take on the responsibility?
- Is the person with VWD or his/her caregiver able to learn to do the intravenous infusions?

Home care has major advantages over treatment at the comprehensive care clinic or at the emergency room. These are:

- quicker treatment when a bleed starts
- a more normal life for the person with VWD and other members of the family
- a greater acceptance of treatment by the young child
- the ability for the person to take care of his/her own health.

Families of children with Type 3 VWD will need to be trained by the comprehensive care program team in how to recognize joint bleeds and then how to infuse the FVIII/VWF concentrate or desmopressin. Children often

learn how to infuse themselves at the age of eight or ten. Then, the person with VWD is able to treat himself/herself at home, at school, at camp or on vacation.

Desmopressin, either as an injection or as a nasal spray, can also be administered at home. However, it has a limited shelf life. People using it at home should carefully check expiry dates.

Cyklokapron and Amicar are drugs that can also be taken at home on a doctor's prescription.

People on home care go to the comprehensive care program once or twice a year for a complete check-up.

Safety of blood products



In the past, HIV and hepatitis C were transmitted through blood products. Are blood products safe now?

The blood products recommended to treat VWD today have never been known to transmit HIV, which causes AIDS, or hepatitis B or C. Four different safety measures are used to make VWF concentrate as safe as possible.

- Blood donors go through a rigorous screening process.
- Each blood donation undergoes a series of tests to detect HIV, hepatitis A, B and C and other viruses which could be present in blood. If any virus is found, the blood donation is not used.
- The plasma from which VWF concentrate is made is again tested for the presence of these viruses.

- During manufacturing, the VWF concentrate is treated to kill any viruses that may have escaped detection.

Despite these safety measures, factor concentrates made from plasma may transmit parvovirus. Parvovirus is a common virus that is not normally dangerous. However, it can, in rare cases, cause miscarriage (spontaneous abortion). (See “*Conception, Pregnancy and Childbirth*” on page 52.)

The risk of transmitting blood-borne diseases, especially unknown ones, still exists. For this reason, scientists are currently developing VWF concentrates made through recombinant technology, without human plasma. These new products are likely to be available within the next few years.



Are there any vaccines a person with von Willebrand disease should take?

Yes, there are. Hepatitis B can still be transmitted by certain blood products, such as plasma and red cells. The vaccine against hepatitis B is recommended for all people who routinely receive blood or blood products.

In very rare cases, hepatitis A has been transmitted by blood products. Therefore, doctors recommend vaccination against hepatitis A for people who receive blood or blood products. This is especially important for people who are infected with hepatitis C. This is because hepatitis A can be a serious, even fatal, disease for people who have hepatitis C.



If a person used blood products in the past, should he/she be tested for HIV and hepatitis C?

Yes. If a person received blood or blood products in the past, he/she should be tested for HIV and hepatitis B and C.

A Hemophilia/Bleeding Disorder Treatment Centre can provide counseling before doing the tests. Anonymous testing, which keeps the results confidential, is available.

Nose bleeds



Are nose bleeds common in von Willebrand disease?

Yes, nose bleeds are the most common symptom of VWD. This is especially true with children.



How can nose bleeds be stopped?

Nose bleeds may be stopped by sitting upright and firmly pinching the widest part of the nostrils together for 10 to 15 minutes. This applies direct pressure to the septum, the cartilage that divides the left and right nostrils. This is the most common site for bleeding. It may be necessary to repeat the procedure a second time. If, after two attempts, the bleeding persists, other treatments may be necessary. The nose can be packed or desmopressin can be taken.

Children should be taught to calm down as much as possible, in the event of a nose bleed.

Some people find that a cold cloth placed on the back of the neck and on the bridge of the nose is helpful in stopping bleeding.

Anti-fibrinolytic agents (Cyklokapron and Amicar) can be given for 5 to 7 days after the nose bleed to prevent re-bleeding.

Drinking hot liquids and strenuous exercise can cause the nose bleed to re-start. Therefore, it is helpful to avoid hot soup, tea or coffee and avoid lifting or straining for 24 hours after a nose bleed.



When should a doctor be consulted for a nose bleed?

If the pinching procedure does not stop the bleeding, and severe bleeding continues for more than 20 to 30 minutes, a doctor should be consulted.



Can anything be done to prevent nose bleeds?

Yes, there are several easy ways to prevent or reduce the frequency of nose bleeds.

It is important to maintain a certain level of humidity in the house, especially in the person's bedroom. This is especially important in the winter when heating makes a house much drier. A humidifier is ideal; however, an open bowl of water can also work very well.

Using petroleum jelly (Vaseline) in the nostrils every day can keep the nostrils from drying and cracking.

In some cases, local clotting agents like Fibrin may be needed to prevent bleeding from re-occurring. The personnel at the Hemophilia/Bleeding Disorder Treatment Centre will be able to help with these treatments.

Cauterizing (burning) the blood vessels in the nose is not usually recommended for people with VWD. A scab forms where the blood vessels are burned. This scab eventually falls off and bleeding can start all over again.

Conception, pregnancy and childbirth



Can women with VWD have children?

Yes. For most women with VWD, conception is not a problem. Of course, hormone therapy to control menorrhagia interferes with conception.

For many women with VWD, pregnancy is a time of few bleeding problems. Fortunately, the level of VWF in the blood for women with Type 1 VWD goes up during pregnancy and at the time of childbirth. However, after childbirth, VWF levels fall quickly and bleeding can continue for many weeks. Treatment may be required to prevent it.

Breastfeeding can help to keep VWF levels raised after childbirth in women with Type 1 VWD.

Women with Types 2 and 3 VWD can also have children. However, more precautions may be necessary.

What kinds of doctors should care for a woman with von Willebrand disease during her pregnancy?

It is very important for a woman with VWD to have confidence in a team of physicians to help her through her pregnancy. This team should include:

- an obstetrician (who specializes in caring for a woman during pregnancy and at delivery)
- a hematologist (who can make sure the woman's bleeding problem is under control).

The anesthesiologist who will be present at the birth needs to know the special needs of a woman with a bleeding disorder. He/she should check with the hematologist before performing invasive procedures like epidural anesthesia.

What problems might arise in the first trimester (the first 3 months of pregnancy)?

Women with Type 3 von Willebrand disease seem to have more frequent miscarriages (spontaneous abortions), especially during the first trimester. It may also be that these miscarriages, rather than being more frequent, are simply more noticeable because they are accompanied by heavier bleeding.

The risk of miscarriage can be lowered with factor replacement therapy. However, this benefit must be weighed against the risk of being exposed to parvovirus through factor concentrates. Parvovirus can, in rare cases, cause miscarriage. (See below.)

In addition, bleeding after a miscarriage may be severe for a woman with VWD.



Are certain blood products dangerous for the fetus?

Yes. Plasma-derived factor concentrates can transmit parvovirus. This common virus is not normally dangerous to people. This is because most people have already been exposed to it. However, in rare cases, it can cause miscarriage (spontaneous abortion). Therefore, pregnant women, or women who might become pregnant, should raise this question with their doctors.



Are women with von Willebrand disease more at risk for bleeding during pregnancy than at other times?

No. In fact, for women with Type 1 VWD the opposite is true. They have less bleeding than they normally do. This is because high hormone levels during pregnancy stimulate the production of blood clotting proteins. As a result, levels of von Willebrand factor and factor VIII rise closer to normal. Most women with Type 1 VWD have few bleeding problems during pregnancy, or during childbirth.

Nevertheless, clotting levels should be monitored, especially as the date of delivery approaches. This way, doctors will know whether to prepare treatments. Some of the clotting factors needed by women with bleeding disorders are rarely used. The hospital blood bank will have to be alerted so that the factor concentrates are on hand if needed.

Desmopressin can also be prescribed to raise VWF and factor VIII levels during delivery for women with Type 1 VWD.

The levels of VWF will not rise for women with Type 3 VWD because they do not make any VWF. In Type 2 VWD, the levels of VWF will rise during pregnancy. However, because the structure of this increased VWF is still not normal, the bleeding disorder will not be corrected.



What special precautions need to be taken during delivery?

If tests have shown that a woman is likely to suffer from bleeding during or after delivery, preventive treatments should be given. These include:

- desmopressin, if effective
- anti-fibrinolytics (Cyklokapron and Amicar)
- FVIII/VWF concentrates.

It should be assumed, unless prenatal testing has shown the opposite, that the fetus is also affected by a bleeding disorder. As a result, delivery should be as gentle as possible for both the woman and the baby. Natural childbirth without the use of instruments is the goal for a woman with a bleeding disorder.

In order to prevent bleeding, the following should be avoided, when possible:

- suction extraction of the baby
- deep intramuscular injections
- episiotomy (cutting of the skin near the vagina to avoid tearing)
- the use of forceps
- scalp electrodes.

An epidural (freezing of the lower body by means of a needle in the spine) may cause bleeding in women with Types 2 and 3 VWD. Before using epidural anesthesia, the anesthesiologist should consult an experienced hematologist. Treatment with FVIII/VWF concentrate may be necessary.

If a Caesarean section is necessary, prior treatment with FVIII/VWF concentrate is necessary.



What should be done after delivery (post partum) to prevent bleeding in the mother and the baby?

Post partum bleeding in women with VWD is more common than in the general population. Therefore, all women should be watched carefully for bleeding in the hours, days and weeks following delivery. The following blood tests need to be done:

- VWF and factor VIII levels
- hemoglobin (iron level in the blood).

If a woman feels her bleeding is excessive, she should immediately notify her obstetrician or hematologist.

Women with Type 1 VWD who breastfeed keep the high hormone levels they had during pregnancy. This may protect them from bleeding in the weeks following delivery (post partum). Women who do not breastfeed see their hormone levels fall. This can lower the levels of clotting factors. They can have bleeding problems in the weeks after giving birth.

Transfusions of factor concentrates, and even red blood cells, may be necessary.

Babies with VWD rarely bleed at birth. Babies with Type 3 VWD, however, may bleed if they undergo surgery, including circumcision.



Is pre-natal testing of the fetus necessary?

No, not usually. Because the symptoms of VWD can be so easily managed, pre-natal testing of the fetus is not recommended. However, if an older child has already been diagnosed with Type 3 VWD, parents may choose pre-natal testing. This can be done starting 10 weeks into the pregnancy.



If von Willebrand disease is suspected, should the newborn be tested immediately?

The diagnosis of VWD in a newborn can be made starting one week after birth. However, there is no danger of bleeding unless the baby needs to undergo surgery. Therefore, most doctors prefer to wait until the child is older – 4-5 years of age – before testing.

Medication to be avoided



Are there any drugs a person with von Willebrand disease should not take?

Yes. Certain drugs affect the way platelets plug holes in blood vessels. A person with VWD should never take drugs containing:

- aspirin (ASA) and other drugs containing aspirin (Alka-Seltzer, Anacin, Aspirin, Bufferin, Dristan, Midol, 222, to name just a few)
- non-steroidal anti-inflammatory drugs (indomethacin, ibuprofen and naproxen), unless prescribed by a physician knowledgeable about VWD
- blood thinners such as warfarin or heparin.

Drugs containing acetaminophen (Tylenol) can be used for fever, headaches and minor aches and pains. However, large doses beyond the levels prescribed on the label should be strictly avoided. They can cause damage to the liver, especially in people who are hepatitis C – positive.

A new generation of anti-inflammatory drugs, called Cox-2 inhibitors, is now on the market. These drugs do not affect blood clotting, and seem to cause less irritation to the stomach. Celebrex is one of the brand names.

Exercise, fitness and sports



Can a person with von Willebrand disease play sports?

Yes. It is important that all people with VWD engage in regular exercise to keep their muscles and joints strong and their health good. Being in good physical condition can actually reduce the number of bleeding episodes a person has, especially in Type 3 VWD.

Another benefit of regular exercise is the raising of the VWF levels.

A person with VWD will have to find out for himself/herself what physical activities he/she can or cannot do. Many people with a mild disorder participate in all kinds of sports including active sports like soccer and high-risk sports like skiing. People with Type 3 VWD may find these activities lead to serious bleeding.

Children, especially, need to be given the chance to discover what activities they can safely do. It is very important for a child's development to participate in the same sports as his/her friends. It is natural, of course, for parents to want to protect their children from harm. With VWD, the best way to protect children is to make sure they follow the latest safety guidelines for all children involved

in sports: for example, helmets for bicycling, rollerblading, skiing and snowboarding; shinpads and helmet for soccer; and full mask for hockey.



Specialists at the Hemophilia/Bleeding Disorder Treatment Centre can advise a person of the risks based on an evaluation of his/her condition. However, in the end, the person living with VWD is the best judge of which activities are appropriate.

Child care and schooling



Will von Willebrand disease affect a child's ability to attend daycare or school?

No, it shouldn't. Bleeding in Types 1 and 2 VWD should not keep a child out of school. Occasionally, with Type 3 VWD, a serious bleed into a muscle or joint could keep a child out of school for a short time. However, with prompt treatment, these absences should last no more than a day or two.



What should daycare and school personnel be told about a child with von Willebrand disease?

It is important to give daycare and school personnel the facts about VWD, but not to over-dramatize the disease. Many nurses in Hemophilia Treatment Centres are willing to talk to daycare and school personnel. This can help to reassure teachers and daycare workers that VWD can be easily managed.

The most common problem encountered at school will probably be nose bleeds. Children with VWD need to be taught at a young age how to handle their own nose bleeds. If the children are very young, the child's teacher or daycare worker will need to learn how to handle them. (See "Nose Bleeds" on page 50.) As with any blood spill, universal infection control precautions should be practiced.

Some parents provide the school with copies of documentation on VWD that can be placed in the child's file and follow him/her from grade to grade.

It is important that daycare or school personnel can contact the parents at all times, in case of emergency. In addition, it is helpful to provide the telephone number of the nearest Hemophilia/Bleeding Disorder Treatment Centre.



Should a child with von Willebrand disease take part in physical education?

Yes. Almost all children with VWD can do the same physical activities as other children. Occasionally, children with Type 3 VWD will be unable to do certain activities or play certain sports. Children with a bleeding disorder know what they can and cannot do on a given day. The gym teacher should let the child choose those activities he/she cannot do. If the gym teacher is overly cautious or, on the other hand, refuses to let the child sit out certain activities, a meeting should be arranged to discuss the issue.



Do the child's classmates need to know about his/her condition?

No, unless the child wants to educate his/her classmates about VWD, there is no purpose in informing classmates. After all, a child with VWD just wants to be treated normally.



What do other people need to be told about a child's von Willebrand disease?

There is no single answer to this question. It will depend on a number of factors, including:

- the child's age
- the severity of the symptoms
- the relationship of the person to the child.



In general, people taking responsibility for the child (babysitters, sports coaches, etc.) need to know what to do in the case of bleeding. If bleeding symptoms are extremely rare, or the child is old enough to take care of any problems himself/herself, informing may not be necessary.

Parents have occasionally been suspected of child abuse when their children with bleeding disorders have been discovered with bad bruises. Being open in talking matter-of-factly about VWD with babysitters, neighbours and others may lessen the chance of this happening.

Employment

A person with VWD can do any job.

The very small number of people with Type 3 VWD may find that certain jobs that are extremely demanding physically will take a toll on their joints in the long run.

Insurance

People with VWD are sometimes refused life insurance. In other cases, they are required to pay higher premiums. Hopefully, as VWD becomes better known by the public and by insurance companies, this situation will change. In the meantime, as different companies have different policies, the best advice is to shop around.

Travelling



What precautions should be taken when travelling?

People with VWD can travel wherever they please.

Following a few simple tips can make travelling even more relaxing.

- Find out from the Hemophilia/Bleeding Disorder Treatment Centre the names, addresses and phone numbers of treatment centres along the route.

- Take along up-to-date written medical information, including:
 - the exact VWD diagnosis
 - the exact prescription for desmopressin or factor concentrates, or other medication
 - the name and phone number of the treatment centre where you are known.

These papers could also prove useful at border crossings if customs officials become suspicious of the drugs, needles and syringes you are carrying.

- Make sure your, or your family member's, MedicAlert bracelet is up to date.
- If you, or your family member, self-infuses desmopressin or factor concentrates, make sure you have more than enough for the whole trip. Check that you have all the supplies (needles, syringes ...) you need.
- If you do not self-infuse, talk to the nurse coordinator at your Hemophilia/Bleeding Disorder Treatment Centre about the possibility of carrying along a supply of desmopressin or factor concentrate. These products are not available everywhere.
- If you do not want to carry these products with you, find out before you leave where they are available.
- Make sure you have a cooler to keep the products at the right temperature.
- Find out if your insurance coverage applies in the province or country you are visiting. If not, take out special travel insurance.
- When travelling by air, bus or train, always keep your medication with you. NEVER check it.

Medical identification



Should a person with von Willebrand disease carry medical identification?

Yes. Every person with VWD should wear a Medic Alert bracelet or necklace. Von Willebrand disease is not well known and not easily diagnosed. In the case of an injury or other emergency, the Medic Alert bracelet will be very helpful to medical personnel.

A person with VWD should always let medical personnel know that he/she has a bleeding disorder. As the person with VWD is very likely to be more knowledgeable about the disease than these non-specialists, he/she should not hesitate to inform them.

Most importantly, he/she should insist on adequate treatment to control bleeding in the event of a serious injury, or surgery. This may mean refusing consent for a dangerous surgical procedure. Medical personnel should be told to get in immediate contact with a Hemophilia/ Bleeding Disorder Treatment Centre for advice concerning VWD.



What is the life expectancy for people with von Willebrand disease?

The life expectancy for people with VWD is normal.

What's more, some researchers are finding that mild VWD could be a health benefit. They explain it this way. Von Willebrand disease makes it more difficult for platelets to stick together. Because of this, people with VWD could have less chance of blood clots blocking arteries (atherosclerosis), and therefore, less chance of heart attacks and strokes.

Can people with von Willebrand disease lead normal lives?

Absolutely! Most people with VWD have only occasional mild bleeding problems. Others bleed more frequently; however, with proper medical care, these bleeding episodes can be controlled.

People with VWD can:

- exercise, play sports and keep physically fit
- get an education
- hold a steady job
- marry and have children.



What are the best ways to manage von Willebrand disease?

- Learn all about von Willebrand disease.
- Find medical care in a centre which specializes in diagnosing and treating the disease.
- Live life to the fullest, knowing that von Willebrand disease can be successfully managed.



Where to get more information

VWD

**For more information about von Willebrand
disease or to get in touch with a
Hemophilia/Bleeding Disorder Treatment
Centre, contact:**

Canadian Hemophilia Society National Office

625 President Kennedy, Suite 505

Montreal, Quebec H3A 1K2

Tel: (514) 848-0503

Toll-free: 1 800 668-2686

Fax: (514) 848-9661

E-mail: chs@hemophilia.ca

Website: www.hemophilia.ca

Hemophilia/Bleeding Disorder Treatment Centres

VWD

MORE
INFORMATION

BRITISH COLUMBIA

Hemophilia Program of British Columbia - Adult Division
St. Paul's Hospital
Room 259, Comox Building
1081 Burrard Street
Vancouver, British Columbia V6Z 1Y6
Tel: (604) 682-2344, ext. 63026/63745
After hours: (604) 682-2344
Fax: (604) 806-8784

Hemophilia/ Hematology
Rm 1A13 - BC Children's Hospital
4480 Oak Street
Vancouver, British Columbia V6H 3V4
Tel: (604) 875-2345 ext. 5335
Pager: (604) 875-2161
After hours: (604) 875-2161
Fax: (604) 875-2533

ALBERTA

Southern Alberta Hemophilia Program
Alberta Children's Hospital
2888 Shaganappi Trail NW
Calgary, Alberta T3B 6A8
Tel: (403) 955-7311
After hours: (403) 955-7211
Fax: (403) 955-7393

Dr. John Akabutu Comprehensive Centre for Bleeding Disorders
8440 112th Street, CSB 7-109
University of Alberta Hospitals
Edmonton, Alberta T6G 2B7
Tel: (780) 407-6588
Pager: (780) 445-1683
Fax: (780) 407-2605

SASKATCHEWAN

Saskatchewan Bleeding Disorders Program
 Royal University Hospital
 103 Hospital Drive Box 113
 Saskatoon, Saskatchewan S7N 0W8
 Tel: (306) 655-6504
 After hours: (306) 655-1000
 Fax: (306) 655-6426

MANITOBA

Manitoba Bleeding Disorders Program
 Health Sciences Centre
 FE349-685 William Avenue
 Winnipeg, Manitoba R3E 0Z2
 Tel: (204) 787-2465
 Pager: (204) 787-2071 #3346
 Fax: (204) 787-1743

ONTARIO

Hamilton Health Sciences Corporation
 McMaster Division
 1200 Main Street West
 Hamilton, Ontario L8N 3Z5
 Tel: (905) 521-2100 #75978/75970
 24 hour: (905) 521-2100 #76443
 Fax: (905) 521-2654

Bleeding Disorders Program
 London Health Science Centre
 Victoria Hospital Rm E4-201
 800 Commissioners Road East
 London, Ontario N6A 5W9
 Tel: (519) 685-8500 ext. 53582
 Pager: #15358
 Fax: (519) 685-8543

Hemophilia Program
 Thunder Bay Regional Hospital Science Centre
 980 Oliver Road
 Thunder Bay, Ontario P7B 6V4
 Tel: (807) 684-6550
 Fax: (807) 684-5906

Comprehensive Hemophilia Care Centre
 St. Michael's Hospital
 30 Bond Street
 Toronto, Ontario M5B 1W8
 Tel: (416) 864-5129
 Pager: (416) 685-9404/9478
 After hours: (416) 864-5431
 Fax: (416) 864-5310

Hemophilia Program
 Hospital for Sick Children
 Hematology/Oncology Clinic
 Ward 8D
 555 University Avenue
 Toronto, Ontario M5G 1X8
 Tel: (416) 813-5871
 Pager: (416) 377-9716
 After hours: (416) 813-7500
 Fax: (416) 813-7221

Hematology Clinic
 Children's Hospital of Eastern Ontario
 401 Smyth Road
 Ottawa, Ontario K1H 8L1
 Tel: (613) 737-7600 ext. 2368
 Fax: (613) 738-4846

Regional Comprehensive Care Centre for Hemophilia and Hemostasis
 The Ottawa Hospital, General Campus
 501 Smyth Road, Box 248
 Ottawa, Ontario K1H 8L6
 Tel: (613) 737-8252
 After hours: (613) 722-7000
 Fax: (613) 737-8157

Hemophilia Program, Sudbury & North-Eastern Ontario
 Laurentian Site of HRSRH
 41 Ramsey Lake Road
 Sudbury, Ontario P3E 5J1
 Tel: (705) 522-2200 ext. 3264
 Fax: (705) 523-7077

SouthEastern Ontario Regional Inherited Bleeding Disorders Program
 Kingston General Hospital, Douglas 3
 Kingston, Ontario K7L2V7
 Tel: (613) 549-6666 ext. 4683
 Fax: (613) 548-1356
 After hours: (613) 548-3232 Page hematologist on call

QUÉBEC

Clinique d'hémophilie CHUS - Hôpital Fleurimont
 3001, 12^e avenue Nord
 Sherbrooke (Québec) J1H 5N4
 Tel: (819) 346-1110 ext. 14560
 Fax: (819) 820-6492 / (819) 564-5434 (hématologie)

Centre d'hémophilie Hôpital de Montréal pour Enfants
 2300, rue Tupper, Bureau A-216
 Montréal (Québec) H3H 1P3
 Tel: (514) 412-4420
 Fax: (514) 412-4424

Centre d'hémophilie - 1^{er} vidéotron Hôpital Ste-Justine
 3175, chemin de la Côte Ste-Catherine
 Montréal (Québec) H3T 1C5
 Tel: (514) 345-4931 #6031
 Pagers: (514) 415-5573/5584/5807
 After hours: (514) 345-4788
 Fax: (514) 514-345-7749

Quebec Centre for Coagulation Inhibitors
 Centre d'hémophilie - 1^{er} vidéotron
 Hôpital Ste-Justine
 3175, chemin de la Côte Ste-Catherine
 Montréal (Québec) H3T 1C5
 Tel: (514) 345-2360
 Fax: (514) 345-4828

Centre régional de l'hémophilie de l'est du Québec
 Hôpital de l'Enfant Jésus
 1401, 18^{ème} Rue
 Local J-S066 (sous-sol)
 Québec (Québec) G1J 1Z4
 Tel: (418) 649-5624
 Fax: (418) 649-5996
 After hours: (418) 649-0252

NEW BRUNSWICK

South East Regional Health Authority
 Hemophilia Clinic
 135 MacBeath Avenue
 Moncton, New Brunswick E1C 6Z8
 Tel: (506) 857-5465 / 857-5467
 Pager: (506) 558-7158
 Fax: (506) 857-5464

Inherited Bleeding Disorder Clinic
 Saint John Regional Hospital
 P.O. Box 2100, 400 University Avenue
 Saint John, New Brunswick E2L 4L2
 Tel: (506) 648-7286
 Fax: (506) 648-7379

NOVA SCOTIA

Pediatric Bleeding Disorder Clinic
 IWK Health Centre
 PO Box 9700
 6th Floor Ambulatory IWK Site
 5850 University Avenue
 Halifax, Nova Scotia B3K 6R8
 Tel: (902) 470-8752 / 470-8819
 Pager: (902) 470-8888 #1982
 After hours: (902) 470-8394
 Fax: (902) 470-7208

Hereditary Bleeding Disorders Program (Adult)
 Victoria General Hospital Site
 Queen Elizabeth II Health Sciences Centre
 Rm: 4020 Centennial Building
 5820 University Avenue
 Halifax, Nova Scotia B3H 1V8
 Tel: (902) 473-5612
 Fax: (902) 473-7596

NEWFOUNDLAND

Eastern Health Corporation
Health Sciences Centre
Janeway Site, Room 2J755
300 Prince Philip Drive
St. John's, Newfoundland A1B 3V6
Tel: (709) 777-4388
Fax: (709) 777-4292

acquired VWD A non-hereditary type of VWD in which a person suddenly develops antibodies, or inhibitors, to the VWF normally produced in the blood.

Amicar An anti-fibrinolytic drug (aminocaproic acid) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

anesthesiologist A physician who specializes in controlling pain and consciousness during surgery.

antibody A substance produced in the blood by the body's immune system to defend against other harmful substances.

anti-diuretic A substance that makes the body retain water.

anti-fibrinolytics Drugs (Cyklokapron and Amicar) that help to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

bleeding disorder A disease in which the body is unable to form blood clots as quickly or as effectively as normal. The family of bleeding disorders includes von Willebrand disease, hemophilia A, hemophilia B, platelet function disorders and a variety of rare factor deficiencies. The disorder may be hereditary or acquired.

bleeding time The time required for a minor cut to stop bleeding. As a test, it is unreliable in diagnosing VWD.

blood clotting The process of forming a permanent clot to repair a damaged blood vessel. It includes four steps: vasoconstriction, platelet aggregation, platelet adhesion, and the formation of a fibrin plug.

blood clotting proteins Substances that circulate in the bloodstream, necessary in blood clotting. They include von Willebrand factor, and factors I, II, V, VII, VIII, IX, X, XI and XIII.

blood type The particular kind of blood each person has. The types are A, B, AB and O.

chromosome A long chain of chemicals known as DNA, which is arranged into about 25,000 units called genes. Genes determine such things as the colour of a person's eyes.

comprehensive care All of the medical services needed by a person with VWD and his/her family for the treatment of VWD and related conditions. This care is provided at a Hemophilia/Bleeding Disorder Treatment Centre.

cryoprecipitate A blood component made from plasma, containing VWF and factor VIII, commonly used to treat VWD in the past. However, because there is no method to kill viruses in cryoprecipitate, it is no longer recommended.

cyklokapron An anti-fibrinolytic drug (tranexamic acid) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

desmopressin A synthetic drug which is a copy of a natural hormone. It acts by releasing VWF stored in the lining of the blood vessels. Desmopressin is not made from blood. It can be called DDAVP, Octostim and Octostim Spray, Stimate and Stimate Nasal Spray.

dilatation & curettage (D&C) An operation to scrape and clean the lining of the uterus.

dysmenorrhea Pain during the menstrual period.

endometriosis A condition in which endometrial tissue forms outside the uterus, for example, around the abdomen. When a woman menstruates, endometrial tissue, wherever it is in the body, bleeds.

epidural A type of local anesthesia in which a needle is placed into the spine to freeze the lower part of the body.

episiotomy A procedure sometimes done during childbirth in which the skin is cut near the vagina to avoid tearing.

factor VIII A protein in the blood that is essential for clotting. Factor VIII levels are low in people with VWD and hemophilia A.

fibrin clot The clot which forms in the last stage of the coagulation process.

gene Tiny structures of DNA which determine such things as the colour of a person's eyes. VWD is caused by an abnormal gene on chromosome 12.

gynecologist A physician who specializes in the woman's reproductive system.

Humate P A blood product made from human plasma, used in the treatment of VWD, containing concentrated von Willebrand factor and factor VIII.

hematologist A physician specializing in diseases of the blood.

hemoglobin A substance in the red cells of blood, responsible for carrying oxygen. When a person has iron deficiency anemia caused by loss of blood, hemoglobin levels are lower than normal.

hemophilia A term used to describe bleeding disorders caused by low levels of factor VIII or IX (hemophilia A and B). The term can also be used more broadly to describe the family of bleeding disorders, including VWD.

hemorrhage The escape of blood from blood vessels, either on the surface of the body or internally.

home care The care of the person with VWD at home, rather than in hospital. This includes the administration of medication by the person with VWD or by a family member.

hormone A secretion in the blood that stimulates organs into action.

hormone therapy The administration of oral contraceptives or other hormones (e.g. progesterone) to raise VWF levels or reduce menstrual bleeding.

hysterectomy An operation to remove the uterus, and in some cases, the ovaries.

inhibitors Antibodies produced to eliminate VWF or other clotting factor proteins, seen as foreign by the body's immune system.

iron deficiency anemia A condition caused by low hemoglobin levels because of blood loss, leading to fatigue and lack of energy.

laparoscopic surgery An operation in which a tiny camera is used to examine the abdominal area. In VWD, this operation is used to remove endometrial tissue that has formed outside the uterus.

leukemia A form of cancer characterized by an excess of white blood cells.

menorrhagia Bleeding during the menstrual cycle which is heavier than normal or lasts longer than normal.

metorrhagia Irregular bleeding throughout the menstrual cycle.

mid-cycle pain Pain occurring during ovulation, which can be due to bleeding from the ovary at the site of ovulation. Also called dysmenorrhea.

mucous membrane An extension of the skin inside the body - for example, the insides of the mouth, the nose, the intestines (the gut) and the uterus (the womb).

multimer A part of the structure of the VWF molecule. In Type 2M VWD, changes in the multimers affect the binding of the VWF to platelets.

obstetrician A physician who specializes in conception, pregnancy and childbirth.

oophorectomy An operation to remove the ovaries.

ovulation The release of the egg from the ovary at the mid-cycle of a woman's period.

partial thromboplastin time A routine blood test which often gives normal results in people with VWD.

parvovirus A human virus carried by a large percentage of the population. Normally harmless, in rare cases, it can cause anemia. It can also cause miscarriage.

plasma The portion of blood that contains clotting factor proteins, including VWF and factor VIII, as well as immunoglobulins and albumin.

plasmin A substance in the blood that dissolves blood clots after the blood vessels have healed.

platelets Small cells less than 1/10,000 of a centimetre in diameter circulating in the blood, which stick to and spread on the walls of the damaged blood vessel to promote clotting.

platelet function tests These tests measure how well the platelets work to control bleeding.

progesterone A natural hormone. As a therapy for VWD, it works by thickening the lining of the uterus and making it less prone to bleed.

prothrombin time A routine blood test which gives normal results in people with VWD.

Type 1 VWD A form of von Willebrand disease in which the von Willebrand factor is present at lower than normal levels, affecting blood clotting.

Type 2 VWD A family of different forms of von Willebrand disease in which the von Willebrand factor does not work properly, affecting blood clotting.

Type 3 VWD A form of von Willebrand disease in which the von Willebrand factor is almost totally missing. This is the most severe form of VWD.

uterine ablation Also called endometrial ablation. An operation to destroy the lining of the uterus. The operation is performed through the vagina. The uterine lining is burned away.

vasoconstriction The first stage in blood clotting in which the blood vessel constricts to reduce the flow of blood to the damaged area.

von Willebrand disease (VWD) A family of inherited diseases in which the blood clots more slowly than normal.

von Willebrand factor (VWF) The clotting protein that is deficient in VWD. The VWF is either present at lower than normal levels or it does not work properly.

von Willebrand factor concentrate A blood product made from human plasma, used in the treatment of VWD, containing concentrated von Willebrand factor and factor VIII. In Canada, the most widely used brand is called Humate P.

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Canadian Hemophilia Society
Help Stop the Bleeding

www.hemophilia.ca