All About von Willebrand disease

Produced by

Bleeding Disorders Canada

Formerly the Canadian Hemophilia Society

June 2026

All About von Willebrand disease

Fourth Edition

This booklet was written by Natalie Philbert, National Programs Director, Bleeding Disorders Canada (formerly the Canadian Hemophilia Society).

The document was reviewed and edited by:

Dawn Goodyear, MD, MSc, FRCPC, Adult Hematologist, Calgary, Alberta

Rochelle Winikoff, MD, FRCPC, CHU Sainte-Justine, Montreal, Quebec

Danielle Meschino, MD, FRCPC, Adult Hematology Fellow, St. Michael's Hospital, Ontario

Vanessa Bouskill, MN, Nurse Practitioner-Paediatrics, The Hospital for Sick Children, Toronto, Ontario

Kay Decker, Hemophilia Nurse Coordinator, Hamilton Health Sciences

Karen Mantya, RSW, Social Worker, Saskatchewan Bleeding Disorders Program, Saskatoon, Saskatchewan

Elia Fong, Physiotherapist, Stollery Children's Hospital, Edmonton, Alberta

Amy Griffith – Person with VWD, Ontario

Trevor Reid – Person with VWD, Ontario

For further information, please contact:

Bleeding Disorders Canada (formerly the Canadian Hemophilia Society)
Telephone: 514-848-0503
Toll-free: 1-800-668-2686
info@bleedingdisorderscanada.ca
bleedingdisorderscanada.ca

ISBN: 978-1-897489-54-3

Preface

In recent years, our understanding and awareness of the challenges, concerns and misunderstandings faced by people with von Willebrand disease (VWD) has grown considerably.

In the year 2000, the Canadian Hemophilia Society (now Bleeding Disorders Canada) 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. In 2007 and 2011 a revised edition was produced as we continued to strive for the most up-to-date information for people living with VWD.

Now in 2026 we are proud to offer the fourth revised edition as we continue to expand our knowledge and share that information with the individuals and families whose lives are touched by VWD. In recent years we have learned much more about the quality of life for people living with VWD and the specific challenges women, girls and people with the potential to menstruate face regularly.

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. We look forward to continuing to grow with you in our understanding and knowledge of VWD. Please note that clinical practices across the country may vary so it is always recommended to speak to your medical team about the best approach for your situation.

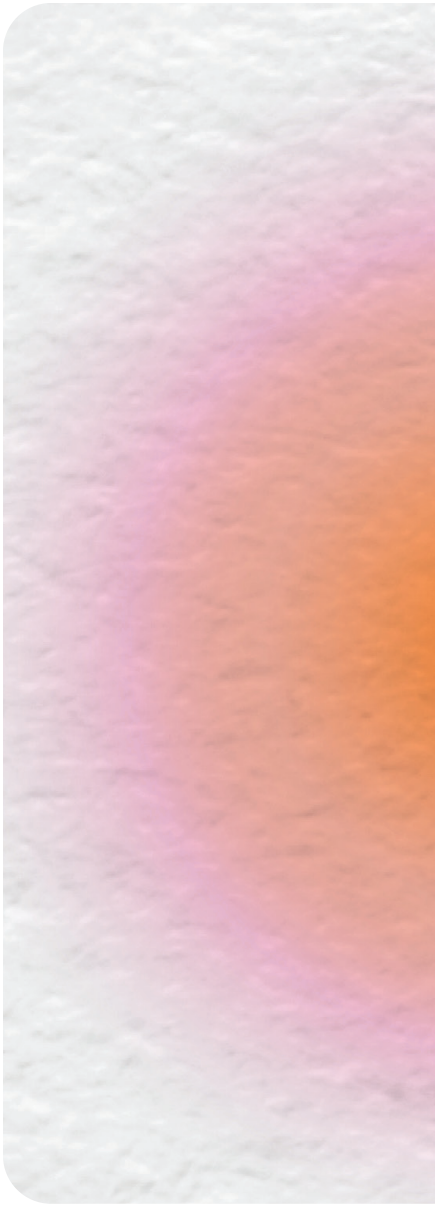


Table of contents

An introduction to von Willebrand disease 1

Types of von Willebrand disease 4

Heredity 7

Symptoms 13

Diagnosis 17

Treatment options 20

Treatment options for Type 1 von Willebrand disease 21

Treatment options for Type 2 and 3 von Willebrand disease . . 24

Treatment options for heavy menstrual bleeding 26

Key questions to discuss with your medical team about treatment options 31

Living with von Willebrand disease 33

Comprehensive care 33

Safety of blood products 36

Nosebleeds 38

Conception, pregnancy and childbirth 39

Medication to avoid 43

Safe treatment options for pain management. 43

Exercise, fitness and sports 44

Childcare and schooling 45

Teens and sexuality 46

Employment 46

Insurance 46

Travelling 47

Medical identification 48

Final word 49

Glossary 50

Contact information 55

An introduction to von Willebrand disease

WHAT IS VON WILLEBRAND DISEASE?

Von Willebrand disease (VWD) was first identified in 1926 by Dr. Erik von Willebrand. The majority of people are born with VWD, inherited from one or both parents, however, in rare cases a person can get VWD without inheriting it from parents or a family history of VWD.

VWD is the most common bleeding disorder that people have worldwide and it is estimated that 1 in 1,000 people have symptoms of VWD that require medical attention.

VWD is due to deficiencies in the von Willebrand factor (VWF). VWF is a protein in blood which is necessary for proper blood coagulation, or clotting. VWF works with another protein factor (Factor VIII) to help control bleeding.

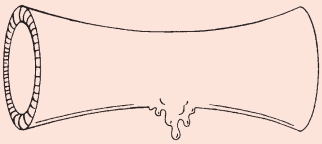
When there is not enough VWF in the blood, or when it does not work the way that it should, the blood takes longer to clot. VWD impacts men and women equally, in terms of numbers affected but not in terms of symptoms or quality of life. More women are typically diagnosed due to gynecological and obstetrical related challenges.

HOW DOES BLOOD CLOT NORMALLY?

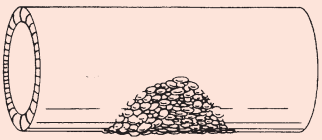
Blood is carried throughout the body within a network of blood vessels. When tissues are injured, damage to a blood vessel may result in leakage of blood through holes in the vessel wall. The vessels can break near the surface, like a cut, or they can break deep inside the body, making a bruise or an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

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

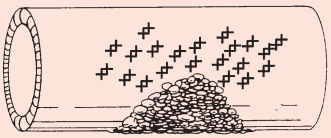
FIGURE 1.



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

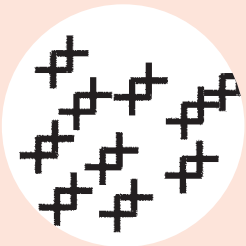


Stage 2:
Platelets, which are small cell fragments circulating in the blood, stick to, spread and fill the injured areas on the exposed collagen wall of the damaged blood vessel.



Stage 3:
Von Willebrand factor acts as a glue to hold the platelets and collagen together at the site of the damaged 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.

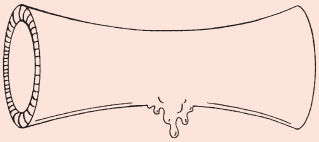


Normal von Willebrand factor (VWF)

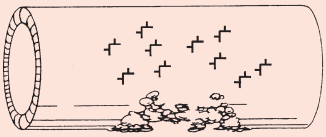
HOW DOES VWD AFFECT THE NORMAL CLOTTING OF BLOOD?

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

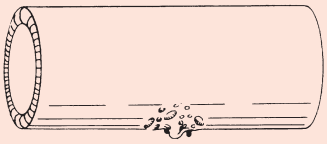
FIGURE 2.



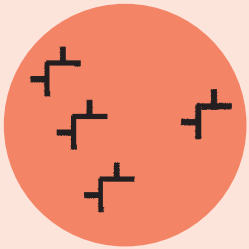
Stage 1:
The blood vessels of people with VWD constrict normally.



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 or the platelets do not stick properly to the collagen of damaged tissue.



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.



Abnormal von Willebrand factor (VWF)

There are various types of VWD that are caused by a problem with the VWF.

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 itself has its own subtypes.

WHAT IS TYPE 1 VWD?

Type 1 VWD is the most common form, accounting for 70-80% of all cases of VWD. In Type 1 VWD, the von Willebrand factor (VWF) works normally, but there is not enough of it.

Some people with Type 1 VWD have no symptoms at all until they experience a bad injury or a surgery. Other people with Type 1 VWD can experience many symptoms.

WHAT IS TYPE 2 VWD?

Type 2 VWD is less common than Type 1. It represents 15-30% of all cases.

In Type 2 VWD, the amount of VWF in people's blood can be normal. The problem is that the VWF does not work properly.

There are several subtypes of Type 2 VWD. It is important to get an exact diagnosis because the subtypes 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 can be 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, VWF binds to platelets in the bloodstream with increased stickiness. Then, the body removes these large bundles of platelets from circulation. This causes a shortage of platelets.

Type 2N VWD is rare. (The "N" stands for Normandy, France where the sub-type was first identified.)

- 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.
- Due to low factor VIII levels, Type 2N is sometimes mistaken for factor VIII deficiency, also known as hemophilia A.
- Type 2N VWD occurs when a child inherits one Type 1 (or Type 2N) gene from one parent and one Type 2N gene from the other parent.

Type 2M is another variant of VWD.

- In Type 2M, binding of the VWF to collagen is normal, but binding to the platelet is impaired.

WHAT IS TYPE 3 VWD?

Type 3 VWD is very rare and the most severe. It affects about 1 in 500,000 people and accounts for <5% of people with VWD. People with Type 3 VWD have very low to no 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. Historically, VWD symptoms have been considered less severe than hemophilia, however some studies have shown that Type 3 VWD and severe hemophilia A can be similar, and in some cases, more severe.

Usually, for a baby to have 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 new or spontaneous mutation in the child's gene inherited from the other parent.

While it is important to understand the type of VWD a person has for the purposes of treatment options, the type does not automatically indicate the symptoms and severity a person will experience. For example, a female with Type 1 VWD may experience severe bleeding symptoms, despite having a milder form of VWD.

Conversations with the medical team about symptoms and severity of bleeds help create a whole picture of the bleeding disorder, in addition to the diagnosis of which type of VWD.

FIGURE 3.

Types and severity of von Willebrand disease

People with VWD have increased bleeding episodes depending on the severity of their disease. When they have a bleeding episode, they bleed for a longer period of time than a person without VWD.

There are several types of VWD: Type 1, Type 2 and Type 3. These vary in severity of symptoms and not all people will experience all symptoms. Some people will have minimal symptoms or have no symptoms at all.

	Diagnosis	Severity	Symptoms
Type 1	Low levels of VWF; may have slightly low factor VIII levels	Mild bleeding symptoms	Nosebleeds, mouth bleeds and heavy menstrual bleeding
Type 2	Levels of VWF may be normal, but it does not work properly	Mild to severe bleeding symptoms	Same as in Type 1, but more severe; Type 2N can experience joint and muscle bleeds
Type 3	Absent or extremely low levels of VWF; factor VIII levels will also be very low	Severe bleeding symptoms	In addition to Type 1 symptoms, will experience joint and muscle bleeds

Type 2 subtypes

Type 2 VWD has four subtypes; treatment varies for each of these subtypes.

	Description
Type 2A	This is the most common subtype. In this subtype, VWF does not work properly to bind platelets together.
Type 2B	In people with Type 2B, the VWF is "extra sticky" and it binds to platelets in the blood stream rather than at the site of the wound. This causes a shortage of platelets in areas where they are needed. Sometimes this type of VWD is misdiagnosed as a low platelet count such as idiopathic thrombocytopenia purpura (ITP).
Type 2N	In this subtype, the VWF binds to platelets at the site of the wound in the same way that it would in healthy people. However, while VWF normally transports clotting factor VIII, this does not occur in people with Type 2N. As a result, low levels of factor VIII lead to poor blood clot formation. Because of low levels of factor VIII, Type 2N VWD may be misdiagnosed as hemophilia.
Type 2M	In this subtype, VWF does not bind properly to platelets. This subtype is similar to type 2A but is distinguished through various specialized laboratory tests.

Heredity

WHO CAN HAVE VON WILLEBRAND DISEASE?

Von Willebrand disease affects both males and females equally. However, because VWD can cause heavy menstrual bleeding and prolonged bleeding after childbirth, more females than males have noticeable symptoms.

HOW DOES A PERSON GET VON WILLEBRAND DISEASE?

Von Willebrand disease is a hereditary disorder. It is caused by an abnormal gene. If one or both parents have VWD, the gene defect can be passed on to their child(ren).

Each cell of the body contains structures called chromosomes that carry DNA. DNA holds instructions, or known as genes, that determines characteristics like eye colour.

Each cell contains 23 pairs of chromosomes, with one set of chromosomes from a parent. Von Willebrand disease is caused by a defect on chromosome 12. The gene defect could be from the mother, father, or both. A person with VWD carries at least one defective gene.

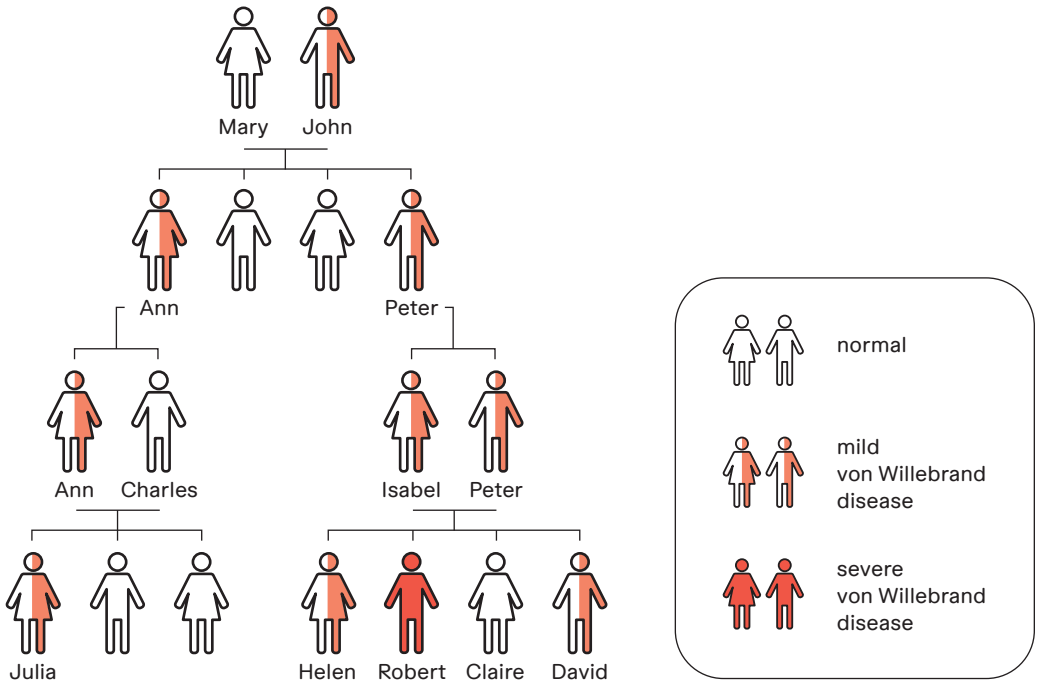
There are two ways of getting the hereditary form of VWD.

- 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 child's genes can undergo a change. This is called a new or spontaneous genetic mutation. The parents do not carry the defective gene, and the child's siblings would not inherit it.

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 4 shows three generations of a family with VWD.

FIGURE 4.



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) partners with 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) partners with 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 (Robert).

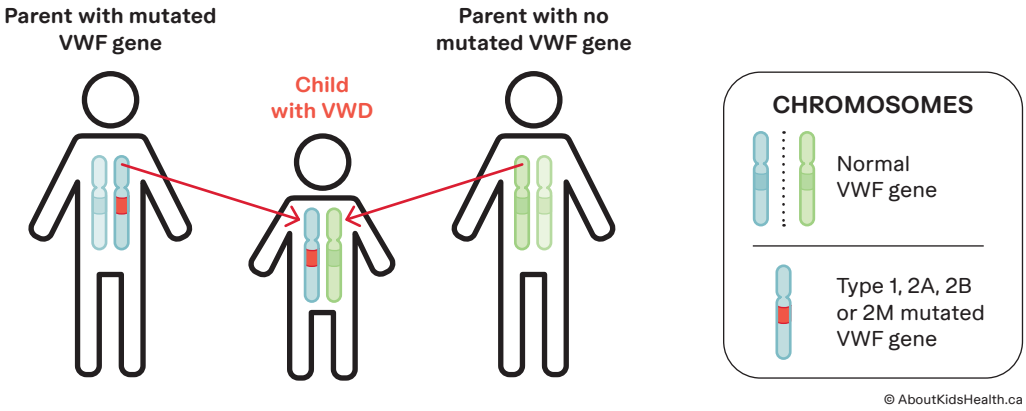
Figure 5 provides an illustration of how VWD is inherited based on the subtypes of VWD.

FIGURE 5.

How is von Willebrand disease inherited?

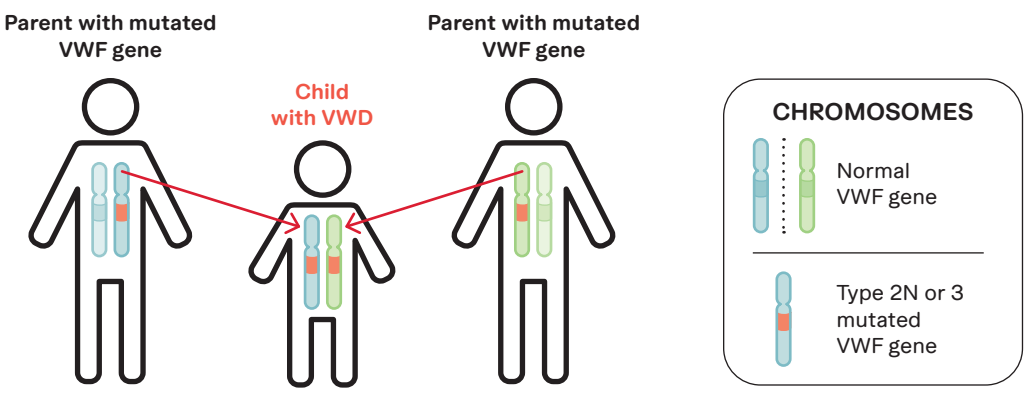
VWD is a genetic condition and it is most often passed on from one or both parents to their children. Occasionally, VWD may occur when one of the child's genes changes randomly; this is called a spontaneous genetic mutation.

Inheritance pattern of Type 1, 2A, 2B and 2M von Willebrand disease



The type of VWD depends on the mutation in the VWF gene. The parent may have mild symptoms or no symptoms at all. Types 1, 2A, 2B and 2M are usually inherited when one parent passes on the mutated VWF gene to their child.

Inheritance pattern of Type 2N and 3 von Willebrand disease

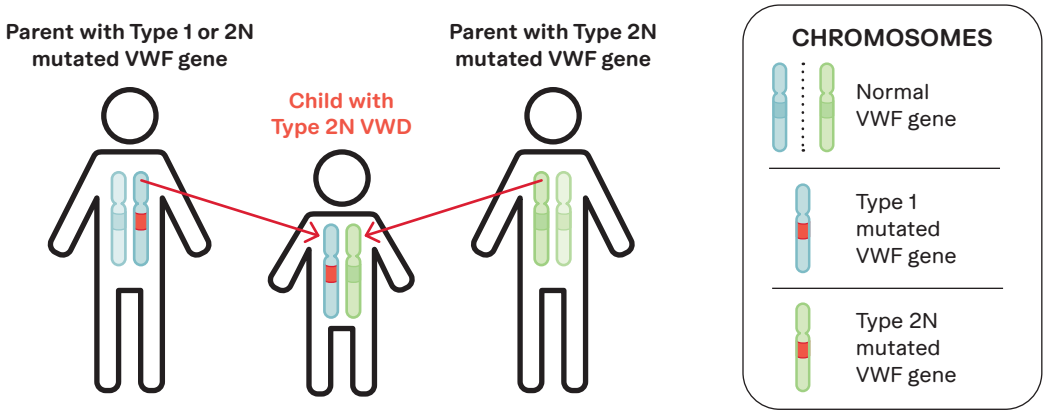


Reprinted with permission from The Hospital of Sick Children Von Willebrand disease

FIGURE 5. (CONTINUED)

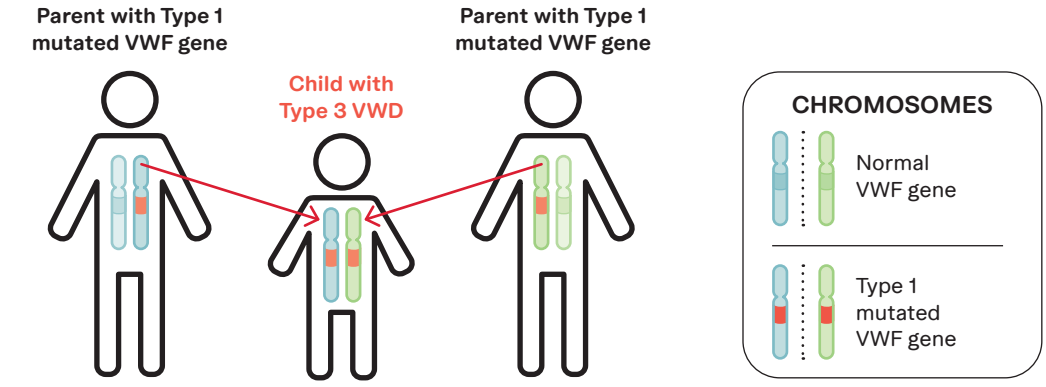
Types 2N and 3 VWD are usually inherited when both parents pass on the mutated gene to their child. A child receives two mutated genes, one from each parent. Each parent may have only mild symptoms or no symptoms at all.

Inheritance pattern of Type 2N von Willebrand disease



Type 2N VWD occurs when a child inherits one Type 1 (or Type 2N) gene from one parent and one Type 2N gene from the other parent.

Inheritance pattern of Type 3 von Willebrand disease



Type 3 VWD occurs when a child inherits a gene responsible for one Type 1 VWD from both parents. In most (but not all) cases parents of a child with Type 3 VWD, despite having a gene for Type 1 VWD, usually have normal levels of VWF and show no signs of bleeding.

Reprinted with permission from The Hospital of Sick Children 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 carries the gene. The child could have gotten the abnormal gene through a new mutation. In this case, chromosome 12 changes at conception or soon after. In the future, this child could pass on the VWD gene to their children.
- A person can develop non-genetic forms of VWD at any stage in life. This is an acquired form of VWD. Neither of the person's parents carries the VWD gene, nor does the affected person. There are several ways in which acquired VWD can develop both in children and in adults. VWF may be physically broken as in the case of various cardiac conditions (aortic/pulmonary stenosis, pulmonary hypertension, cardiac assisted devices) or abnormally eliminated from the circulation (antibody or platelet complexes). Acquired VWD may occur because of certain medications a person has taken in immune diseases, 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 some cases, parents of a child with severe Type 3 VWD do not show symptoms. Occasionally one, or both parents, will have mild bleeding problems.

IS VON WILLEBRAND DISEASE ALWAYS THE RESULT OF A GENETIC CONDITION?

No. As explained above, 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. There may also be feelings of shock or grief about what a parent had pictured for their child's future. 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.

Symptoms

WHAT ARE THE SYMPTOMS OF VON WILLEBRAND DISEASE?

Symptoms of VWD may include:

- easy bruising and/or longer period of healing
- frequent or prolonged bleeding from nose and gums
- prolonged bleeding from minor cuts
- bleeding from the gums when baby teeth erupt, fall out or after tooth extractions
- spontaneous or prolonged bleeding in the mouth
- excessive or prolonged bleeding following injury, surgery, dental work, or childbirth
- bleeding in the upper and lower gastrointestinal tract. Symptoms would include:
  - abdominal pain
  - black stools or passing bright red blood
- Potential bleeding into joints and muscles. Symptoms would include:
  - morning stiffness
  - increasing pain, even with rest
  - a feeling of tightness in the joint or muscle
  - swelling and a puffiness to the touch
  - heat at the site of the bleeding
  - decreased joint movement
  - injuries taking longer to heal
- Symptoms related to menstruation (see the below section "***Specific Issues Related to Menstruation***")

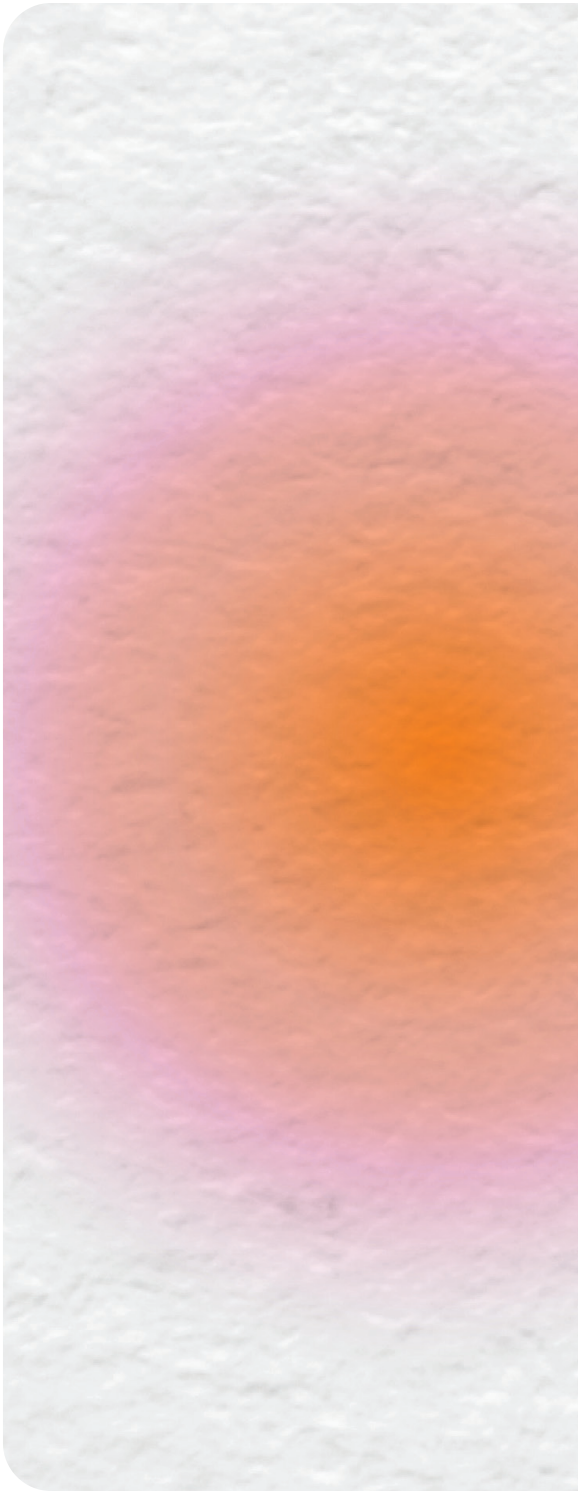
However, some people have few or no symptoms. They realize they have a bleeding problem only when another person in the family is diagnosed with VWD or after they have a serious injury or surgery. Symptoms also vary greatly from person to person. Even members of the same family can have different symptoms.

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
- frequent and/or prolonged nosebleeds
- prolonged bleeding from minor cuts
- heavy menstrual bleeding at the start of menarche (the first menstrual period).

In cases of severe Type 3 VWD, bleeding can occur in newborns, especially from the umbilical cord and at the time of circumcision. There can also be a risk of ICH (Intracranial hemorrhage) from delivery or assisted delivery with the use of forceps or vacuum. These types of bleeding can also occur with Types 1 and 2 VWD, however it is more common in Type 3 VWD.



SPECIFIC ISSUES RELATED TO MENSTRUATION

It can be hard to tell if menstrual bleeding is unusually heavy compared to others. In a family with several members who have VWD, heavy menstrual bleeding may just be the "norm" in the family. Sisters, mothers, aunts and grandmothers often have the same type of bleeding. Nobody sees it as special or, if they do, they say, "All the women in our family bleed a lot during their periods." Other families may not talk about menstruation at all, so normalcy and family history may be difficult to obtain.

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

- heavy or prolonged bleeding during menstruation
- menstruation lasting longer than 7 days
- passing clots larger than a quarter
- changing menstrual products every hour or regularly flooding
- requiring double menstrual product protection
- changing menstrual products during the night
- iron deficiency/anemia
- heavy menstrual bleeding impacting daily activities and/or quality of life.

Menstrual bleeding can be especially heavy at the time of a first period. For this reason, when there is a family history of a bleeding disorder connecting with a medical team to help treat the bleeding is a good idea. The medical team should include:

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

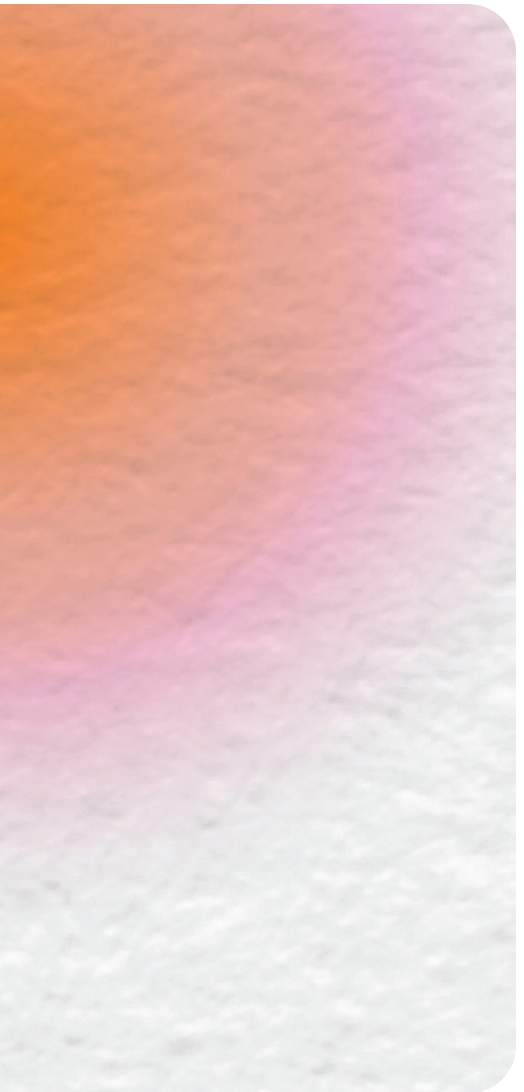
DYSMENORRHEA AND MID-CYCLE PAIN

Some women with VWD have pain during their menstrual periods. This is called dysmenorrhea. Dysmenorrhea is usually worse when combined 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. With VWD this bleeding can be significant and cause severe pain.

VWD can also complicate matters for women who have endometriosis. With this condition, endometrial tissue forms outside the uterus, for example, around the abdomen. During menstruation, endometrial tissue, wherever it is in the body, bleeds. The bleeding can be particularly heavy if a person also has VWD. The blood can irritate the abdominal wall, causing pain. Typically, this pain starts a few days before the onset of vaginal bleeding.

HOW DOES MENOPAUSE IMPACT VON WILLEBRAND DISEASE?

After many years of battling heavy periods, there can be the assumption that VWD will be better after menopause. In fact, hormone regulation becomes more chaotic closer to menopause, and this increases the risk of unpredictable and heavy bleeding. It is important to maintain a strong relationship with a gynecologist even after childbearing to anticipate the combined issues of menopause and VWD.



HOW DOES HEAVY MENSTRUAL BLEEDING IMPACT QUALITY OF LIFE?

Heavy menstrual bleeding can significantly impact quality of life. Here are some examples. Heavy menstrual bleeding caused by VWD may:

- cause pain during menstrual periods and at times of ovulation
- cause fatigue from iron deficiency anemia
- cause embarrassment and/or anxiety due to worry over leaking menstrual blood or staining clothes when at work or school or out socially, which can cause real or perceived isolation
- result in mental health challenges, such as depression, anxiety or isolation
- impact relationships and sexual intimacy
- limit the amount of time at work or school or struggle to work effectively during menstruation
- result in unnecessary procedures or surgeries such as a hysterectomy due to doctors not diagnosing or misdiagnosing a bleeding disorder
- create mistrust in the medical profession after being told for years that their bleeding was normal
- impact job and career choices.

Fortunately, with proper diagnosis and treatment, these challenges can be dramatically reduced and even eliminated.

Diagnosis

IS DIAGNOSING VON WILLEBRAND DISEASE EASY?

Diagnosing VWD can be complicated and relies on multiple steps. Even with the knowledge about VWD, many doctors are still not familiar with it. As a result, VWD is often underdiagnosed or misdiagnosed.

For example:

- a person with a low platelet count because of Type 2B VWD could be misdiagnosed with Immune Thrombocytopenia (ITP)
- a woman with heavy, prolonged menstrual bleeding because of VWD, who has not responded to other effective treatment modalities that are available, could be advised to have a hysterectomy.

For this reason, a person who thinks they have 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 several 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 monthly menstrual cycle
- being on hormonal therapy, sometimes referred to as the birth control pill or oral contraceptive
- having an infection
- having recently undergone surgery
- having recently had a blood transfusion
- being stressed
- 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 VWD.

WHAT TESTS ARE RELIABLE IN DIAGNOSING VON WILLEBRAND DISEASE?

A doctor who suspects VWD will first take a complete family history. This can sometimes be challenging as the family history depends on the understanding of the patient of the family history and on being aware of what constitutes abnormal bleeding. The doctor will then do several blood tests to find out:

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

Ideally, VWD testing should be carried out at a specialized centre when needed. There may be a cost for VWD testing when done at community labs, depending on geographic location and these tests will likely need to be repeated at least once by a specialized lab. The tests may include...

- **Factor VIII:C:** This measures the amount of factor VIII clotting activity.
- **VWF: antigen:** This measures the amount of von Willebrand factor.
- **VWF: activity:** This measures how well the VWF works.
- **VWF multimers:** This examines the structure of the VWF.
- **Genetic testing:** This helps distinguish subtypes of VWD.

HOW LONG DOES IT TAKE TO GET THE RESULTS?

Routine VWD results can take several weeks to be obtained in most centres. However, in urgent situations results can be obtained within several hours (when needed) at a specialized lab. Genetic testing results may take several months. Genetic testing is not required to make a diagnosis of VWD in most cases but may be helpful for Type 2 and Type 3 VWD.

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 due to the multiple factors that can influence VWF levels.

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 they have a bleeding problem should discuss the need for being re-tested with their doctor.

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. 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 recognize that there is no "right" response and to get support from others. Medical people, organizations that specialize in bleeding disorders, such as your local chapter of Bleeding Disorders Canada (formerly the Canadian Hemophilia Society), 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 assistance in managing fears.

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 hematomas can often be controlled by applying pressure and elevating the limb.
- Bleeding from minor cuts can be stopped by applying pressure.
- Nosebleeds may be stopped with simple first aid techniques (See "***Nosebleeds***" on page 38.)

However, sometimes medical treatment is necessary. The type of treatment depends in part on the type of VWD a person has and on the type of bleeding they experience. Treatment also depends on how bleeds are impacting the quality of life of a person. Even if the bleeds are considered minor but are frequent or impacting everyday life, treatment can be discussed with your medical team.

WHERE IS THE BEST PLACE FOR A PERSON WITH VON WILLEBRAND DISEASE TO GET TREATMENT?

Few doctors are familiar with VWD, however, through efforts in education and raising awareness this is slowly beginning to change. Even hematologists, who deal with blood disorders, are rarely experts in diagnosing and treating VWD. Many obstetricians and gynecologists remain unaware of the consequences of VWD, particularly related to menstruation, childbirth and menopause. Therefore, the best place for a person with VWD to get treatment is a hemophilia/bleeding disorder treatment centre. (See "***Comprehensive Care***" on page 33.)

Once diagnosed and an individualized treatment plan has been made, the doctors at the hemophilia/bleeding disorder treatment centre can work with the individual and their primary care provider to ensure that it is part of the whole care plan.

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, except for a variant of VWD Type 1 called VWD Type 1 C in which the response to desmopressin is short-lived. 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. The nasal spray form has not been available for quite some time and is unfortunately off the market for an undetermined period of time.

Desmopressin is effective for many people with Type 1 VWD. However, different people respond to desmopressin in different ways. Therefore, a doctor may do tests to find out each individual's response to the drug. This is called a DDAVP Challenge. These tests may be 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 concentrate*** on page 24.)

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

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

Desmopressin is also an anti-diuretic, which retains water in the body. Therefore, doctors recommend restricting liquids for 24 hours following a dose.

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, the person should go to the hemophilia/bleeding disorder treatment centre or emergency room for help. In infants and young children, due to fluid retention DDAVP should be used with caution and as such is not given to children under 2 years of age. However, it is important to note that some medical teams may choose not to give young children DDAVP under 3 years of age. It is best to have a discussion with the medical team about the right choice for your specific situation.

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

ANTIFIBRINOLYTIC AGENT (TRANEXAMIC ACID)

Tranexamic acid (i.e. TXA or Cyklokapron) is an example of a drug that stops the activity of an enzyme, called plasmin. Plasmin dissolves blood clots. Antifibrinolytic agents put the brakes on plasmin, allowing more opportunity for the clot to form.

Because these drugs do not help make a clot, it cannot be used in place of desmopressin or factor VIII/VWF concentrate in cases of serious bleeding.

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).

Tranexamic acid has proven very useful for people with VWD. It can be used:

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

Tranexamic acid (i.e. Cyklokapron) comes in tablet and intravenous form. It 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. Of note, tranexamic acid (Cyklokapron) does not increase the risk of blood clots. Tranexamic acid (Cyklokapron) can also be available in a topical intranasal cream and as a liquid mouthwash.

FIBRIN GLUE

While Fibrin glue is not often used anymore, it is a product that is removed from blood and manufactured as a natural clotting agent. In the coagulation process, the final clot is made up of fibrin. 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.

FACTOR VIII/VON WILLEBRAND FACTOR CONCENTRATE

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 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 receives the blood product 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. The screened plasma is then treated to destroy any viruses that may still be present. Factor concentrates used today have an excellent safety record. Compared to screening in 2011, plasma sent for fractionation is now screened for Parvovirus B19 virus using a nucleic acid amplification test (NAT). This testing is done by plasma fractionators and follows established protocols for pre-pooling testing. Any units testing positive are discarded. The risk of B19 transmission in fractionated plasma is considered very low.

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.

Humate P and Wilate are examples of concentrates of FVIII/VWF that are available for the treatment of VWD in Canada.

FVIII/VWF concentrate is given through intravenous (IV), or injected into a vein. It can be administered at a clinic, doctor's office or emergency room. Many people learn to treat themselves at home. (See "***What is home care?***" on page 34.)

A recombinant form of von Willebrand factor, called Vonvendi, was approved by Health Canada in 2019 to control bleeding and for surgery in VWD; however, it is not routinely available. In rare cases, it is available through provincial drug plans' exceptional access programs.

Other factor VIII concentrates such as monoclonal factor VIII and recombinant factor VIII contain no von Willebrand factor. Therefore, they are of limited value in treating VWD. It is important to note that there can be a risk of inhibitor formation in VWD (the development of antibodies that neutralize clotting factors, making treatment less effective) in people with Type 3 VWD.

TRANEXAMIC ACID (CYKLOKAPRON)

Bleeding in mucous membranes is often treated with tranexamic acid (Cyklokapron) and sometimes Fibrin Glue. (See the treatments for Type 1 VWD.)

DESMOPRESSIN

Desmopressin can be recommended for Type 2 VWD. Desmopressin releases stored VWF, which does not function properly in Type 2 VWD.

This drug could make platelet clumping worse in Type 2B VWD, resulting in lower platelet count or even cause unwanted clots in the blood stream. Formation of an effective clot would be even more difficult.

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.

Desmopressin is not recommended in the treatment of Type 3 VWD, as these individuals will have no storage of VWF in their body and therefore will not respond.

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. Hormone therapy and antifibrinolytic drugs are considered first line treatment for many for heavy menstrual bleeding.

*** Desmopressin should be used with caution in Type 1C due to its short response time**

Not generally recommended treatment

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 currently no method to kill viruses in cryoprecipitate, it is no longer generally recommended in Canada. However, it is used in refractory bleeding because it is rich in high molecular multimers and for this reason it may also be used in some acquired VWD cases.

Treatment options for heavy menstrual bleeding

WHAT ARE THE MEDICAL TREATMENT OPTIONS FOR HEAVY MENSTRUAL BLEEDING?

HORMONE THERAPY

Tranexamic acid and hormonal therapy are considered to be first line therapy for women and girls with heavy menstrual bleeding. Hormonal therapy may take the form of a pill that combines estrogen and progesterone hormones (it is also commonly referred to as a contraceptive pill). Alternatively, hormone therapy may be administered by a skin patch or vaginal ring although this is less common. Hormone therapy works by helping to control anovulatory bleeding and uterine shedding so it is very effective and should be considered as a first line of treatment for all types of VWD. The estrogen component also has an effect on increasing the coagulation factor VIII which can decrease bleeding and although this is not its main mechanism, it may explain why some individuals with type 2 and 3 may have less benefit.

High dose progesterone derivatives can also be used in certain situations but their effectiveness in controlling uterine blood loss is somewhat unpredictable as is progesterone delivered in the form of a small device inserted under the skin in the arm. Low dose oral progesterone is generally not helpful. However, progesterone delivered directly to the uterine lining through the placement of an IUS (levonorgestrel-releasing intrauterine system). It is very effective and as such many guidelines recommend an IUS as a first line of treatment. The benefits of such therapy should be discussed with gynecologists with knowledge in bleeding disorders. Some examples of brands of IUS are the Mirena and the Kyleena. The IUS is a small, flexible device that is inserted into the uterus through the vagina. It releases low amounts of hormones (progesterone) locally and in many cases, this reduces the amount of blood lost during a menstrual period. It must be inserted by a gynecologist or other specially trained health care provider and can last up to approximately 7 years. It is important that the placement is checked a few months after insertion to be sure it is in the right place.

Advantages

- Insertion can be done in a clinic and does not require an operation.
- It can last for many years.
- It is an effective method of birth control, however, pregnancy can still occur after it is removed.
- Individuals do not have to remember to take pills on a regular basis.
- It does not contain estrogen so can be used in women and girls at risk of blood clots or who have experiences with blood clots.

Disadvantages

- Some people experience discomfort and cramping during insertion. Menstrual periods can be irregular for the first 3-6 months after insertion. Prolonged spotting may also occur.
- Some studies are conflicting but there may be a higher rate of expulsion and malposition in bleeding disorders. Doctors may recommend preparation with Tranexamic Acid (TXA) prior to insertion and for the following period thereafter.

Other hormone therapies may be prescribed when hormonal therapy does not work well. These include a GnRH analogue to shut down the hormones of the ovary at the level of the brain.

Although the use of hormonal therapy in the form of a combined estrogen and progesterone or progesterone only pill (sometimes referred to as oral contraceptives) can be very effective, for some adolescents it can raise some difficult issues.

- Some parents may not want their adolescent to take this form of hormone therapy, fearing it will lead to early sexual activity. However, studies have shown that therapeutic use of this hormone therapy 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 may hesitate to go against the parents' wishes, even when hormone therapy may be the only effective way to control heavy menstrual bleeding.
- Cultural differences significantly impact attitudes toward hormone therapy, driven by factors including religious beliefs, socioeconomic status, historical context, and gender norms. These factors influence not only the acceptance of hormone therapy but also knowledge, access, and consistent use across different populations.
- Parents and adolescents also fear use of hormone therapy may lead to cancer, infertility, blood clots or stroke, although the risks are small. These concerns can be lessened with accurate information.

The medical team at the hemophilia/bleeding disorder treatment centre can help families work through these issues.

Concerns also arise regarding the idea of placement of a hormonal IUS in an adolescent. These concerns should be raised with a pediatric gynecologist, however, the procedure is considered safe and effective and should be considered as an upfront treatment choice even in adolescents. Local anesthesia may be required for placement in some adolescents.

Non-hormonal treatments

ANTI-FIBRINOLYTIC AGENTS (TRANEXAMIC ACID)

Tranexamic acid (Cyklokapron) can be very effective for treating heavy menstrual bleeding. It can be started on the first day of menstrual bleeding and taken for heaviest days or the entire period. It can be combined with the use of combined estrogen and progesterone or progesterone only pills.

What are the other gynecological treatment options for heavy menstrual bleeding?

For some individuals, medical treatments alone will not work. Heavy, prolonged bleeding during the menstrual cycle will continue. For these situations, gynaecological procedures, including surgery, may be considered. It is important to have all the information before making decisions.

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. Various techniques to obliterate the uterine lining can be used, such as thermal or laser. Hormone therapy can be 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, so no hospital stay is required provided everything goes as planned.
- The recovery time is much shorter than with a hysterectomy.
- The success rates are high.

Disadvantages

- Unlike with medical treatment or the IUD, the individual will no longer be able to have children.
- The operation may have to be repeated.
- For a small percentage of people, 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 due to heavy menstrual bleeding even before testing for von Willebrand disease or other bleeding disorders. This unfortunately means that the person loses the ability to have children, when their bleeding could be successfully treated without the need for such an invasive procedure and there can be greater risks of bleeding with undiagnosed VWD. Hysterectomy should be reserved as a last resort treatment for women who have failed other therapies/treatments and for those who do not wish to have future pregnancies.

Advantages

- Hysterectomy completely stops menstrual bleeding.
- It may be the only option for individuals who do not respond to medical treatment or the IUD, and for whom endometrial ablation is not effective.
- Individuals can do well with proper planning.

Disadvantages

- Unlike with medical treatment or the IUS, the person can no longer have children.
- A hysterectomy is a major operation and the risks of bleeding are increased for people with VWD both during and after the operation. This can be managed with hemostatic agents and/or FVIII/VWF concentrates.
- The operation requires a stay in hospital.
- The recovery time is much longer than with an endometrial ablation.
- If there is a full hysterectomy (removal of the uterus and the ovaries), long-term hormone therapy is required.

LAPAROSCOPIC SURGERY (FOR ENDOMETRIOSIS, OR 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 for individuals who do not respond to hormone therapy or other medical treatment.

Disadvantages

- While not a major operation, a person with VWD may need appropriate preparation with hemostatic agents and/or FVIII/VWF concentrates.
- May require a hospital stay.

OOPHORECTOMY (REMOVAL OF ONE OR BOTH OF THE OVARIES)

The purpose of this operation is to stop bleeding from an ovarian cyst, however it is rarely done. This bleeding may happen even though:

- a person is taking hormone therapy to reduce heavy menstrual bleeding
- 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.
- Bleeding may occur and thus requires appropriate preparation with hemostatic agents and/or FVIII/VWF.
- Individuals may no longer have children (if both ovaries are removed).
- Hormone replacement therapy is required.

Not recommended treatment

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 or in the case of a miscarriage, however, it is unlikely that it will be effective in reducing heavy menstrual bleeding. In fact, the opposite may be true. The D&C will remove any existing platelet plugs and fibrin clots can make the bleeding worse. In addition, the endometrial tissue can grow back. Like for the other invasive procedures mentioned above, due to the risk of bleeding, hemostatic preparation is also required.

Key questions to discuss with your medical team about treatment options

These are some of the key questions to ask when you discuss treatment options with your health care providers.

How long has the treatment been in regular use?

Is the treatment ...

1. a prophylactic therapy (a prophylactic treatment is taken on a routine schedule to prevent bleeding)?
2. a therapy to stop active bleeding (on-demand)?
3. both?

How is the treatment administered:

1. intravenously?
2. subcutaneously?
3. orally (a pill)?
4. in some other way?

Can the treatment be administered at home?

How easy is the treatment to administer? How long does it take?

Does the product need to be refrigerated? How long can it be at room temperature?

How well does the treatment work?

- How quickly does it stop bleeding?
- Is it used for prevention of bleeding (prophylaxis) only, or can it be used to treat bleeding too?
- How many bleeds does a person have in a year, on average, if on prophylaxis?
- What physical activities might a person be able to do or not do when on this treatment?

What safety concerns or complications have been reported?

- Are there any side effects to be aware of?
- Were there any issues with unusual blood clots (venous or arterial thrombosis) while on treatment?
- How did they happen?
- How were they treated?
- Are there therapies that should be avoided while on this treatment?

Are there any limitations on who can receive the treatment? Age limits? Disease severity? Other medical conditions that would stop someone from receiving this treatment?

Available treatments for VWD are increasing and will likely continue to increase over time. Bleeding Disorders Canada (formerly the Canadian Hemophilia Society) has an online booklet, All About Novel Therapies, that is a guide to these emerging therapies. Its goal is to provide people with bleeding disorders and their caregivers a basic understanding of novel therapies. This will help in shared decision-making with their health care providers when discussing and making treatment choices. As treatment options are changing at a rapid pace, this booklet will be regularly updated. You can access that guide on the Bleeding Disorders Canada website – Novel Therapies Education Program | Hemophilia

Living with von Willebrand disease

COMPREHENSIVE CARE

What is comprehensive care?

Comprehensive care is: all the medical services needed by a person with VWD and their 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 an individual with VWD can receive all the care they need at one time. It is called comprehensive care because it offers a complete range of services. The following people work there:

- **The medical director. This doctor is often a hematologist who specializes in the area of blood clotting who:**
  - prescribes the lab tests to find out the exact bleeding problem
  - prescribes or recommends the proper treatment to control and prevent bleeding
  - monitors the overall health of the person with VWD.
- **The nurse coordinator is the front-line person in the clinic. The nurse coordinator:**
  - 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?***" on page 34.)
  - organizes the delivery of blood products for home use
  - coordinates appointments with other members of the comprehensive care team.
- **The obstetrician/gynecologist:**
  - works with the hematologist to prevent and control heavy menstrual bleeding
  - helps to avoid complications during pregnancy and childbirth.

- **The dentist:**
  - provides dental care
  - works closely with the hematologist to prevent bleeding during dental work.
- **The physiotherapist:**
  - bone and musculature system to make sure the joints and muscles stays healthy and 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
  - Helps women/girls understand pelvic health and function in relation to their bleeding disorder.
  - Advocates for an orthopaedic consult if indicated.

The comprehensive care team may include other individuals as needed, such as:

- a social worker (to help families deal with the challenges of a bleeding disorder)
- a genetic counselor (to give information to carriers)
- an orthopedic specialist (for joint problems and joint surgery)
- laboratory specialists.

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.

Key questions to consider:

- Is treatment needed often enough to justify home care?
- Is the person with VWD or their caregiver willing to take on the responsibility?
- Is the person with VWD or their 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, such as:

- 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.

For families of children with Type 3 VWD who are on prophylaxis, they 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. 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. Some younger children may have permanent venous access such as ports inserted.

Desmopressin can also be administered at home. However, it has a limited shelf life and needs to be refrigerated. People using it at home should carefully check expiry dates.

Tranexamic acid (Cyklokapron) is a drug 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.

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.

Factor concentrates used today have an excellent safety record. The plasma used for FVIII/VWF concentrate is screened for blood-borne viruses such as HIV, hepatitis A, B and C, and parvovirus B19. 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. Some viruses, such as parvovirus B19 or hepatitis A, are more difficult to remove or inactivate. Parvovirus B19 may most seriously affect pregnant women, or immune-compromised individuals. While there have been rare reports of these infections associated with the use of some plasma-derived products, the overwhelming numbers of hepatitis A and parvovirus B19 cases are acquired in the community, and not through blood products.

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 the person be tested for HIV and hepatitis C?

Yes. If a person received blood or blood products in the past, the individual 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.

NOSEBLEEDS

Are nosebleeds common in von Willebrand disease?

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

How can nosebleeds be stopped?

Nosebleeds 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. Tranexamic acid is often given in this situation. The nose can also be packed by a specialist or desmopressin can be taken.

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

Anti-fibrinolytic agents (Tranexamic acid/Cyklokapron) can be given for 5 to 7 days after the nosebleed to prevent re-bleeding.

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

When should a doctor be consulted for a nosebleed?

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. If nosebleeds are recurrent then the person could have tranexamic acid at home.

Can anything be done to prevent nosebleeds?

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

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 topical tranexamic acid may be needed to prevent bleeding from re-occurring. The medical team at the hemophilia/bleeding disorder treatment centre will be able to help with these treatments.

Can women with VWD have children?

Yes. For most women with VWD, conception is not a problem. Of course, hormone therapy to control heavy menstrual bleeding interferes with conception so this treatment must be adapted with a clear plan once trying to conceive. Ideally, the person should speak to the specialized medical team when planning a pregnancy.

Pregnancy is often a time with few bleeding problems due to the level of VWF in the blood. Fortunately, the level of VWF in the blood for most individuals 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. The increase in VWF in pregnancy is less predictable in Type 2 and therefore levels need to be followed in pregnancy. There is no expected increase in VWF factor levels in Type 3 VWD. Those with both types may experience increased risk for both mucosal bleeding and gum bleeding during pregnancy. Additionally, more precautions may be necessary.

What kinds of doctors are important for a person with von Willebrand disease during pregnancy?

It is very important to have confidence in your medical team during pregnancy. This team should include:

- an obstetrician (who specializes in pregnancy and delivery)
- a hematologist (who can ensure any bleeding issue is under control and who will monitor the evolution of the VWF levels in order to plan the childbirth)
- an anaesthesiologist (who specializes in bleed abnormalities)
- a pediatric hematologist (who can ensure the safe delivery of baby until diagnosis can be determined)

All necessary physicians and specialists should together create a birth plan. This birth plan encompasses procedures for a safe labor and delivery for both mother and baby and should be accessible to anyone supporting and caring for mother and baby during birth. Often the birthing plan is made in the third trimester when most VWF reach their peak in pregnancy.

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

Bleeding in the first trimester may be more common in people with VWD, however, recent data does not support an increase in miscarriage rate although very few people with VWD type 3 have been included in these studies due to its rarity. Appropriate precautions and monitoring should be offered to all women with VWD who are bleeding in the first trimester and appropriate treatment ensured.

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

Is bleeding more common during pregnancy than at other times?

No. In fact, for individuals with Type 1 VWD the opposite is true. They often 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 individuals with Type 1 VWD have few bleeding problems during pregnancy and immediate bleeding following childbirth is only slightly increased compared to people without VWD. However, most women must be followed and monitored for delayed post-partum hemorrhage that may begin after 24 hours and that can even worsen in the 2 weeks following delivery as VWF levels fall back to their baseline levels before pregnancy.

Nevertheless, clotting levels should be monitored, especially as the date of delivery approaches. This way, doctors will know whether to prepare treatments. It is important to alert the hospital so the hospital blood bank can have the factor concentrates on hand if needed.

Desmopressin can also be prescribed to raise VWF and FVIII levels during delivery for women with for individuals with Type 1 VWD although caution is required due to the higher risk of volume overload and hyponatremia (low sodium) in pregnancy. The risk and benefit of this treatment must be considered and discussed with the medical team.

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 may not be corrected. Platelets may drop lower in individuals with type 2B VWD due to the higher circulating levels of VWF, which bind and subsequently clear platelets. The levels of VWF will not rise for women with Type 3 VWD because they do not make any VWF and thus will require FVIII/VWF replacement.

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 or presumed effective in cases where a desmopressin trial has not been done)
- anti-fibrinolytics (tranexamic acid/Cyklokapron)
- FVIII/VWF concentrates.

It should be assumed, unless prenatal testing has shown the opposite, that the fetus has a 50% chance to be 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 an individual with a bleeding disorder.

To prevent bleeding, the following should be avoided, when possible:

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

Bleeding is a serious but rare complication of an epidural anesthesia (freezing of the lower body by means of a needle in the spine) in individuals with severe forms of VWD. Before using epidural anesthesia, the anesthesiologist should consult an experienced hematologist. Treatment with FVIII/VWF concentrate may be necessary. In some cases the doctor may recommend an alternative pain control method when the risk of an epidural is considered too high even with factor concentrate.

For those people whose VWD levels do not correct in the third trimester or in people with Type 3 VWD, factor replacement is likely required before a cesarean section and sometimes for vaginal delivery.

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

Postpartum bleeding 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:

- Hemoglobin and iron levels.
- VWF and factor VIII levels (in more severe cases)

If bleeding post-partum is heavier than a menstrual cycle, and/or with any of the features of heavy menstrual bleeding listed above, she should immediately notify her obstetrician or hematologist. Tranexamic acid may be prescribed for home use to treat heavy bleeding. Anemia postpartum may be present and should be monitored closely. Treatment options may include IV iron or as a last option, red blood cell infusions. Ongoing oral iron may also be prescribed.

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 (postpartum). 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 or iron infusions, may be necessary and homecare could be organized for the mother.

Babies with VWD rarely bleed at birth. However, babies with more severe Type 1 VWD, Type 2 VWD and most Type 3 VWD, may bleed if they undergo surgery, including circumcision. There is also the risk of ICH (Intracranial hemorrhage) because of FVIII levels potentially being lower than 1%. It is crucial that the hematologist be advised of any need for surgery required for the baby.

Is prenatal testing of the fetus necessary?

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

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

For severe forms of VWD it is easy to diagnosis at birth on cord blood, however, those that are not severe is harder and so if cord blood is tested at birth, it is typically repeated 6-12 months of age to confirm the accuracy of levels. It can be difficult to diagnose milder forms of VWD in babies, therefore, discuss the timing of testing with the medical team to determine what is the best time for your specific situation.

MEDICATIONS TO AVOID

Nonsteroidal anti-inflammatory drugs (NSAIDs) can act as mild blood thinners by making platelets less sticky, which interferes with blood clotting. These medications include Aspirin® (also called acetylsalicylic acid or ASA), ibuprofen (Advil®, Motrin®) and Naproxen (Aleve®). Low-dose 81 mg ASA may be needed in some patients for heart or blood vessel health. However, taking full-dose NSAIDs for headaches, fever or muscle aches may increase the risk of bleeding in people with VWD. Before taking any NSAIDs, talk to your treatment centre physician to discuss your health profile, and the risk benefit balance of taking these medications.

Some cold medicines and antidepressants can also interfere with clotting. Always check with your treatment centre team and/or pharmacist to make sure a drug is safe.

Herbal remedies can offer helpful complementary care in some cases. However, it is important to recognize that "natural" does not mean "safe." Some herbs may have an effect on bleeding. Before you decide to try an herbal remedy, get advice from your treatment centre team.

SAFE TREATMENT OPTIONS FOR PAIN MANAGEMENT

It is important to assess and evaluate the cause of joint and muscle pain. Strategies such as use of heat and transcutaneous electrical nerve stimulation (TENS) can be helpful to lessen pain. In the case of a muscle or joint bleed, immobilization, compression or splinting may help minimize pain. Additionally, treatments such as DDAVP or factor replacement therapy might be appropriate depending on individual treatment plans. Physical activity should be restricted until pain resolves. Exercises and/or physical therapy should begin after pain subsides to gradually restore muscle strength and mobility.

For muscle tension, massage therapy can be soothing and relaxing. Topical anti-inflammatory creams (even if they contain salicylates, like Voltaren®) and ointments are safe to use for muscle and joint-related pain relief, with no impact on blood clotting. Acetaminophen (Tylenol®) is an effective and safe treatment for pain and fever. Acetaminophen does not affect blood clotting and does not increase the risk of bleeding in people with VWD. Celecoxib (Celebrex®) is an anti-inflammatory medication that does not block platelet function, and can be effective for managing joint and muscle pain in people with VWD. However, it does require a prescription. If you have persistent pain which is not resolving, please contact your treatment centre team to discuss your symptoms.

Can a person with von Willebrand disease play sports?

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

Regular exercise can raise VWF levels, resulting in more protection to joints and muscles.

A person with VWD will have to determine what physical activities to participate in. Many people with a mild disorder participate in all kinds of sports including active sports like soccer and high-risk sports like skiing. It is recommended to have conversations with the medical team to create individualized plans and tailor therapies to assist with desired activities. People with Type 3 VWD or severe bleeding subtypes, may find these activities lead to serious bleeding and are encouraged to avoid contact sports and dangerous activities that may lead to head injury.

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 activities they enjoy and those of their 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. Injuries are bound to happen. Injuries can help a child to learn more about what they handle, learn from the mistake/accident, how their body heals, and to think of risks and benefits of their activity choices. Risk taking, within reason, is part of a child's development to allow them to be more resilient.

Specialists at the hemophilia/bleeding disorder treatment centre can advise a person of the risks based on an evaluation of the bleeding disorder. However, in the end, the person living with VWD is the best judge of which activities are appropriate.

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

No, it should not have an impact. 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 and preventative therapy, these absences should be minimized.

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 bleeding disorder. 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 nosebleeds. Children with VWD need to be taught at a young age how to handle their own nosebleeds. If the children are very young, the child's teacher or daycare worker will need to learn how to handle them. (See "***Nosebleeds***" on page 38.) 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.

It is important that daycare or school personnel can always contact the parents, 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, such as combative 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. 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 the child's bleeding disorder?

No, unless the child wants to share with their classmates about VWD, there is no need to inform 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 manage their bleeding disorder, 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.

TEENS AND SEXUALITY

Adolescents may begin to experiment with sexuality. Teens should be encouraged to talk openly with the medical team about potential bleeding concerns during sexual activity so they can be informed and know how to manage any bleeding episodes appropriately.

EMPLOYMENT

A person with VWD can do any job.

A 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 any medications, needles and syringes you are carrying.

- Take along the Travel Card from Bleeding Disorders Canada (formerly the Canadian Hemophilia Society) that contains information about how to access care and treatment when away from home. Among other things, the wallet card contains 16 key phrases in English, French and Spanish that can be used in an emergency situation. To obtain a copy of the travel card, please contact Bleeding Disorders Canada.
- Make sure your, or your family member's, MedicAlert bracelet is up to date.
- If you, or your family member, self-inject desmopressin or self-infuse 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.
- A letter from your hematologist explaining your medication and why it is imperative or stay with you, with their contact information on hospital letterhead

Should a person with von Willebrand disease carry medical identification?

Yes. Canadian hemophilia/bleeding disorder treatment centres provide *FactorFirst* cards to many of their patients. The *FactorFirst* card contains basic information on treatment of bleeding disorders, specific information on an individual's particular condition, and contact information for the person's treatment centre. If you do not have one, ask about it at your treatment centre. Similar to a *FactorFirst* card, there is also a *TreatFirst* card for people who use other treatments besides factor.

It is also recommended that a person with VWD wear a MedicAlert bracelet or necklace. Von Willebrand disease is not well known and not easily diagnosed. In the case of an injury or other emergency, the *FactorFirst* card, *TreatFirst* card and MedicAlert 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.

Final word

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
- be in a partnership or 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 bleeding disorders.
- Live life to the fullest, knowing that von Willebrand disease can be successfully managed.

Glossary

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.

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.

antifibrinolytics
Drugs (tranexamic acid/Cyklokapron) 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.

collagen
The structural protein for body tissues. VWF binds to this structure and platelet for clotting.

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 in Canada.

cyklokapron
An antifibrinolytic 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. Due to the fact that the D&C will remove any existing platelet plugs and fibrin clots, it is no longer recommended for women with bleeding disorders.

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.

hematologist
A physician specializing in diseases of the blood.

hematomas
Bleeds into tissues or muscles.

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.

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

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.

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

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

intrauterine system
A small, flexible device that is inserted into the uterus. It releases low levels of hormone locally in order to reduce the amount of blood lost during menstrual periods.

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.

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.

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.

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

Contact information

For further information, please contact:

Bleeding Disorders Canada (formerly the Canadian Hemophilia Society)
National Office
400-1255 University Street
Montreal, Quebec H3B 3B6
Tel: 514 848-0503
Toll-free: 1 800 668-2686
Fax: 514 848-9661
info@bleedingdisorderscanada.ca
bleedingdisorderscanada.ca

For a complete list of all the Canadian hemophilia/bleeding disorder treatment centres, please go to the Bleeding Disorders Canada Web site at
bleedingdisorderscanada.ca.

Produced by
Bleeding Disorders Canada