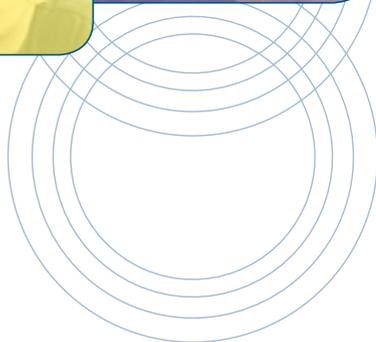
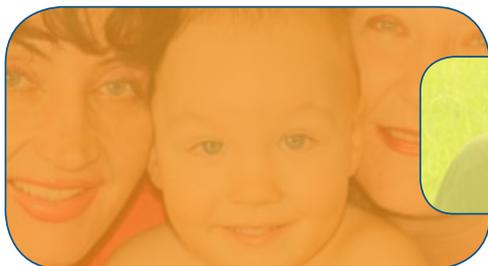


A guide for people living with von Willebrand disorder



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WHAT IS VON WILLEBRAND DISORDER (VWD)?

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

VWD was named after the Finnish doctor Erik von Willebrand who discovered the condition and published his findings in 1926. Dr von Willebrand described this new type of bleeding disorder after observing the bleeding problems of a large family who were living on one of the Åland Islands between Sweden and Finland. VWD is also known as *von Willebrand disease*.

HOW COMMON IS VWD?

VWD is the most common inherited bleeding disorder worldwide. It affects both males and females from all racial backgrounds.

Most people with VWD are born with the disorder as VWD is inherited genetically. Sometimes VWD will show up when the person is a child. Others don't find out until they are adults and have a bleeding problem, or until a relative is diagnosed and it is suggested that they are tested as well.

It is thought that up to 1 in 100 people have VWD, but most people will have few symptoms. Many people with VWD may not know they have the disorder because their bleeding symptoms are very mild.

Research suggests that many people with VWD have not yet been diagnosed.

Usually VWD is less severe than other bleeding disorders, such as haemophilia. The form causing moderate bleeding problems is uncommon, and the severe form of VWD is rare.



HOW SERIOUS IS VWD?

It depends on the type of VWD and the level of von Willebrand factor in the person's blood. Most people have such mild symptoms that they are not aware they have the disorder. Others only realise they have a bleeding problem when they have heavy bleeding after a serious accident or a dental or surgical procedure.

However, with all forms of VWD there can be bleeding problems. Some people with VWD bleed quite often, eg with nosebleeds, bruising and heavy periods. A smaller number of people have the severe form of VWD and may also experience joint and muscle bleeds, similar to haemophilia.

There is no cure for VWD. It is a lifelong condition, usually with mild symptoms.

There are safe, effective treatments for all types of VWD.

(see What is the treatment for von Willebrand disorder?)

Haemophilia

Although VWD is more common, haemophilia is often better known. Haemophilia is a rare inherited genetic bleeding disorder. It occurs when an essential blood clotting factor - factor VIII (8) or IX (9) - is missing in a person's blood or doesn't work properly. Bleeding is mostly internal and can cause muscle and joint damage. To stop or prevent bleeding episodes (or "bleeds"), people with haemophilia can be treated with replacement clotting factor products, which are injected into a vein. Although both men and women can carry the haemophilia gene and pass it on to their children, usually only males with the gene have symptoms of haemophilia.

SYMPTOMS



The symptoms of VWD vary greatly from person to person. Even members of the same family may have different symptoms.

Most people with VWD have few or no symptoms.

The more common symptoms are:

- Having nose bleeds often or that are difficult to stop
- Easy bruising
- Very heavy or long menstrual periods
- Bleeding for a long time with minor cuts
- Bleeding from the gums
- Bleeding after injury, surgery or dental work that continues for a long time.

Bleeding in people with VWD usually involves the mucous membranes, the delicate tissues that line body passages such as the nose, mouth, uterus, vagina, stomach and intestines.

Less common symptoms that older people might experience are:

- Blood in faeces (bowel motions/poo) from bleeding in the intestines or stomach
- Blood in urine from bleeding in the kidneys or bladder.

People with severe forms of VWD, particularly type 3 VWD, may also have other bleeding problems similar to haemophilia, such as:

- Bleeding episodes that are spontaneous or happen for no obvious reason
- Bleeding into joints and muscles which can cause swelling and pain.

The types of symptoms a person with VWD experiences can change over their lifetime. For example, they may have nosebleeds and easy bruising as a child and find this occurs less often as they grow older. However, their type of VWD will not change.

We all have VWD in my family – some have minimal symptoms, some mild, some severe. All have had issues with blood noses, but seem to have grown out of the nasty blood noses that used to go on for hours. All have bruising which varies and our youngest daughter has had problems with periods.

Women are more likely to show symptoms of VWD than men. Without treatment, women with VWD often bleed more or for longer than normal with menstruation (their period). Some women with VWD also have heavy bleeding a few days or weeks after giving birth and some have a lot of period pain or irregular periods. However, these symptoms are not always related to VWD and may have other causes. An assessment by a gynaecologist is an important part of understanding and treating these symptoms effectively.

TYPES OF VWD

There are three main types of von Willebrand disorder. Bleeding symptoms can vary from person to person within each type.

- **Type 1 VWD** is the most common form. Around 80% of all people with VWD have this form. In type 1 VWD, the von Willebrand Factor (VWF) works normally, but there is not enough of it. Symptoms are usually mild, depending on the level of VWF in the blood.
- In **type 2 VWD**, the amount of VWF in people's blood is often normal but the VWF doesn't work properly. Type 2 VWD is divided into subtypes 2A, 2B, 2M and 2N. Certain subtypes may be treated differently, which makes knowing the exact type of VWD you have very important.
- **Type 3 VWD** is very rare. People with type 3 VWD have very little or no VWF in their blood. Symptoms are more severe and can also include joint and muscle bleeding. Bleeding can occur more often.

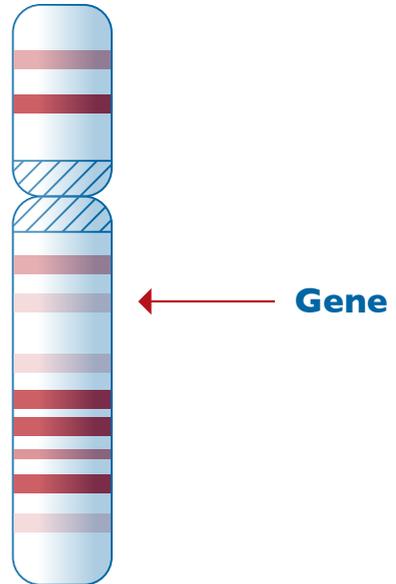
HOW DO YOU GET VWD?

Von Willebrand disorder (VWD) is usually inherited.

The way each of us makes von Willebrand factor (VWF) in our body is coded into one of our genes and has been inherited from genetic information from both our mother and father. This gene is called the VWF gene. Genetic information or coding in our genes guides how our body grows, develops and works.

If there is an alteration causing VWD in the gene, this may then be passed down from parent to child, in the same way as other genetic information like the colour of their hair or their eyes. The altered gene is often called a VWD gene. A parent can pass on the altered VWD gene even if they don't have symptoms.

All of us have two copies of each gene, one inherited from each parent. Genes are carried in our chromosomes, which are tightly coiled strands of genes located in the centre of the cells in a human body. The VWF gene is located on an ordinary chromosome (autosome), not on a sex chromosome, like haemophilia. Sex chromosomes decide whether we are male (XY) or female (XX). This means that VWD affects males and females in equal numbers, unlike haemophilia which usually only occurs in males – so both sons and daughters can inherit VWD. Their symptoms can be different to their parent's or to each other.

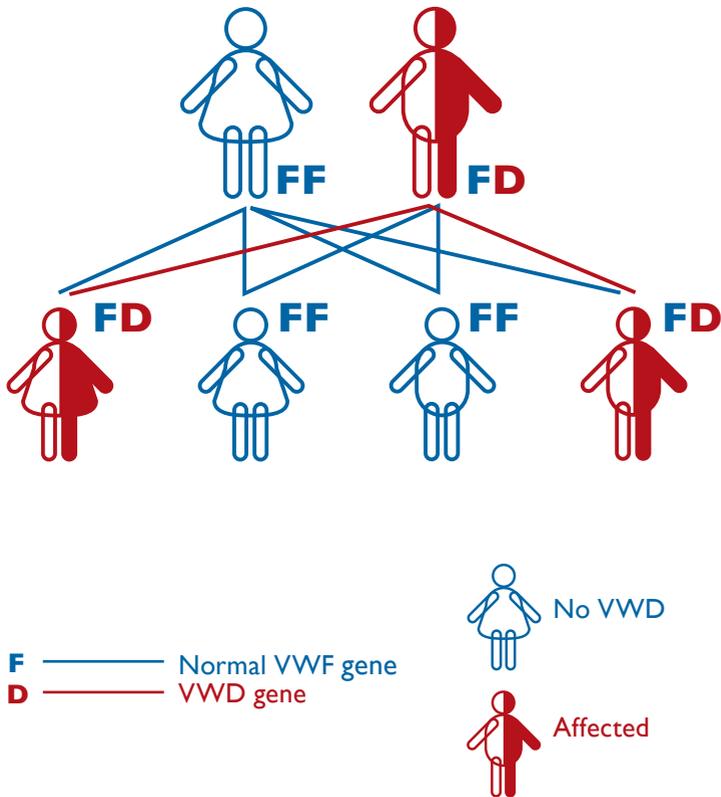


Chromosome

HOW IS VWD INHERITED?

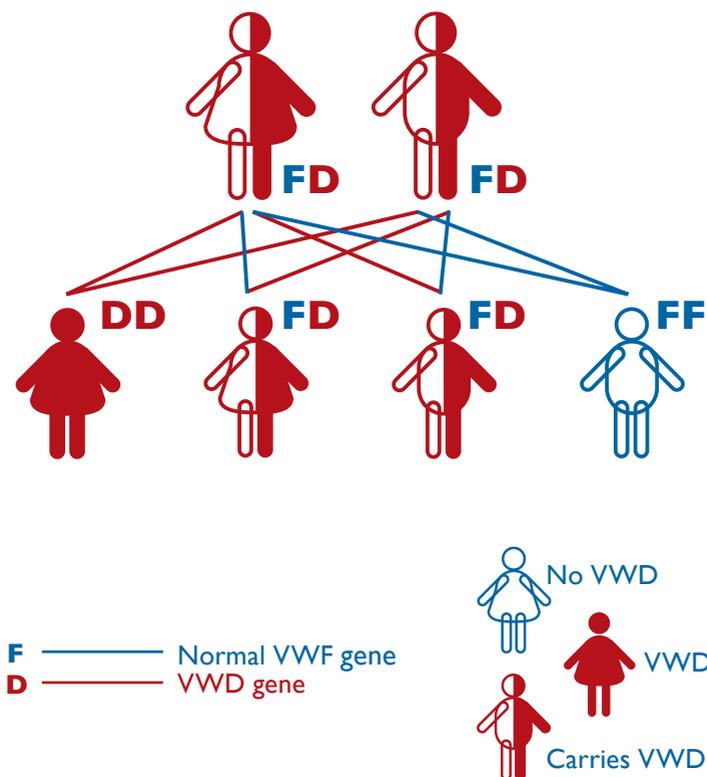
There are two main inheritance patterns for VWD:

Inheritance of von Willebrand disorder type I, 2A, 2B, 2M



In most type I and type 2A, 2B and 2M VWD, the VWD gene is *dominant*. This means that if one parent has a VWD gene, they have a one in two (50%) chance of passing the gene on to each of their children. They or their children may or may not have symptoms.

Inheritance of von Willebrand disorder type 2N and 3



In type 3 and type 2N and some type 1 and 2A VWD, the VWD gene is recessive. If both parents carry this type of VWD, they may not have symptoms but there is a one in four (25%) chance that their children could inherit a copy of the VWD gene from both of them and have symptoms, usually severe. There is a one in two (50%) chance that their children will inherit only one copy of the VWD gene from them and carry the gene but not have symptoms, like their parents. There is also a one in four (25%) chance that their children will not inherit the VWD gene at all.

Source: Adapted from Goodeve AC, James P. Von Willebrand disease. GeneReviews 2009 Jun 04, viewed 15 March 2010. <http://www.ncbi.nlm.nih.gov>

If one member of a family is diagnosed with VWD, the doctor may recommend testing other members to see whether they have VWD as well.

My mother was diagnosed with Von Willebrand's about 15 years ago after an operation and advised my sister and me to get tested. It turned out that I have Type 1 but my sister doesn't.

In my family it seems as though everybody has it! Myself and two of my three brothers were diagnosed at birth, and recently we found out that my oldest brother also has it, although much more mildly. Since I have a more severe form, it's likely that my mum is also a carrier – she doesn't experience any bleeding problems herself, but her own mother was a severe bleeder.

My husband found out he had VWD in his thirties after a minor haemorrhage having a wisdom tooth out. He'd always had lots of little bruises here and there, but had had no other bleeds following things like tonsillectomy as a child.

No family history of VWD

Sometimes there is no family history of VWD. A baby can have a *genetic mutation*, or change in one of their genes, before they are born. Although their parents and their brothers and sisters do not have the VWD gene, the child will be able to pass the VWD gene on to their own children in the future.

CAN YOU ACQUIRE VON WILLEBRAND DISORDER?

Some people develop a form of VWD later in life due to other medical problems that affect their von Willebrand factor. This is called Acquired von Willebrand Syndrome (AVWS) and is very rare.

VWD AND BLOOD CLOTTING

How does blood clot normally?

Blood circulates throughout the body in a network of blood vessels. When body tissue is injured, damage to a blood vessel can cause blood to leak through holes in the vessel wall. Blood vessels can break near the surface, eg in a cut. Or they can break deep inside the body, making a bruise or an internal haemorrhage.

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

Normal blood clotting stages



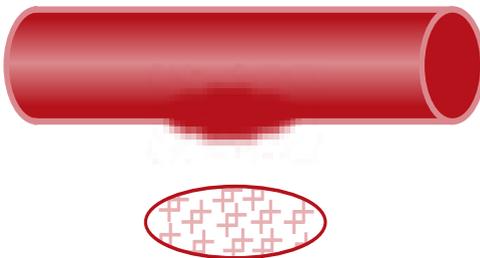
Stage 1: The blood vessels constrict to slow the flow of blood to the injured area.



Stage 2: Platelets, small cells circulating in blood, stick to and spread on the walls of the damaged blood vessel.



Stage 3: Von Willebrand factor (VWF) acts as a glue to hold the platelets in place where the damage to the blood vessel occurred.



Stage 4: The platelets then make a surface for blood clotting to take place. Clotting proteins circulating in the blood gather on the surface of the platelets to make a mesh-like clot, like a scab.

Normal von Willebrand factor (VWF)

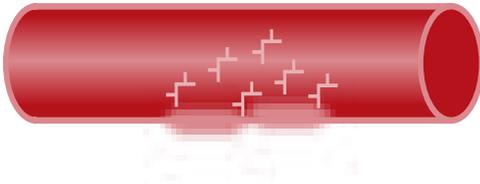


How does VWD affect the normal clotting of blood?

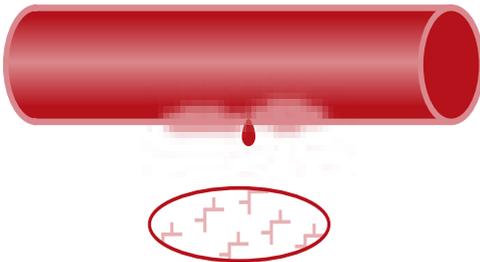
Von Willebrand disorder (VWD) affects the last three stages in the blood clotting process.



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



Stages 2 and 3: A person with VWD may not have enough von Willebrand factor (VWF) in the blood or it may not work properly. Because of this the VWF cannot act as a glue to hold the platelets in place where the damage to the blood vessel occurred. The platelets do not stick to the lining of the blood vessel.



Stage 4: VWF carries factor VIII (8) in the bloodstream. Factor VIII is one of the proteins needed to make a solid clot. If the factor VIII levels are also low, a solid clot may take many days to form.

Abnormal von Willebrand factor (VWF)

Source: Adapted from Page D. All about von Willebrand Disease... for people with von Willebrand Disease and their families. 2nd ed. Montreal: Canadian Hemophilia Society, 2007.

DIAGNOSIS

VWD can be difficult to diagnose and repeated testing may be needed to confirm the diagnosis.

Many people's symptoms are mild and they may not be diagnosed until they have a major bleeding problem, eg from surgery or an injury. However, if they have a severe form of VWD, they will usually have major bleeding problems as a baby or small child and will often be diagnosed within their first year of life.

Diagnosing VWD involves:

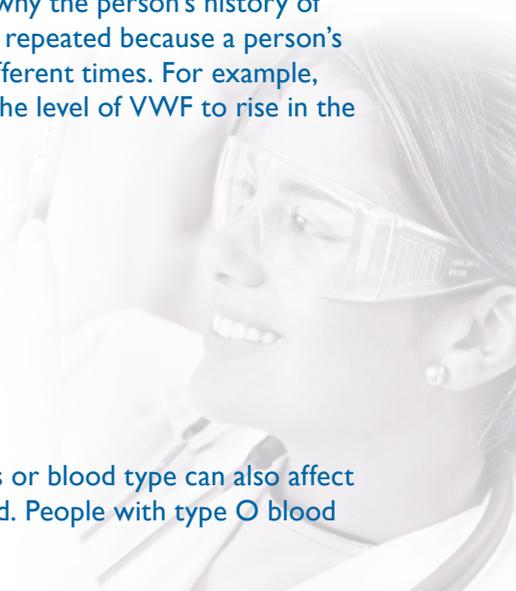
- A personal history of bleeding or bruising more than normal from mucous membranes or skin after injury, trauma or surgery **and**
- A family history of bleeding more than normal **and**
- Specialised laboratory test results for VWD.

Tests for VWD

The doctor may order several laboratory tests on blood samples:

- **Von Willebrand factor antigen:** to measure the amount of von Willebrand factor in your blood
- **Von Willebrand factor ristocetin cofactor activity** and/or the **collagen binding assay:** to show how well the von Willebrand factor works
- **Factor VIII clotting activity:** to measure how well the von Willebrand factor binds to the factor VIII protein and maintains the level of factor VIII in the blood. Some people with VWD have low levels of factor VIII, while others have normal levels
- **Von Willebrand factor multimers:** if some tests suggest you have VWD, this test is used to show the makeup or structure of the von Willebrand factor and helps to diagnose the type of VWD
- **Platelet function test:** to measure how well your platelets are working.

Understanding the laboratory test results is complex and needs to be done by a haematologist (specialist doctor) and laboratory with experience in VWD.



VWD can only be diagnosed with specialised blood tests. Routine blood tests often give normal results, which is why the person's history of bleeding is so important. Testing is often repeated because a person's VWF and factor VIII levels can vary at different times. For example, several common experiences can cause the level of VWF to rise in the blood and appear to be normal:

- Stress
- Exercise
- Pregnancy
- Normal hormonal changes during a woman's monthly menstrual cycle
- Hormone treatment.

Inflammation with other health problems or blood type can also affect levels of VWF and factor VIII in the blood. People with type O blood often have lower levels of VWF.

If you think you have a bleeding problem, it is important to see a haematologist who specialises in bleeding disorders. In Australia, these haematologists can be found at Haemophilia Centres or Services which are at some major hospitals. Talk to your general practitioner or your gynaecologist about a referral.

There are a variety of issues related to diagnosing VWD, including:

- Other family members may need to be tested for VWD
- Possible implications for employment, health insurance cover and carrying the VWD gene
- Decisions will need to be made, in consultation with your Haemophilia Centre or Service, about which treatment options will be the most suitable.

Haemophilia Centre social workers, counsellors and nurses are available to help people deal with these issues.

TREATMENT

Specialist Haemophilia Centres or Services have a team of health professionals with expertise in providing treatment and care to people with bleeding disorders including VWD. They can work with the person with VWD to make a treatment plan and advise on ways to live well with VWD.

The team includes:

- Haematologists: doctors who specialise in blood disorders
- Haemophilia nurses
- Social workers or counsellors
- Physiotherapists
- Other specialist health professionals

The recommended treatment for VWD can depend on the type of VWD a person has and how severe it is. Usually people with mild VWD will only need treatment if they have surgery, dental work or an accident or injury.

For some minor bleeding problems, like bruising, treatment may not be necessary.

If people do need treatment at times, there are several treatments available. Which treatments will be used on particular occasions will depend on the person's medical needs, what works best for them and the situation. The haematologist will consider all of this when they work with the person to decide the best treatment option.

Desmopressin (DDAVP) is a synthetic hormone. It works by releasing the body's stored VWF and factor VIII into the bloodstream to help blood clot. These stores are limited, so a person may need to wait for the body to rebuild its stores of VWF before taking another dose, usually about 24 hours.

Desmopressin can be given as a slow injection into a vein, but may also be given as an injection subcutaneously (into the fatty tissue under the skin), or in special circumstances as a nasal spray.

Desmopressin can help to prevent or treat bleeding in many people. It is not suitable for everyone. The haematologist at the Haemophilia Centre may decide to give a test dose of desmopressin and evaluate whether it will work for that particular person. This test may need to be repeated at times as people's responses to DDAVP change at different times in their life.

Clotting factor concentrate made with von Willebrand factor (VWF) and factor VIII (FVIII) replaces the missing VWF and FVIII in the blood and helps blood to clot. This clotting factor concentrate is made from the plasma (pale yellow fluid part) in human blood and is produced from blood donations.

This clotting factor concentrate is used when DDAVP is not suitable, or when it is likely the person will need treatment for more than 2-3 days, eg after major surgery. The treatment is infused (injected) into a vein in the arm.

Tranexamic acid and **aminocaproic acid** are medicines that stop blood clots being dissolved once they have formed. They can be used to stop bleeding in the mouth or nosebleeds, gut bleeding, bleeding after dental work, minor surgery or an injury.

Most commonly they are taken as tablets, syrup or as a mouthwash. They may be used by themselves or together with DDAVP or a clotting factor concentrate.

Fibrin glue is a medical gel made from fibrinogen and thrombin, which are proteins in the body that help blood to clot. It can be applied directly onto a wound to stop bleeding.

Hormone treatment, such as **oral contraceptives** (birth control pills), can help women who have heavy menstrual bleeding. The hormones can increase VWF and factor VIII levels.



Plasma-Derived Treatment Product Safety

VWD treatment product safety is a high priority for Australian regulatory authorities, blood bank services, manufacturers and the bleeding disorders community. Recombinant clotting factor products, which are genetically engineered and contain little or no human material, are not yet available to treat VWD and some people with VWD need to use clotting factor concentrates made from human blood plasma.

Manufacture of human plasma factor concentrates is carefully regulated and monitored to make sure the concentrates are now as safe as possible from infections that can be transmitted by blood:

- In Australia blood donors are screened and blood donations are tested for HIV, hepatitis B and C, human T-cell lymphotropic virus (HTLV) and syphilis
- Strict controls are applied when selecting blood donors to reduce the risk of transmitting infections, including vCJD (variant Creutzfeldt-Jakob disease, the human form of “mad cow disease”)
- When they are manufactured, human plasma factor concentrates are treated with several processes to remove or inactivate HIV and viral hepatitis and, as far as possible, exclude other known infectious agents which may be passed on by blood, such as the prion causing vCJD.

In Australia during the 1980s and early 1990s some people with bleeding disorders, including a small number of people with VWD, acquired hepatitis C, and some also HIV, through contaminated plasma clotting factor concentrates they used for their treatment. As a result a number of safety measures have been developed and put in place. Work to prevent the transmission of infections through blood products is ongoing. The cases of vCJD in the United Kingdom have highlighted a new infectious agent, the vCJD prion, and there has been considerable research to develop a reliable screening test and manufacturing processes to exclude vCJD. In Australia people who resided in the UK from 1980 to 1996 for more than six months are not permitted to donate blood.

The risk of new infections from using human blood products is now thought to be extremely low. In spite of this, the risk cannot be entirely excluded, particularly if the risk came from a new or unknown type of blood-borne virus or other micro-organisms causing disease. Because of this, people using these products and patient advocacy organizations such as Haemophilia Foundation Australia (HFA) continue to take a strong and watchful interest in product safety.

Hepatitis B vaccination is recommended for people with bleeding disorders who use plasma-based concentrates.

More information:

Australian Red Cross Blood Service – <http://www.donateblood.com.au>

Australian Government. Department of Health & Ageing;
National Health and Medical Research Council.

Australian immunisation handbook. – <http://www.health.gov.au>

TAKING CARE OF YOURSELF OR YOUR CHILD

Knowing as much as you can about your own or your child's health and what to do in particular situations can help with your confidence to enjoy life and its ups and downs, particularly if you have a plan for managing any health problems that may come along.

Diagnosis with VWD can be a confronting experience at first and all sorts of fears may run through your mind. This is a very normal experience and don't be afraid to ask the Haemophilia Centre team any questions, especially things that are worrying you, or to talk with them about your anxieties. If you haven't understood the answer, let them know – it's important to get proper explanations in language you understand.

Sometimes you might feel a bit confused, or overwhelmed by your thoughts and feelings, or just want to talk some things through in a non-judgemental and confidential environment, and might not be sure who to turn to. Many Haemophilia Centres have Haemophilia Social Workers or Counsellors who are very happy to help with this or can refer you to other counselling services. You may also find it useful to contact your local Haemophilia Foundation, who may be able to put you in touch with other people with VWD or parents of children with VWD.

Over time people with VWD and parents of children with VWD often find that they become experts on their own situation and can educate others. They work with their medical team and make decisions together. This can help to have a greater sense of control and choice in treating the condition and living a full and fulfilling life.

People without bleeding problems often don't understand what the experience is like for you or what it means – and that includes GPs, medical specialists, friends and family - so it's good to be informed and educated for your own sake, and so you can communicate your needs to others.



EDUCATING YOUR CHILD ABOUT VWD

Involving your child in learning about their bleeding disorder is an important part of their growth and development. As your child grows up, you can teach them about their type of VWD, the treatment they use, medications they should avoid, when to seek treatment, genetic inheritance and how to speak out and get help to achieve what they need. The type of information and how you teach it will depend on their stage of development – your Haemophilia Centre can help you with this.

It will be a gradual learning process throughout their childhood, which gives them an opportunity to understand and adapt to their VWD and take responsibility for managing their bleeding disorder. When they reach adulthood and take on the management of their own health, they will then have enough information and the confidence to make independent decisions about their health and life choices.

HOW DO YOU FIND OUT ABOUT VWD?

- Talk to your Haemophilia Centre team about VWD, including any questions and concerns, and what treatments and self-management strategies would be best for you or your child
- Talk to other people with VWD or parents of children with VWD
- Check the web sites of local and overseas Haemophilia Foundations for up-to-date and accurate information, eg:
 - Haemophilia Foundation Australia – www.haemophilia.org.au
 - Canadian Haemophilia Society - www.hemophilia.ca
 - The Haemophilia Society (UK) - www.haemophilia.org.uk
 - National Hemophilia Foundation (USA) - www.hemophilia.org
- Everyone's experience of VWD is different, so in some ways you will be your own best guide – over time you will become skilled at recognising your or your child's patterns.

SELF MANAGEMENT

All bleeding and bruising should be dealt with promptly to make sure no permanent injuries result. Some minor bleeding episodes can be managed at home without a visit to the doctor. At other times it is important to know how to treat yourself or your child properly for bleeds and injuries until you can get expert help.

Talk to your Haemophilia Centre team for advice on what would work best for you or your child.

Standard first aid techniques

- Bruising easily is common with VWD. Small bruises may look lumpy and unattractive but are not serious and usually fade over a few weeks
- Larger bruises or minor bleeding into muscles and joints can often be controlled by applying a cold pack (eg, an ice pack wrapped in a towel or a cool relief gel) and elevating the leg or arm. Apply the cold pack for about 10-15 minutes, then remove and reapply every two hours or so
- Bleeding from minor cuts can be stopped by applying pressure and keeping the cut dry until healed
- To treat a nosebleed, sit upright and pinch your nostrils together below the bridge of the nose in the soft tissue for 10 -15 minutes. If the bleeding doesn't stop, repeat this procedure a second time. Try to remain calm. A cold cloth on the back of the neck or on the bridge of the nose may also help
- Drinking hot liquids and strenuous exercise can cause the nose bleed to restart. Avoid hot soup, tea or coffee and lifting and straining for 24 hours after a nose bleed
- Bleeds in the mouth or tongue can be treated at home with tranexamic acid or aminocaproic acid tablets, syrup or mouthwash. These are available on prescription from your doctor.



Our son plays all sorts of non-contact sports – the specialists at the Children’s Hospital said they wanted to see him with little bruises, because it meant he has a life and isn’t wrapped in cotton wool all the time. We liked that. Once our anxiety settled (with lots of information), life got back to normal again.

If your child has an injury and a bleed, show them that being calm helps. Our son panicked at the slightest bleed at first, thinking he might die, but is now more relaxed.

Encourage your kids to tell you about any knocks or injuries as you can find some nasty bruises a couple of days later when it is too late to treat.

When to seek medical attention

In general, seek medical attention if:

- The nostril pinching procedure does not stop a nose bleed and severe bleeding continues for more than 20 to 30 minutes
- There is blood in the urine or stools
- Bleeding is excessive or lasts for hours
- There are broken bones
- There are cuts that do not stop bleeding with pressure and may require stitches
- There is a suspected bleed into a muscle or joint
- There has been a blow to the head or a head injury.

If you are at all uncertain about when or how to treat bleeding, contact your Haemophilia Centre team. Learning to self-manage treatment for your bleeding disorder happens over time.

Tips - useful first aid items to have on hand

Suggestions from people with VWD

- *Ice packs — keep a couple of the bendable cold gel-packs or packets of frozen peas or corn in the freezer. It's also handy to have instant ice packs in the glove box of the car or in your bag*
- *Compression bandages for sprains or bandaging over wounds*
- *Band-aids/plasters*
- *Tranexamic acid tablets – can be really useful when you have minor bleeding issues or can't get to treatment immediately*
- *Crutches – surprisingly inexpensive to buy from most pharmacies, and incredibly useful to have on hand, especially if you get muscle or joint bleeds; the physiotherapist at the Haemophilia Centre can also supply them when needed*
- *Gauze and surgical tape - for wound dressings*
- *Contact details for your hospital and haemophilia centre. Keeping them on the fridge at home can be helpful.*

Tips for dealing with medical emergencies

- Tell the medical and nursing staff that you or your child has VWD and what it means
- Have the contact details of your Haemophilia Centre on hand. You may wish to contact the Centre before you seek emergency treatment or ask the doctors or nurses where you are getting care to contact the Centre for advice
- If possible, organise a protocol through your Haemophilia Centre for handling emergencies with your local emergency department
- Consider wearing a medical alert bracelet and carrying a HTC Centre ID card in your wallet and/or having an ICE (In Case of Emergency) number in your mobile phone – in an emergency, even if you are conscious, you may be shocked or not well enough to communicate clearly
- Consider having your or your child's medical information on file at local hospitals providing emergency care.

We tell any medical caregiver ASAP that our child has VWD, and ensure the staff know what this means! When our son has a cut that needs stitching, we ensure they ring the Haematology Registrar at the Children's Hospital before treating him - then he has superglue instead of stitches, etc.

Now that protocols are in place, rather than just walking into the Emergency Department, we ring the Haematology Department first and they call down to the Emergency Department to expect us.

PREPARATION BEFORE MEDICAL, SURGICAL AND DENTAL PROCEDURES

It's important to inform your doctor, dentist or surgeon that you have VWD before undergoing any procedures. Without proper management, wound healing may be delayed as bleeding that occurs with surgery or procedures may last much longer in people with VWD. Before having surgery or medical or dental procedures, consult with your Haemophilia Centre so that you can discuss the medical support you may need to prevent excessive or unnecessary bleeding from the procedure.

Where possible, plan this well ahead of time. The Haemophilia Centre team may also need to liaise with the surgeon, dentist or other health professionals involved in your care to discuss the best approach for you individually and any pre- or post-treatment care you may need.



MEDICATION TO BE AVOIDED

Some medicines, vitamins and herbs interfere with the way platelets promote clotting and may delay healing. If you have VWD, consult with your haematologist before taking:

- Medicines containing aspirin
- Non-steroidal anti-inflammatory drugs, unless prescribed by a doctor with expertise in VWD (ie, ibuprofen, indomethacin and naproxen – these have many brand names; ask your local pharmacist to check for you)
- Other blood thinners such as warfarin and heparin
- Capsules of fish oil containing omega-3 fatty acids (however, normal serves of fish should not cause a problem)
- Herbal or homeopathic medicines that affect platelet function or clotting, such as ginkgo biloba, ginger, ginseng and chondroitin
- Other medicines that claim to treat bleeding, bruising or improve clotting.

Also check with your doctor when starting new medications that could irritate your mucous membranes such as your nasal passages or stomach lining – any bleeding could be complicated by VWD.

LIVING WELL WITH VWD

Von Willebrand disorder may be part of life, but it doesn't need to rule it. If you have VWD, you can still go to school, have adventures, play sports, raise a family, work, travel, and do the things that inspire you or enjoy the fun activities you love.

BEING ACTIVE

Exercising regularly and being physically active is a great way to keep muscles and joints strong and stay in good health. Exercise can help to boost VWF levels and may even reduce the number of bleeding problems and episodes you experience.

Whether at work or play, being active can be a balance between taking on physical challenges and preventing injuries or dealing with accidents quickly. Know your limits and be prepared.

We live on a farm and bruises tend to be part of every day life when dealing with animals. I have an ice pack in the fridge which is often used weekly.



What about sports?

People with VWD generally have to find out for themselves what physical activities they can and can't do. Many people with mild VWD take part in all kinds of sports, including active sports like soccer and moderate-risk sports like downhill skiing. People with type 3 or severe VWD might find that contact sports or sports with a high risk of injury, such as football or boxing, lead to serious bleeding and may prefer less injury-prone sports or to modify their activities.

It's all about understanding your limits. At a recent event I decided that tandem skydiving was "safe" but because of the tightness of the seatbelts, rally driving wasn't.

It is important to give children the chance to discover what activities they can safely do. As they grow up, they will want to take part in the same sports as their friends. Naturally parents may want to protect their children from harm, but they can do this by making sure their children follow the current safety guidelines for all children in sport and are prepared for accidents.

Choose sports wisely but don't wrap them in cotton wool. Our child with the most serious VWD is our "tree-climber"!

Most kids with VWD are normal kids who should be able to do normal activities. My kids seem to know their limitations but at the same time VWD has never stopped them. The kids ride motor bikes and horses, play football, netball, cricket, tennis and basketball.

Sometimes kids do have to stop and think about themselves - a sprained ankle from netball can take a little longer to recover from due to severe bruising. Team mates might be pushing to get them back on the court but that ankle has to carry them around for lots of years.



Tips

- Ask the Haemophilia Centre team for advice on risks with sport and work, based on your or your child's individual health and situation
- Follow the latest safety guidelines and use the protective equipment recommended for your sport
- Be ready for accidents, just in case: always have your first aid kit on hand (this might include emergency treatment product); wear a medical alert bracelet; and tell someone with you what to do if you are injured
- Treat injuries, bruising or bleeding promptly and give yourself time to recover fully.

You just have to weigh up the risks and make sure that you completely understand the consequences and are prepared for what could possibly happen. I have type 1 VWD and ride a motorbike. I wear a medical alert bracelet, carry DDAVP with me and advise my riding partners of my situation.

Common sense is very important - the kids need to understand what VWD is all about and realise they have to set their own limits as mum and dad are not always watching over them.

Further reading

Egan B. Boys will be boys: a guide to sports participation for people with haemophilia and other bleeding disorders.

Melbourne: Royal Children's Hospital, 2005.

(available from HFA and your Haemophilia Centre)

TRAVEL TIPS

Pre-planning can make all the difference – travelling is a wonderful opportunity for activity and adventure, and it can be much more enjoyable if you are prepared for all eventualities:

- Let your Haemophilia Centre know if you are travelling interstate or overseas
- You may not need them, but just in case, find out the names, addresses and phone numbers of Haemophilia Centres along your route – hours of operation and where to go in the hospital may also be helpful, eg directly to the Haemophilia Centre, or to the Emergency Department
- The Haemophilia Centre can also advise on medication and blood products, documentation and travel insurance
- Overseas travel may require further documentation for customs and security – talk to your Haemophilia Centre about this well in advance and allow plenty of time to prepare the documentation!

Useful travel web sites:

- Smarttraveller – www.smarttraveller.gov.au
- World Federation of Hemophilia Passport (global treatment directory) – www.wfh.org
- Medicare Australia (Travelling Overseas section) – www.medicareaustralia.gov.au



SCHOOL

Having VWD shouldn't affect a child's ability to attend school. Most bleeding with Types 1 or 2 VWD won't be serious enough to keep a child out of school. Occasionally, with type 3 or severe VWD, a serious bleed into a muscle or joint might mean that a child is absent from school for a short time – but with prompt treatment, this should only be for a day or two.

- When they are old enough, a child needs to know how to handle their own common bleeding problems, such as nosebleeds. If the child is very young, a staff member will need to learn how to manage them
- Provide the day care centre or school with information on VWD and on the child's condition and how to manage it
- It is important that the day care or school staff can contact parents or guardians at all times, in case of emergency. It may also be helpful to provide the telephone number of their Haemophilia Centre.

Well-informed day care and school staff can be very helpful. It is important that they have the facts about VWD, but that the information isn't over-dramatised. You may find it useful to organise information sessions for staff at day care or school – Haemophilia Centre staff are usually very experienced in this and happy to assist you. A child may or may not want to educate their classmates – most young people want to be treated normally, so the value of this would depend on each individual situation.

Helping school teachers be more relaxed (yet always wary, without the kids knowing) helps heaps.

Most of our worry about VWD has been other people's anxiety. People panic if my son has the slightest scratch – it is important that they are fully informed and that they show my son they are not frightened of him.

DO I NEED TO TELL OTHERS?

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

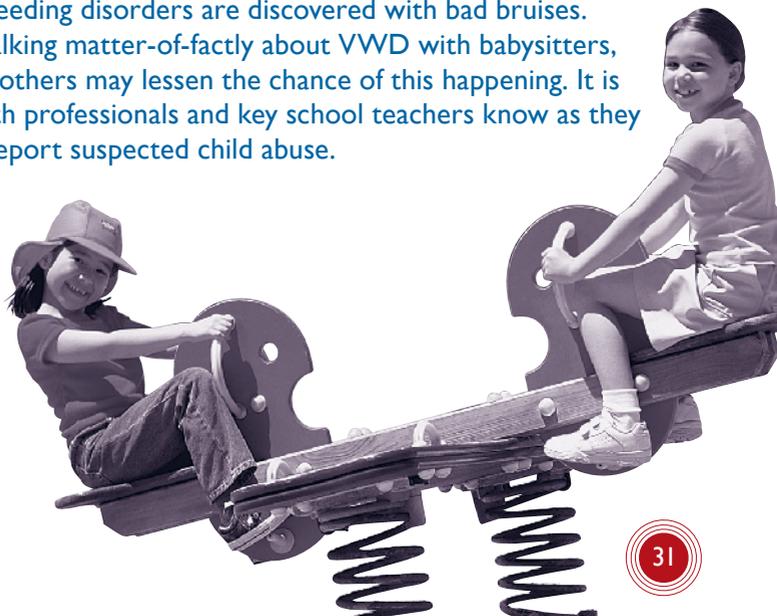
- The age of the person with VWD and whether they can look after themselves
- How severe the symptoms are
- Their relationship to the person with VWD.

Generally, those taking responsibility for the person with VWD (babysitters, sports coaches, carers, etc) need to know what to do in case of bleeding. If bleeding symptoms are extremely rare or the person can take care of any problems themselves, informing may not be necessary.

Consider whether you might want to tell your work manager, close friends or travelling companions in case of an emergency.

If the question is asked on an insurance or superannuation form, you are obliged to answer it. This could mean you are refused insurance or have to pay a higher premium. As different companies have different policies, shop around for your best option – talk to your Haemophilia Social Worker or Counsellor or Haemophilia Foundation for more information.

Parents have occasionally been suspected of child abuse when their children with bleeding disorders are 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. It is wise to let health professionals and key school teachers know as they are obliged to report suspected child abuse.



WORK

Most people with VWD will be able to take up their chosen career. An exception to this is the armed forces, which has very strict medical entry requirements.

If you have type 3 or severe VWD, you may find that jobs that are extremely demanding physically may be more likely to lead to severe bruising or bleeds. Talk to your Haemophilia Centre team about whether you can manage or prevent this with treatment and/or options for modifying activities.

SPECIAL ISSUES FOR WOMEN AND GIRLS

Heavy bleeding with menstrual periods (menorrhagia) is a common symptom of VWD for women and girls. It may involve:

- Heavy menstrual periods (eg, soaking through a tampon and pad around two hourly, or needing to change during the night)
- Menstrual bleeding for longer than normal (eg, longer than 8 days)
- Bleeding with clots bigger than a 50 cent piece in size.

Heavy menstrual bleeding can lead to anaemia (low red blood cell count/low blood iron levels), with symptoms of fatigue, paleness, lack of energy and shortness of breath.

Some women and girls with VWD also experience:

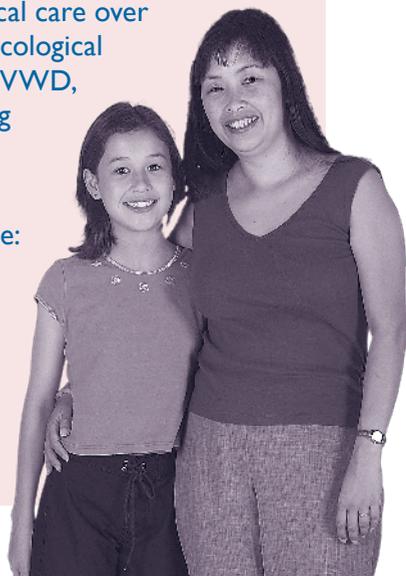
- Pain during their menstrual periods (dysmenorrhoea)
- Abdominal pain and sometimes bleeding during ovulation (when an egg is released from the ovaries, around the middle of the menstrual cycle).

Although these can be symptoms of VWD, they can also be symptoms of a gynaecological disorder, so it is important to consult a gynaecologist.

If you are a woman or girl with VWD, a holistic or comprehensive care approach to your health care can help you to achieve better health and quality of life. Specialist gynaecological care over your lifetime is important to manage any gynaecological issues that occur. These may not be related to VWD, but in some cases VWD may make the bleeding problems worse.

Ideally your medical care team should work together on your health care and should include:

- A gynaecologist
- A haematologist specialising in bleeding disorders
- A GP or paediatrician or obstetrician, if relevant at the time.



HOW IS THIS TREATED?

With diagnosis and appropriate treatment, these problems can be dramatically reduced and sometimes even eliminated. Treatments include:

- **Tranexamic acid** and **aminocaproic acid**, antifibrinolytic drugs which can reduce bleeding by slowing the breakdown of blood clots
- **Oral contraceptives** (“the Pill”) **combining the hormones oestrogen and progesterone**. The hormones increase VWF and factor VIII in the blood and reduce menstrual blood loss. Although it also has the effect of preventing pregnancy, in this case the treatment’s aim is to manage VWD symptoms and so it may also be suitable for teenage girls who are not sexually active and women who are not specifically seeking birth control
- **An intrauterine device (IUD), releasing the hormone progesterone** which reduces bleeding
- **Desmopressin (DDAVP)**, a synthetic hormone which stimulates the body to release VWF and factor VIII
- **Iron supplements** for anaemia.

See also the “*What is the treatment for von Willebrand disorder?*” section

Generally, treatment options with medication will be exhausted before considering surgery such as hysterectomy (surgical removal of the uterus) or procedures such as endometrial ablation, where the lining of the uterus is destroyed to reduce menstrual blood loss. Surgery and some procedures have their own risk of bleeding complications for women with VWD.

However, some women with VWD may need to have gynaecological surgery or procedures for other reasons. If this happens, it is important that this is managed in a team, with discussion between the woman, the Haemophilia Centre and the gynaecologist and/or surgeon.

HOW MIGHT THINGS CHANGE OVER A LIFETIME?

Puberty: menstrual bleeding can be especially heavy when a girl first starts having periods. When there is a family history of VWD or it is known that she has VWD, a girl should be followed closely by her medical team during puberty and may need treatment if she has heavy bleeding.

Sexual intercourse – some women with VWD may experience bleeding if there are small tears in their vagina after sexual intercourse. This can happen during their first sexual experience when the hymen is broken. It can also occur after childbirth and menopause when the vaginal wall may be thinner and dryer due to a drop in oestrogen levels - oestrogen creams for the vaginal wall and/or lubricants can help with this.

Pregnancy and childbirth: most women with VWD do not have a problem with delivering a healthy baby. Pregnancy can cause blood levels of VWF to increase, decreasing the likelihood of bleeding complications during pregnancy and delivery. However, this needs to be monitored as women with VWD can have heavy bleeding for an extended period after delivery when their factor levels return to their usual levels.

To minimise the chances of complications:

- Discuss VWD with a genetic counsellor, your haematologist and an obstetrician before you become pregnant
- Before you have any invasive procedure, such as amniocentesis, ask your haematologist if you are at risk of bleeding and whether anything needs to be done to prevent it
- During your third trimester, you should have blood tests to measure VWF to help plan for delivery and for any treatments to prevent potential post-delivery bleeding
- Discuss your choices for anaesthesia, especially an epidural, with your haematologist, obstetrician, and if possible, your anaesthetist
- Unless prenatal testing has shown the opposite, it should be assumed that the baby may have VWD and delivery methods should be as gentle as possible. A caesarean section is not usually required.

Menopause: When a woman begins menopause, her body's erratic hormone regulation can increase her risk of unpredictable and heavy menstrual bleeding. However, for some women with VWD, levels of VWF rise as they age and normalise so that bleeding problems reduce. Keeping a close relationship with her gynaecologist in the years before menopause will help a woman with VWD be prepared to manage any problems that might occur.

There are specific resources for women with bleeding disorders – contact HFA for more information or check the HFA web site – www.haemophilia.org.au

CONNECTING WITH OTHERS

People with VWD or parents of children with VWD often comment on how helpful it is to talk to others in a similar situation and know they are not alone. To pick up tips on how to manage VWD and contact others affected by VWD, stay in touch with your Haemophilia Foundation.

State and territory Haemophilia Foundations have:

- Newsletters and web sites to update people with bleeding disorders and their partners, families, friends and carers
- Social activities where people can meet, talk about common experiences and enjoy a meal or a day out, eg family camps, Christmas parties, men and women's groups, grandparents' groups.

Haemophilia Foundation Australia also supports:

- A youth program run by young people affected by bleeding disorders
- Internet-based communities and social networking sites for people affected by bleeding disorders.

The best thing to do is to chat with other people in a similar situation. That would have been great when we were first diagnosed as I could have learned from someone else's mistakes instead of my own!

Talking to other parents of kids with VWD and using their experiences to help your own situation makes a huge difference.

CAN I LIVE A NORMAL LIFE WITH VON WILLEBRAND DISORDER?

Absolutely! Most people with VWD only have occasional and mild bleeding problems.

Others may bleed more often or sometimes have a more serious bleeding problem, but with appropriate management, these bleeding episodes can be controlled or prevented.

People with von Willebrand disorder have a normal life expectancy.

With awareness and planning, most people with VWD live full, active and independent lives.



MORE INFORMATION

For more information about von Willebrand disorder (VWD), or to find out how to get in touch with your local Haemophilia Foundation or a specialist Haemophilia Centre, contact:

Haemophilia Foundation Australia

P: 1800 807 173 **E:** hfaust@haemophilia.org.au

W: www.haemophilia.org.au

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IMPORTANT NOTE

This booklet was developed and fully funded by Haemophilia Foundation Australia for education and information purposes only and does not replace advice from a treating health professional. Always see your health care provider for assessment and advice about your individual health before taking action or relying on published information.

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