Liverpool University Hospitals
NHS Foundation Trust

Patient information

Type 1 Von Willebrand Disease

Haematology Liverpool

Introduction

This leaflet aims to provide you with general information about the signs and symptoms you may have with Type one Von Willebrand Disease. If you are ever worried about your symptoms please contact the Haemophilia team or present to the Emergency Department.

What is von Willebrand Disease?

Von Willebrand Disease (VWD) is an inherited disorder of the clotting (coagulation) system. A protein in the blood called von Willebrand factor (VWF) helps blood to clot. In VWD either the level of VWF is low or the VWF does not work very well, or both. VWF is one of many clotting factors that work in combination to form a stable clot at the site of injury. VWF also helps to stabilise clotting factor VIII (eight) in the blood. If your blood does not clot properly, you can bleed more than most people and have symptoms such as easy bruising, frequent or long-lasting nosebleeds or bleeding from your gums. Women with VWD often experience heavy menstrual periods (menorrhagia)
There are three main types of VWD:

- **Type 1 is found in between 60-80% of people with VWD. This is a reduction in the amount of VWF, but not in the function of the VWF that is present.**
- **Type 2** is found in between 15 -30% of people with VWD. This type is characterised by reduced function of the VWF. The levels on blood tests may be almost within normal limits but the clotting protein does not work properly.
- **Type 2** VWD is split into further classifications according to the natures of the functional change: type 2A, type 2B, type 2M and type 2N. Bleeding symptoms can be mild to moderate.
- **Type 3** is associated with a significant reduction or complete absence of VWF and affects around 1 in 500,000 people. As a result of this significant reduction or absence of VWF, your platelets will not be able to form a clot and your factor VIII level will be low. This will put you at risk of severe bleeding that is difficult to stop. Bleeding from the mouth, nose and gut is common, and joint and muscle bleeds can occur after an injury; because of this increased bleeding frequency Type 3 is usually diagnosed at a very young age. Prophylactic factor replacement will be required to prevent bleeding.

What are the signs and symptoms of VWD?

- frequent or large bruises from minor bumps or injuries.
- frequent nosebleeds, or nosebleeds that are hard to stop.
- heavy, delayed or prolonged bleeding after surgery.
- prolonged bleeding from gums after dental procedures.
- heavy bleeding from a cut.
- prolonged or heavy bleeding after an injury.
- bleeding gums.
- heavy or prolonged menstrual periods (menorrhagia).
- bleeding during or after labour and childbirth.
- spontaneous bleeding, e.g. internal or joint bleeding (only in the most severe of cases).

How is the diagnosis made?

Because symptoms can be mild, VWD can be difficult to diagnose and may go undetected. Your diagnosis will usually be made in a Haemophilia Comprehensive Care Centre or a Haemophilia Treatment Centre, where you will see a Haematologist (doctor specialising in blood disorders) or a Haemophilia specialist nurse.

You will be asked if you bruise or bleed easily, and about problems with any surgery or dental procedures. If these bruising or bleeding episodes happened when you were very young you may not be able to answer some of these questions, and it would be important to speak to close relatives where possible. You will also be asked if other family members are affected by any bleeding or bruising or are known to have inherited bleeding disorders. As well as asking you questions, we will need to collect a sample of your blood, which we will send to a laboratory for testing. Due to the specialised nature of these tests, it may take up to two or three weeks to get your results. Many people need multiple tests over a period of time to make a clear diagnosis, including which type of VWD they have.

Blood tests may include

- von Willebrand factor antigen test: this measures the amount of VWF in the blood.
- von Willebrand factor activity tests: these show how well your VWF works.
- factor VIII level: this is checked because if your VWF is low, your factor VIII level may also be low.
- blood group.

Sometimes these bloods will have to be repeated as the levels can fluctuate. Further blood tests including genetic bloods may be taken to confirm the type of VWD and to help us to provide you with accurate information regarding treatment that may be required. Genetic tests can only be undertaken with consent.

How did I get von Willebrand Disease?

VWD is most commonly inherited from your parents, passed from one parent who is affected by VWD to their child. Less commonly, a spontaneous genetic error can occur and you may be the first person in the family to have VWD.

The type of VWD a person is born with mostly depends on whether they inherit copies of this faulty gene from one or both parents.

If one parent has a genetic fault that causes VWD, there is a one in two (50%) chance of a child having type 1 VWD.

Once there is laboratory confirmation of your bleeding disorder, we will be able to advise you if you or other family members need to attend for genetic investigations. We can arrange for the necessary genetic tests to be carried out at the Liverpool Haemophilia Comprehensive Care Centre.

What does VWD mean for day-to-day life?

There is no need to amend your usual activities, although would advise against contact sports such as rugby, boxing, judo and karate. Check with your Haemophilia specialist nurses if you have any questions regarding this.

People with Type 1 VWD have few day-to-day problems relating to VWD but may require some treatment for injuries, surgery, and dental extractions. The most common symptoms in VWD are bleeding from the nose and mouth. Women are more likely to experience symptoms and complications due to the increased risk of bleeding during menstruation, pregnancy, and childbirth. It may also be hard to stop bleeding after an injury, dental procedure, or surgery.

How often will I see the haemophilia team?

You will be reviewed regularly by the Haemophilia team. These appointments are a good time for us to get to know you and for you to get to know the team, and for you to ask any questions and talk a little about VWD. You will also be registered with our centre on the National Haemophilia Database who will issue a Bleeding Disorder Alert Card (see NHD patient information leaflet).

You should contact the haemophilia team in between appointments if you:

- have had a head injury.
- have a prolonged nosebleed which does not seem to be slowing after 10-15 minutes.
- have dental issues with bleeding from the gum that is hard to manage.
- have difficulty in managing heavy periods.
- become pregnant.
- need an operation.

Please let us know if you need surgery or dental extractions or treatment. It is important for us to know the planned date as soon as possible so that we can make a treatment plan to make sure the procedure runs smoothly.

Can I join in with sports?

Yes. Most sports are encouraged as they can help to keep you fit and healthy long term. Contact sports such as rugby, mixed martial arts and boxing should be avoided, tag rugby can be participated in. Other sports including football, swimming, dancing, basketball, and cycling are encouraged.

Treatments for von Willebrand Disease

Nose / gum bleeding: Use usual first aid for nose and mouth bleeds. Apply pressure to the affected area, for example, where a tooth has fallen out a finger wrapped in gauze pressing firmly over the socket, or for nose bleeds apply firm pressure below the bridge of the nose. Sucking on an ice pop helps both nose and mouth bleeds to slow down as it cools the area. Seek advice if bleeding is ongoing after 10-15 minutes of this initial management, or you start to feel unwell.

Injuries and heavy bruising: use normal first aid measures first; this includes Rest Ice Compression Elevation otherwise known as **RICE** – if you aren't happy that you are able to adequately self-manage an injury please do contact the Haemophilia team for advice or attend the Emergency Department or Urgent Treatment Centre.

- **Rest-** Rest the affected joint or muscle.
- **Ice-** Apply ice, wrapped in a towel or a cold pack to the affected joint for 10-15 minutes. Ice helps to reduce pain and muscle spasms as well as reducing swelling and redness. Ice alternating with heat can also help to reduce pain, particularly if the bleed is into a muscle.
- **Compression-** Applying pressure to the injured area will help slow blood flow; the pressure can also provide comfort.
- **Elevation-** Raise the injured arm or leg above the heart although this may seem difficult you can prop an arm and leg up with cushions or pillows. This helps to reduce the blood flow to the area, lowering the blood pressure to slow the bleeding.

Medications Currently there is no cure for von Willebrand Disease but there are medicines and treatments available that help to keep symptoms under control.

Tranexamic acid is an anti-fibrinolytic agent and helps stabilise the blood clots that form whilst the body heals. It is often used to prevent or treat bleeding from mucous membranes such as the inside of the mouth, nose, gut or womb. It is usually taken as tablets, three or four times a day, but may also be given as an intravenous infusion (drip into a vein).

Desmopressin (DDAVP) stimulates release of your own clotting factors (factor VIII and VWF) from storage sites in the body into the blood. Levels of the clotting factors are increased by three to six times your baseline level for 12-24 hours. If necessary, you may have a repeat dose after 12 hours. In some people repeated infusions may not be as effective because the body doesn't have the chance to rebuild its stores. The dose is calculated according to body weight.

It cannot be used if you have certain medical conditions and does not work for everyone. We may perform a DDAVP trial prior to any emergencies or surgical interventions to see if this medicine will work for you.

Von Willebrand Factor concentrate (Veyvondi) is given as an injection into the vein to replace the missing VWF and allow clotting to take place. It is a man-made product (recombinant), which means it is not made from blood donations. It is given before surgery or dental work and for bleeding that is difficult to control. You may require repeat doses (top-ups) depending on the type of surgery or injury.

Who do I tell about my diagnosis?

It is helpful for other healthcare professionals to be informed, including your GP and any other specialists involved in your medical care.

You should talk to family members who may need to ask a doctor to check if they also could be affected by von Willebrand Disease.

You might find it helpful to tell friends about VWD so that they can support you and so that if you develop bruises they can understand why. Your workplace should also be informed so that appropriate risk assessments can be carried out.

Medicines that should be avoided

Please avoid using pain killers known as NSAIDs (e.g. ibuprofen, naproxen, diclofenac) or Aspirin as these medicines interfere with the way the blood clots by affecting the function of platelets. These can make the symptoms of VWD worse.

You can take paracetamol for pain relief or to reduce a temperature, or please seek advice from a pharmacist regarding other pain relief that could be suitable.

Links

The Haemophilia Society website https://haemophilia.org.uk/bleeding-disorders/von-willebrands/ Liverpool University Hospitals NHS Foundation Trust http://www.liverpoolft.nhs.uk/

Feedback

Your feedback is important to us and helps us influence care in the future.

Following your discharge from hospital or attendance at your outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point.
Your co-operation is greatly appreciated.

Further information

If you have any further questions, please ask a member of our medical / nursing team. Please contact the Haemophilia Team, available Monday - Friday 9am - 5pm
Tel: 0151 706 3397

Useful websites

The Haemophilia Society:
https://haemophilia.org.uk/

World Federation of Haemophilia:
www.wfh.org

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All Trust approved information is available on request in alternative formats, including other languages, easy read, large print, audio, Braille, moon and electronically.

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