East Kent Hospitals University NHS Foundation Trust

von Willebrand's Disease (vWD)

Information for parents, patients, and carers from the Haemophilia Centre

What is von Willebrand's Disease (vWD)?

Von Willebrand's Disease (vWD) is the most common bleeding condition that affects blood clotting. It is thought to affect around one in every 100 of the population in the UK.

People with vWD do not bleed any faster, but may bleed for longer than expected.

Having vWD should not affect your everyday life. Most of the bleeding problems that happen tend to be after an injury, surgery, or dental treatment.

How does von Willebrand's Disease affect clotting?

Von Willebrand factor (vWF) is a protein that helps to make your blood clot when there is blood vessel injury. If you have von Willebrand's Disease you do not have enough of this protein for your blood to clot normally, or what you do have of the protein might not work as well as it should.

vWF also helps the clotting system by carrying another of the clotting factors (Factor VIII), so some people with vWD may also have low levels of Factor VIII.

How do you get von Willebrand's Disease?

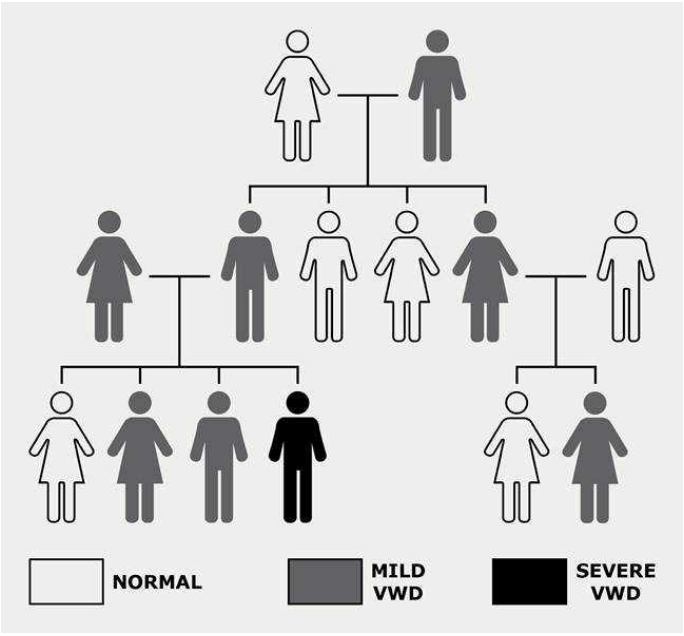
Von Willebrand's Disease is an inherited condition, you are born with it. Both men and women can inherit it.

One set of our genes is inherited from our mother and the other set from our father. Both parents must have the defective gene in order for the child to be more severely affected.

Mutations can happen, which mean that either the gene is unable to make enough vWF or that what it does make does not work as well as it should.

The diagram shows how von Willebrand's Disease may be passed down through a family.

Family members can be affected differently depending on whether they have inherited the defective vWD gene from one or both parents. This is why testing them is important to establish the severity of the condition.



Von Willebrand Disease. © World Federation of Hemophilia 2012. http://elearning.wfh.org/elearning-centres/vwd/#how_do_you_get_vwd

What are the different types of von Willebrand's Disease?

There are three main types of von Willebrand's Disease, depending on whether the vWF is missing or it is not working properly - Type 1, Type 2, and Type 3.

- **Type 1** is the most common (affecting around eight in 10 people with vWD). People with Type 1vWD have lower levels of the von Willebrand factor. The symptoms are usually mild (see symptoms below).
- **Type 2** is second most common (affecting up to two in every 10 people with vWD). In people with Type 2 vWD the von Willebrand factor does not work properly. There are four different subtypes of Type 2 vWD (2A, 2B, 2M, and 2N). There are generally more bleeding problems with Type 2 vWD than with Type 1. The type of treatment you need may be different depending on the subtype you have.
- **Type 3** is much rarer (affecting one in every 100 people with vWD). People with Type 3 vWDhave little or no vWF and will have more serious bleeding problems, including spontaneousbleeding (bleeding which happens for no apparent reason).

What are the symptoms of von Willebrand's Disease?

Common symptoms

- In women and girls, heavy and prolonged bleeding during menstruation (known as menorrhagia)
- Nose bleeds
- Bleeding from gums
- Easy bruising
- Bleeding after surgery
- Bleeding after trauma or injury

- Bleeding in the mother at childbirth.

Rare symptoms

- Blood in your urine (known as haematuria)
- Bleeding in your stomach or intestines (blood in your stools (poo) or black tarry stools).

How is von Willebrand's Disease treated?

Day-to-day treatment is not needed for mild or moderate von Willebrand's Disease. However it may be needed before and after surgery, dentistry, or injury.

The need for treatment will depend on:

- how severe your bleeding is
- the type of surgery or dental treatment you are having
- your previous history of bleeding
- your family's history of bleeding; and
- your level of vWF.

What are the treatments for von Willebrand's Disease?

There are three main treatments for vWD.

- Antifibrinolytic Agents (Tranexamic Acid)
- DDAVP (Octim)
- Veyvondi
- Voncento
- **Antifibrinolytic Agents (Tranexamic Acid)**

  Tranexamic Acid works by stopping the early breakdown of a clot that has been made after injury to your blood vessel. Fibrin is a protein that gives the blood clot stability. Tranexamic Acid stops the substances that destroy the fibrin within the clot.

  Often it is the only treatment needed for some people with vWD and for some procedures. It is particularly useful for mouth bleeding and therefore very good preventative treatment when you are having a dental procedure.

  This treatment is usually given as tablets. You may be asked to crush the tablets in a small amount of water and swish it around your mouth, before either swallowing it or spitting it out (you will be advised by the haemophilia team which is best for you).

  Tranexamic Acid is not used when there is blood in your urine, as small clots can occur which can then block your urinary tract and cause you pain.

Are there any possible side effects?

- Nausea (feeling sick) and vomiting
- Diarrhoea
- Joint or muscle pain
- Muscle cramps
- Headache or migraine
- Runny or stuffy nose
- Stomach or abdominal pain.

Other side effects may include skin rash and changes to your colour vision.

- **DDAVP (Octim)**

  DDAVP (Octim, also called Desmopressin) is a synthetic (artificial) medicine given as a small injection under the skin. It helps your blood to clot by releasing your own stores of von Willebrand factor. Please see the DDAVP patient information sheet for more information. Ask a member of staff for a copy or go to the haemophilia web page (https://www.ekhuft.nhs.uk/services/haemophilia-and-thrombosis-centre/).

  Are there any possible side effects?

  - Red or flushed face (this is only temporary)
  - Headaches
  - Dizziness
  - Mild stomach pain
  - Nausea (feeling sick)
  - Allergic reactions (rare)
  - Fits (very rare).

- **Veyvondi**

  DDAVP is not suitable for all patients (either because of other medical conditions or DDAVP does not increase your vWF level enough when it is given).

  If there is a bleeding problem, Veyvondi replaces the low levels of von Willebrand factor. It does not come from blood donors.

  It is given to you either through a small butterfly needle or a cannula (a small tube into a vein in your arm) directly into the vein, over two to five minutes.

  Are there any possible side effects?

  - Allergic reaction
  - Headaches are common

    - Nausea (feeling sick) and vomiting
    - Dizziness / vertigo
    - Muscle twitching
    - Unusual taste.

- **Voncento**

  DDAVP is not suitable for all patients (either because of other medical conditions or DDAVP does not increase your vWF level enough when it is given).

  Voncento is made from donated blood, so there is a very small risk that you will get a blood-borne virus. There are ways in which these risks are minimised by the drug company. The first is that donors are carefully screened to make sure they do not carry these viruses. Secondly, testing for signs of viruses at each donation and finally, the Voncento is heat treated to inactivate or remove any possible viruses.

  Voncento is given to you either through a small butterfly needle or a cannula (a small tube into a vein in your arm) directly into the vein, over two to five minutes.

  Are there any possible side effects?

    - Allergic reaction
    - Headaches
    - Raise in body temperature
    - Changes to the way things taste.

Where can I find more information about vWD?

There are several sources of useful information about von Willebrand's Disease, including the following.

- The Haemophilia Society (https://haemophilia.org.uk/)
- Living with vWD Patients Support Community (https://livingwithvwd.org/)
- NHS: von Willebrand Disease (https://www.nhs.uk/conditions/von-willebrand-disease)

This leaflet has been produced with and for patients.

If you would like this information in **another language, audio, Braille, Easy Read, or large print** please ask a member of staff. You can ask someone to contact us on your behalf.

Any complaints, comments, concerns, or compliments please speak to your doctor or nurse, or contact the Patient Advice and Liaison Service (PALS) on 01227 783145 (tel://+441227783145), or email ekh-tr.pals@nhs.net (ekh-tr.pals@nhs.net)

Patients should not bring large sums of money or valuables into hospital. Please note that East Kent Hospitals accepts no responsibility for the loss or damage to personal property, unless the property has been handed into Trust staff for safe-keeping.

Further patient leaflets are available via the East Kent Hospitals website (https://www.ekhuft.nhs.uk/information-for-patients/patient-information/).

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