NHS
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Von Willebrand Disease and Transition to the Adult Services

Information for patients & families

leeds children's hospital
caring about children

This leaflet is for patients with von Willebrand disease who are aged 16 or over and who are ready to be transitioned to the Adult Services.

What is von Willebrand disease?

Von Willebrand disease (vWd) is a condition that affects blood clotting. If you have von Willebrand disease you may bleed from cuts for longer than normal and bruise more easily.

The severity of von Willebrand disease varies, but it usually affects people only mildly. Therefore, most people with von Willebrand disease live completely normal, active lives. It is the most common inherited bleeding disorder.

What goes wrong in blood clotting

There are proteins and cells in blood that make it clot. When a blood vessel is cut, these proteins and cells become active, and quickly form a blood clot to seal the cut and stop the bleeding.

If you have von Willebrand disease, one of the clotting factors, called von Willebrand factor is missing or does not work properly.

Therefore, blood clotting is less efficient than usual.People with von Willebrand disease usually have some von Willebrand factor in their blood, just lower levels than normal.

In these people, the disease is only mild, and the **common symptoms are easy bruising, frequent and sometimes severe nose bleeds and, in women, heavy periods.**

People with severe forms of von Willebrand disease have little or no von Willebrand factor in their blood. This can cause symptoms like those of haemophilia, such as bleeding into the muscles and joints. This is, however, a rare condition.

Why do I have von Willebrand disease?

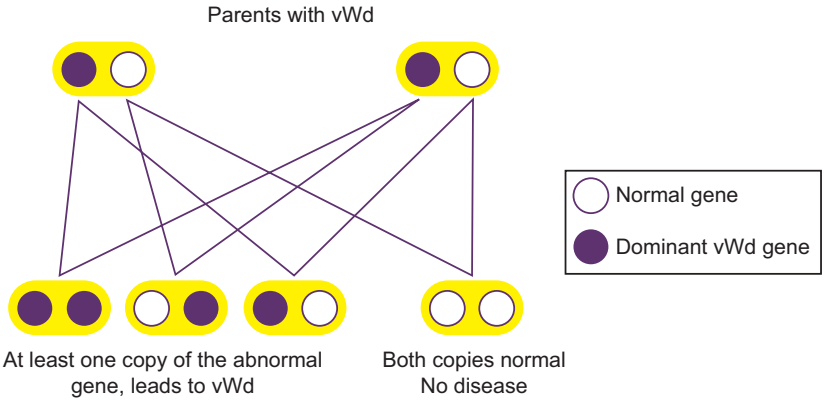
Von Willebrand disease is a genetic disorder and is passed on from your parents through an abnormal gene, rarely it can develop for the first time in the affected child so not be inherited. Von Willebrand disease affects both males and females and can be passed on by either your mother or father. Other genetic factors such as blood group can also influence the levels of von Willebrand clotting factor.

There are constant reviews as to how von Willebrand disease is inherited. The current thinking is as follows:

The gene for von Willebrand factor is very large and a variety of mistakes in it can produce von Willebrand disease. The abnormal gene that causes von Willebrand disease is usually "dominant" over its other copy.

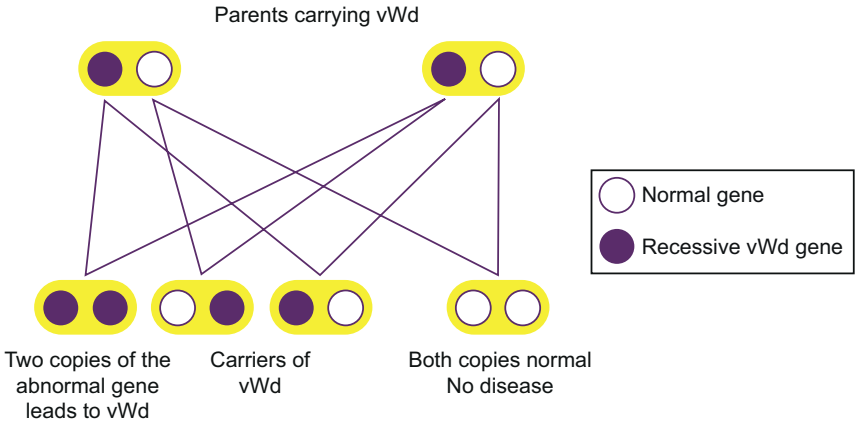
This dominant inheritance produces variable results - it cannot be predicted that all affected individuals will be affected to the same degree.

Inheritance of the dominant von Willebrand disease gene



Rarely, the gene for von Willebrand disease is hidden ("recessive"). In this case, it is possible for parents who do not have von Willebrand disease to carry it and pass it on to their children. For the recessive gene to produce von Willebrand disease, a person must have two copies of an abnormal gene, and this usually produces severe disease.

Inheritance of the recessive von Willebrand disease gene



Treatment of von Willebrand disease

Most people with von Willebrand disease do not need any treatment for most of the time.

When it is needed there are three main sorts of treatment available.

Tranexamic Acid

- Is a medicine that controls bleeding. It helps to stop blood clots from breaking down, which slows down bleeding. It is used for nose and gum bleeds and can also be used to reduce blood loss if you have heavy periods. It is often prescribed for dental procedures and is used as a mouth wash and then the solution is swallowed.

Desmopressin (DDAVP)

- You are likely to have had a desmopressin trial when you were younger to see if this treatment is effective as it does not work for everyone. The medicine is usually given as an injection under the skin but can also be given into the vein or as a nasal spray.
- It is often used to prevent bleeding during minor procedures such as dental treatments and also for nose bleeds.

Factor Concentrate

- Concentrated von Willebrand factor can be given to treat severe bleeding (e.g. after an accident) or if you have a severe form of von Willebrand disease. This is given by injection into a vein and replaces the missing von Willebrand factor, so blood clotting takes place at the normal speed.

Things to remember:

- It is important even if you do not have any symptoms of bleeding, if you have a planned intervention such as dental surgery you must inform your dentist that you have von Willebrand disease and contact the adult team who will be able to advise you. It is likely that you will need tranexamic acid and/or desmopressin prior to treatment to reduce the risk of bleeding. Any dental treatment or surgery should take place at the Dental Institute in Leeds.
- If you are involved in an accident or injure yourself you should also notify the Accident and Emergency staff that you have von Willebrand disease and show them your bleeding disorder card. You should carry your bleeding disorder card with you at all times. If you require a new card, this can be obtained from the adult team.
- When you are thinking of starting a family or you become pregnant it is important to let your Haematology Consultant know so that plans can be put in place for obstetric care. Extra monitoring may be required throughout pregnancy and interventions at delivery. Make sure your midwife is also aware that you or the baby's father has von Willebrand disease.

One last thing to remember!

It is important that you **DO NOT TAKE N**on **S**teroidal **A**nti **I**nflammatory **D**rugs **NSAID's** (pronounced en-said) as they can cause bleeding into the stomach.

NSAID's include:

- Ibuprofen, nurofen, diclofenac potassium, diclofenac sodium, aceclofenac, acemetacin, celecoxib, dexibuprofen, dexketoprofen, etodolac, etoricoxib, felbinac, fenoprofen, fluribiprofen, indometacin, ketoprofen, mefanamic acid.
- We don't expect you to remember the list! **Just remember NSAID's**. If you are prescribed them it must be under the supervision of your Haematology Consultant.
- When you are prescribed or buy drugs over the counter always inform your doctor or pharmacist about your **von Willebrand disease diagnosis and show them your bleeding disorder card**.

When you are asked - **'Do you have any allergies?'**

Your response should be - **'I have von Willebrand disease so cannot take NSAID's.'**

If you would like more information about von Willebrand disease a good resource is The Haemophilia Society website:

- **www.haemophilia.org.uk**

Rechecking Von Willebrand Levels

Von Willebrand levels can increase with age so before you are transitioned to the adult services we will perform a blood test to check your levels. If the new results fall within the normal ranges you may be discharged from our care rather than transitioned to the adult services.

Transition to Adult Services

While you have been cared for by the children's services, in general your parents or guardians would have taken charge of your care. Around 16 - 18 years of age, patients who have been under the care of the children's services must be transferred to the adult service. This is important as your needs will have changed from when you were younger. As a child your care was provided with a child/family centred approach but as you get older and are making independent life choices it is important that the responsibility for managing your von Willebrand disease shifts to you as an individual.

Now you are old enough to transition to adult services you are expected to be in charge of your care. However, you will always have the support of your nurses and doctors at the hospital.

In due course, you might be thinking about career choices, moving out of the family home and eventually starting a family of your own. The adult team will be able to help you with any queries you may have about your diagnosis and how this will affect your future.

It is likely that previously you will have had yearly telephone reviews if your symptoms were mild and well managed; it may have been that your parent or guardian undertook these consultations for you.

However, for your first appointment in adult services it will be a face to face appointment. This is important so that you can meet the staff involved in your care and familiarise yourself with Bexley Wing at St James's Hospital in case you need to attend for treatment or emergency review. After this initial appointment it is likely that you will be reviewed by telephone every 6 - 12 months.

You are still a patient of the children's team UNTIL you have had your first appointment with your new adult haematologist. Before this appointment, if you have a problem or query you can direct it to the children's team.

To contact the children's team call

- **0113 392 7179** - For appointments
- **0113 392 6863** - For advice - Haemophilia Nurse Specialists

The Adult Haemophilia Centre is on Level 3 at Bexley Wing, St James's University Hospital. There are several bus routes that take you there from Leeds city centre and there is a multi-storey car park on site.

To contact the adult team call

- **0113 206 8458** - For appointments
- **0113 206 8321** - For advice - Haemophilia Nurse Specialists
- Out of hours contact the inpatient wards 88-89 on: **0113 206 9188/ 0113 206 9189**
- For contact information in an emergency refer to your bleeding disorder card.
- Good luck from all the children's team members!

Notes

What did you think of your care?

Scan the QR code or visit **bit.ly/nhsleedsfft**

Your views matter

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