

Living life *with* von Willebrand

Information about von Willebrand disease.
How it can affect you. What treatments are available,
as well as suggestions and advice to enable
you to live your life in the best way possible.





This is von Willebrand disease

Both women and men can have von Willebrand disease. Women are more likely to notice the disease, as it can cause heavy menstrual periods and abnormal heavy bleedings after childbirth.

The disease is caused by a lack of or an impaired function of the von Willebrand factor. It is a protein found in the blood and is necessary for normal coagulation. When there is not enough, or if the von Willebrand factor does not work correctly, it takes longer for the blood to clot and for bleeding to stop.

There are various long chains of von Willebrand factor in the blood and these also act as carrier proteins for coagulation factor VIII. A lack of von Willebrand factor can also lead to a low level of Factor VIII.

From a mild to severe form

There are several different types of von Willebrand disease, ranging from mild to severe. Which type depends on how much the amount of – or function of – von Willebrand factor is affected.

Hereditary causes

Von Willebrand disease is hereditary and congenital, but new mutations can cause the disease in some individuals. It is caused by a change in the genes coding for von Willebrand factor which makes it function poorly or cause it to not be produced in normal amounts.

Each cell in the body contains 46 chromosomes arranged in 23 pairs. There are 20,000–25,000 different genes on the chromosomes, that determine how a person looks and functions. A gene serves as a template for a particular protein. Each pair contains one chromosome from the mother and one from the father. The genetic predisposition to von Willebrand disease is found on chromosome number 12.

Inherited in two ways

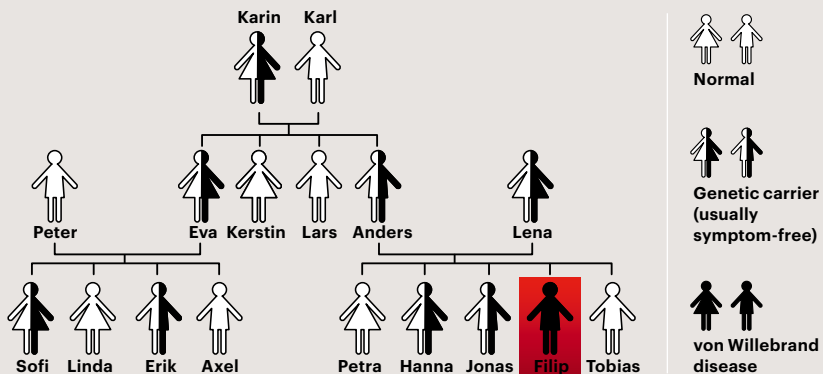
A child may inherit the gene responsible for von Willebrand disease from one or both parents. Since the disease is hereditary, it is often found in several members of the same family and relatives.

A child can be born with von Willebrand disease without either parent being genetically predisposed to the disease. In this case a change has occurred in the genetic make-up (new mutation), which causes the disease in the foetus.

Boys and girls are at equal risk of inheriting von Willebrand disease.

Recessive heredity

The genetic heredity of VWS type 3 and some type 2 variants is recessive. This means that you must have a double set of genes for the disease to manifest itself. That is, both parents have given you a gene. If you only have one gene, you are a carrier, and usually have no problems.



Symptoms of von Willebrand disease

Symptoms vary greatly from one person to the next, even within the same family. Here are some examples of common symptoms.

Bruising from minor blows and bleeding from the nose and oral mucous membrane.

Prolonged bleeding from skin wounds.

Important to get a diagnosis

Von Willebrand disease is under-diagnosed and many doctors are unaware of the diagnosis. It is therefore important that people are properly evaluated and referred to one of the country's three coagulation clinics (in Malmö, Gothenburg or Stockholm). To provide a basis for evaluation and assessment, it is important that your doctor is informed of all your symptoms and whether you have any known illnesses or are taking any medication. It is also helpful if your doctor knows how often you experience bleeding and if others in your family have bleeding problems.

Avoid ...

- ... painkillers containing acetylsalicylic acid or NSAID.**
- ... certain medications for panic disorder and depression.**
- ... blood-thinners**
- ... natural remedies such as omega-3 fatty acids.**



Prolonged bleeding after tooth eruption or extraction.

Abundant or prolonged menstrual periods (menorrhagia).

Abnormally heavy bleeding after surgery, childbirth or trauma of any kind.

Treatment depends on the severity of the symptoms

The treatment depends on the type of disease and the severity of the symptoms. Minor bleeding can often be managed without medication.

Examples of minor bleeding:

- Small bruises usually disappear on their own.
- Large bruises and minor muscle bleedings can often be stopped by applying a cooling agent and keeping the affected body part elevated.
- Bleeding from small wounds can be stopped with pressure.
- Nosebleeds can be stopped by pinching the soft parts of your nose with your fingers for ten minutes. It is a good idea to sit upright and lean forward slightly. Cotton wool that can help stop the bleeding can be bought in pharmacies.

However, in case of severe symptoms, medication or other medical treatment is necessary. The type of treatment depends, among other things, on the type of von Willebrand disease.

- Desmopressin is a medicine that stimulates clotting by releasing von Willebrand factor and Factor VIII from cells in the walls of blood vessels.
- Tranexamic acid does not stimulate coagulation in the same way as desmopressin, but it prevents the breakdown of a blood clot.
- Women can be treated with birth control pills, IUDs or other forms of hormone therapy such as vaginal rings or contraceptive rods.
- Treatment with factor concentrates containing von Willebrand factor may be needed for people with more severe forms of von Willebrand disease (especially type 2 and type 3). Patients can inject their own medication at home to prevent bleeding.

Children with von Willebrand disease

Symptoms of von Willebrand disease can occur in children of any age. It is often detected when the child falls and hits the mouth and bleeds for a long time from the gums. Many people do not notice an increased tendency to bleed. They discover that they bleed easily only after a physical injury or surgery. In some cases, von Willebrand disease is only discovered in connection with a relatives investigation.



One in five women

Bleeding disorders such as von Willebrand disease are much more common in women with heavy menstrual periods than in the general population. Scientific studies have shown that nearly one in five women who seek medical attention because of heavy periods, have some form of bleeding disorder. Many of them also have relatives with bleeding problems. 80–90% of women with von Willebrand disease have heavy menstrual periods compared to 10% of the general population.

Pregnancy and childbirth

For women with type 1, bleeding problems often decrease during pregnancy, as the level of von Willebrand factor in the blood rises sharply. The levels of von Willebrand factor and factor VIII should still be measured, especially as the time of delivery approaches, in case treatment needs to be initiated. After childbirth, the levels drops rapidly and there is a risk of heavy bleeding, which can sometimes continue for weeks. Treatment with tranexamic acid in combination with desmopressin, or concentrates of von Willebrand factor, is usually needed during labour and in the first few weeks afterwards.

Abundant periods and quality of life

Heavy menstrual periods can have a significant impact on a woman's quality of life. This may mean:

- That your work situation is affected and you have to reduce your working hours.
- That you get iron deficiency.
- That you are bothered by pains in connection with periods or ovulation.
- That you can get endometrial cysts or internal bleeding from the ovaries and often bleed through the menstrual pads.

Please contact your doctor, gynaecologist or healthcare centre if you experience these symptoms. You should not suffer unnecessarily and help is available. Problems can be reduced or eliminated with proper diagnosis and treatment.



If you are unsure about anything related to your von Willebrand disease – always ask your doctor!

Become a member. It makes a difference!

It is easy to register at fbis.se

Together we are stronger!

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