CONDITION

Von willebrand disease

A usually inherited bleeding condition in which von Willebrand factor is low or does not work properly.

WHAT IT IS
A long-term blood-clotting condition

BODY SYSTEM
Blood and clotting

MAIN SYMPTOMS
Easy bruising, nosebleeds, heavy periods, prolonged bleeding

WHAT CAUSES IT
Usually inherited changes affecting von Willebrand factor

HOW IT IS DIAGNOSED
Von Willebrand factor activity, antigen, and factor VIII tests

MAIN TREATMENT
Clotting medicines, antifibrinolytics, and procedure planning

CAN IT BE CURED?
No - inherited forms remain lifelong

IS IT AN EMERGENCY?
Sometimes - uncontrolled or internal bleeding needs emergency care

LONG-TERM OUTLOOK
Usually a full life with planned bleeding care

Overview

Von Willebrand disease, or VWD, is a bleeding condition. It is sometimes called von Willebrand's disease. Most forms are inherited, meaning they are linked to a gene change passed through a family.

People with VWD have too little von Willebrand factor, or the factor does not work properly. Von Willebrand factor is a protein that helps blood platelets stick to an injured blood vessel. It also protects another clotting protein called factor VIII.

When this process does not work normally, a clot may take longer to form. Bleeding is often from the nose, mouth, skin, uterus, or a cut. The severity ranges from mild bleeding that is noticed only after surgery to more frequent or serious bleeding.

There are three main types:

Knowing that you have VWD matters before surgery, dental work, pregnancy, or starting medicines that can affect bleeding.

- Type 1: a partial shortage of von Willebrand factor
- Type 2: von Willebrand factor is present but does not work properly
- Type 3: very little or no von Willebrand factor, usually causing more severe bleeding

Symptoms

Symptoms can begin in childhood, but mild VWD may not be recognised until later. Some people first notice it after a tooth removal, operation, injury, or childbirth.

Common symptoms include:

Heavy periods may mean changing period products very often, bleeding for longer than usual, passing large clots, or bleeding through clothes or bedding. Blood loss over time can cause iron-deficiency anaemia. This may lead to tiredness, weakness, headaches, pale skin, or shortness of breath.

People with severe VWD can have bleeding into muscles or joints. This is uncommon in milder forms.

Symptoms may change with age, hormones, illness, stress, and pregnancy. A person can still have VWD even if they have long periods without obvious bleeding.

- Frequent or long-lasting nosebleeds
- Easy bruising, sometimes with large bruises
- Bleeding from the gums
- Bleeding from cuts that lasts longer than expected
- Heavy or prolonged menstrual periods
- Excessive bleeding after dental work, surgery, miscarriage, or childbirth

Causes and risk factors

Most VWD is caused by an inherited change affecting the production or function of von Willebrand factor. It can affect people of any sex.

Many cases of type 1 and type 2 follow a dominant inheritance pattern. This means one altered

copy of the relevant gene can cause the condition. Type 3 usually follows a recessive pattern, in which both copies are altered. Some type 2 subtypes have different inheritance patterns.

A family history supports the diagnosis, but its absence does not rule VWD out. Relatives may have mild or unrecognised symptoms, and new genetic changes can occur.

Rarely, VWD develops later in life without an inherited gene change. This is called acquired von Willebrand syndrome. It may be associated with certain heart, immune, blood, or thyroid conditions, or with some medicines.

Aspirin, non-steroidal anti-inflammatory drugs such as ibuprofen, and anticoagulants like warfarin or apixaban can increase bleeding. Do not stop a prescribed medicine without medical advice.

Diagnosis

Diagnosis is based on your bleeding history, family history, examination, and blood tests. A clinician may use a standard bleeding questionnaire to assess the pattern and severity of past bleeding.

Tests used to confirm and classify VWD include:

Further tests may examine the size and structure of the factor or how it binds to platelets and factor VIII. Genetic testing is useful in selected cases, particularly when a specific subtype or type 3 is suspected.

A full blood count and iron tests can look for anaemia. Routine clotting tests may also be done, but they can be normal in VWD. A normal screening test therefore does not rule it out.

Von Willebrand factor levels can rise temporarily because of stress, exercise, inflammation, pregnancy, or oestrogen. Testing may need to be repeated when you are otherwise well. A haematologist, a doctor specialising in blood conditions, may interpret the results and determine the type.

- Von Willebrand factor antigen: measures how much factor is present
- Von Willebrand factor activity: measures how well it works
- Factor VIII activity: measures the clotting protein carried by von Willebrand factor

Treatment and management

Treatment is matched to the VWD type, bleeding history, and situation. Some people need

treatment only before a procedure or after an injury. Others need treatment for frequent bleeding.

Options include:

Desmopressin does not work for every type. A supervised test may be used to see whether you respond. Because it can affect the body's water and salt balance, instructions about fluids and monitoring are important.

Von Willebrand factor concentrate may be used for major bleeding, surgery, childbirth, severe VWD, or when desmopressin is unsuitable. Treatment around an operation may also include laboratory monitoring.

Tell your dentist, surgeon, and other healthcare professionals about VWD before a procedure. Keep a record of your type, previous treatments, and treatment response. A medical identification card or bracelet can be useful.

If you have periods, tracking bleeding can help show whether treatment is working. Planning with haematology and obstetric teams is important before pregnancy and childbirth.

- Desmopressin: a medicine that releases stored von Willebrand factor into the blood
- Von Willebrand factor concentrate: replaces the missing or ineffective factor
- Antifibrinolytic medicines: help stop clots from breaking down, particularly in the mouth, nose, or uterus
- Hormone-based treatment: may reduce heavy menstrual bleeding
- Local treatment: pressure, surgical techniques, nasal care, or dental measures at the bleeding site
- Iron replacement: treats iron deficiency caused by repeated blood loss

Complications

Repeated blood loss can cause iron-deficiency anaemia. Heavy periods may interfere with school, work, sleep, exercise, and daily activities.

Serious bleeding can occur after major injury, surgery, or childbirth. Internal bleeding and bleeding into joints or muscles are more likely in severe VWD. Repeated joint bleeding can eventually damage a joint.

Treatment usually reduces these risks. Some clotting treatments can rarely contribute to excessive clotting, so higher-intensity treatment may require monitoring.

Prevention

Inherited VWD cannot currently be prevented. It is not caused by diet, exercise, or anything a parent did during pregnancy.

Bleeding episodes can often be prevented or reduced through planning. Tell healthcare professionals about the condition before surgery, dental work, pregnancy, or childbirth. Ask before taking non-prescription medicines that may affect bleeding, especially aspirin or anti-inflammatory pain medicines.

Genetic counselling can explain your inheritance pattern and the chance of passing VWD to a child. It provides information rather than directing reproductive decisions.

Acquired von Willebrand syndrome may improve when its underlying cause is treated, although this is not always possible.

Living with it and what to expect

Inherited VWD is lifelong, but the underlying type usually does not change. Bleeding symptoms and measured factor levels can change over the years. Levels may rise with age or pregnancy, but this does not necessarily remove the diagnosis or future bleeding risk.

Most people with mild or moderate VWD can take part in school, work, travel, exercise, and family life. People with severe VWD may need more regular specialist care and a detailed plan for injuries and procedures.

Routine follow-up may include reviewing bleeding, checking for iron deficiency, updating procedure plans, and discussing periods or pregnancy. Contact sports or activities with a high risk of head injury may need individual planning, especially with severe VWD.

During pregnancy, von Willebrand factor often rises. Levels can fall quickly after birth, creating a risk of delayed postpartum bleeding. Testing and a delivery plan may therefore be needed even if pregnancy itself has been uncomplicated.

Keep emergency and haematology contact details available when travelling. Carry enough information for an unfamiliar medical team to identify your condition and usual treatment.

When to get medical help

Seek emergency care for:

Use the emergency treatment plan provided by your specialist if you have one.

Contact your healthcare team promptly for bleeding that keeps returning, a period that becomes much heavier, bleeding after a procedure, or symptoms of anaemia. Medical assessment may be needed even if the bleeding has stopped.

Routine contact is appropriate before planned surgery, dental extraction, pregnancy, or starting a medicine that affects clotting. Regular follow-up is also useful if your bleeding pattern changes.

- Bleeding that is heavy, rapidly worsening, or will not stop with firm pressure
- A significant head injury, especially with headache, vomiting, confusion, weakness, or unusual sleepiness
- Vomiting blood, coughing up blood, or passing black or bloody stools
- Sudden severe pain or swelling that could represent internal, muscle, or joint bleeding
- Heavy vaginal bleeding with fainting, marked weakness, chest pain, or breathing difficulty
- Heavy bleeding during pregnancy or after childbirth
- Swelling or bleeding in the mouth or neck that affects breathing or swallowing

Common questions

Is von Willebrand disease the same as haemophilia?

No. Both are inherited bleeding conditions, but they involve different clotting proteins. Haemophilia usually involves factor VIII or IX. VWD involves von Willebrand factor and may also reduce factor VIII. Nosebleeds, bruising, and heavy periods are especially common in VWD.

Can I have VWD if my routine clotting tests are normal?

Yes. Common screening tests can be normal, particularly in mild VWD. Diagnosis requires tests that measure von Willebrand factor amount and activity, along with factor VIII. Because levels fluctuate, repeat testing may be needed.

Will my child inherit it?

This depends on the type and the gene changes involved. With many dominant forms, each child has a one-in-two chance of inheriting the altered gene. Type 3 usually requires an altered copy from both parents. Genetic counselling can explain the pattern in your family.

Can I take ibuprofen or aspirin?

These medicines can interfere with platelets and increase bleeding. Aspirin may also be prescribed for important heart or circulation reasons, so do not stop prescribed treatment on

your own. Ask a clinician or pharmacist which pain relief is appropriate for you.

Can I exercise with VWD?

Most people can exercise. Activity supports general health, strength, and joint stability. The safest choices depend on bleeding severity and injury risk. People with severe VWD may need a plan for higher-impact activities or sports involving collisions.

Does VWD affect periods and pregnancy?

It can. VWD may cause heavy periods and increase bleeding after miscarriage, procedures, or childbirth. Factor levels often rise during pregnancy but fall after delivery. Early planning with maternity and haematology teams can reduce risk.

Sources

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