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What is von Willebrand's disease?

Von Willebrand's disease (vWD) is one of the most common bleeding disorders that occur due to the low levels of or improper functioning of the von Willebrand factor (vWF) in the blood. This protein helps the blood cells to stick together to form a clot and thus, stops bleeding. An inadequate level of this factor can inhibit the clot formation resulting in excessive bleeding.

When von Willebrand factor doesn't work correctly, it takes a longer time for bleeding to stop. vWF also supports the clotting processes by carrying the clotting factor VIII. Hence, patients with vWD may have a low level of factor VIII.

Are von Willebrand's disease & Haemophilia the same?

Von Willebrand's disease and Haemophilia are similar since both these conditions occur due to the deficiency of clotting factors.

While von Willebrand's disease occurs due to the deficiency of von Willebrand's factor, Haemophilia occurs due to the inadequate levels of clotting factors VII and IX.

Haemophilia affects men more commonly than women, while von Willebrand disease can affect both men and women equally.

Also, von Willebrand's disease is the most common hereditary disorder linked to coagulation abnormalities. Haemophilia, on the other hand, is usually transmitted to male offspring through maternal lines with nearly one-third of the cases caused due to spontaneous mutation without any family history.

What are the symptoms of von Willebrand's disease?

The symptoms of vWD can be either too mild to be noticed or be extremely severe and frequent. The symptoms may begin at any age.

The common symptoms of vWD include:

- A tendency for easy bruising
- Lumpy bruises
- Long-lasting and heavy bleeding from injuries or cuts
- Long-lasting bleeding after a surgery or tooth removal
- Frequent bleeding from the gums and nose
- Long-lasting nosebleeds
- Bleeding in the gut, joints, and muscles
- Blood in the urine and stools
- Heavy menstrual periods in women
- Heavy bleeding during and after labour
- Symptoms of anaemia such as excessive fatigue, tiredness, and shortness of breath

What causes von Willebrand's disease?

Von Willebrand's disease occurs due to the deficiency of von Willebrand's factor in the blood.

Von Willebrand factor is a protein, which supports clot formation in the event of an injury to the blood vessels. A lack of adequate levels of this factor can prevent your blood from clotting normally. Therefore, the bleeding from the injured or ruptured blood vessel continues for a longer period.

Since vWD is an inherited condition, it occurs due to the abnormalities linked to the genes involved in the synthesis of the von Willebrand factor.

There are three types of vWD and the type of vWD a patient can develop usually depends on whether they have inherited copies of the abnormal gene from one or both parents.

The risk of inheriting type 1 vWD may also be affected by the child's blood group. It has been observed that people with 'O' blood are more likely to be affected than people with 'A' or 'B' blood.

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

The main types of vWD include:

- Type 1: It is the most common and the mildest form of vWD that occurs due to the reduced secretion of von Willebrand factor in the blood. Patients with type 1 vWD have excessive bleeding only when injured, have surgery, or tooth removal.
- Type 2: Type 2 vWD occurs when the von Willebrand factor does not work correctly due to which the bleeding occurs more frequently and heavily than in type 1 vWD.
- Type 3: It is the most severe but rarest form of vWD. Patients with type 3 vWD have a very low level of von Willebrand factor. They develop frequent bleeding from the nose, gums, mouth, and gut. Bleeding from joints and muscles may occur only after an injury.

How is von Willebrand's disease treated?

Currently, there is no cure for vWD. However, the condition can be controlled using medicines and other therapies, as discussed below.

The treatment suitable for a patient depends on factors such as:

- The severity of bleeding
- The level of vWF
- The type of surgery or dental treatment the patient needs to undergo
- History of bleeding
- Family's history of bleeding

What medications are used to treat von Willebrand's disease?

Antifibrinolytic agents

Patients with vWD are usually treated with antifibrinolytic agents such as Tranexamic Acid. Tranexamic Acid works by inhibiting the early breakdown of clots that have formed after an injury to the blood vessels.

Tranexamic Acid also blocks the action of substances that destroy fibrin, a protein responsible for providing stability to the blood clot. The use of antifibrinolytic agents is particularly useful for patients who suffer from frequent bleeding from the mouth.

It is also an effective preventative treatment for patients planning to undergo a dental procedure. Antifibrinolytic agents are available as tablets to be taken orally.

Side effects of Tranexamic Acid include:

- Nausea and vomiting
- Diarrhoea
- Muscle cramps
- Pain in the joints and muscles
- Pain in the stomach or abdomen
- Runny or stuffy nose
- Skin rash
- Changes in colour vision
- Headaches and migraines

The use of Tranexamic Acid should be avoided by patients who usually have blood in the urine as clots formed in the urine may block the urinary tract causing pain.

DDAVP (Desmopressin)

Desmopressin is another medication recommended for the management of Von Willebrand's disease. Desmopressin is a synthetic medicine usually given as an injection under the skin to help your blood form a clot by releasing the body's von Willebrand factor. It is also available as a nasal spray.

Side effects of Desmopressin include:

- Temporary redness and flushing of the face
- Dizziness
- Nausea
- Mild stomach pain
- Headaches
- Convulsions
- Allergic reactions

Replacement therapies

Replacement therapies for vWD include the infusion of concentrated blood-clotting factors, including von Willebrand factor and clotting factor VIII.

The administration of von Willebrand factor concentrate as an injection before a surgical or dental procedure is recommended to reduce the risk of bleeding. Patients with severe vWD might need to use these medications regularly to prevent excessive bleeding.

Your doctor may recommend the administration of von Willebrand factor concentrate if desmopressin was ineffective or not suitable for you.

Another replacement therapy for adult patients above 18 years of age includes the use of a genetically engineered product of von Willebrand factor. It is a recombinant factor made without plasma that can minimise the risk of viral infections and allergic reactions.

What complications can arise from von Willebrand's disease?

In rare cases, von Willebrand disease may cause uncontrollable bleeding leading to life-threatening consequences.

Other complications of vWD include:

- Pain and swelling in the joints and soft tissues may occur due to the abnormal bleeding in these tissues.
- Women may get heavy menstrual periods due to which they can develop iron deficiency anaemia.

What precautions can be taken?

- Inform your surgeon or dentist that you have vWD before undergoing any procedures. You may need to take medications to reduce your risk of bleeding during and after the procedure.
- Inform your doctor that you have vWD before receiving any vaccinations. Vaccines can be given just under your skin to avoid bleeding in the muscles.
- Avoid painkillers or anti-inflammatory drugs such as ibuprofen or aspirin as these drugs can increase the risk of bleeding. You may use acetaminophen to relieve pain.
- Avoid strenuous physical activities that can cause bruising such as wrestling, football, and hockey.