

Von Willebrand Syndrome (VWF): A Prolonged Bleeding Disorder

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DESCRIPTION

Von Willebrand Disease is most common inherited bleeding disorder (blood disorder) in which the blood does not clot properly. This is majorly caused due to the low concentration of Von Willebrand Factor (VWF) which is required for blood clotting.

When a person is injured and starts to bleed, VWF binds to factor VIII (8) (another clotting protein), and platelets in blood vessel walls, resulting to form a clot at the site of injury and stop the bleeding. But in case of von Willebrand Disease, the clot might take longer time to form or form incorrectly, leading to heavy bleeding. Cause for the disease may be either genetic (like haemophilia).

Diagnostic tools

VWD is often diagnosed by using series of blood tests along with your personal history with bleeds. Some of the tests have been listed below

- Von Willebrand factor antigen test-measures the amount of von willebrand factor present in blood.
- Von Willebrand activity test (ristocetin cofactor or RCF activity test) measures the working efficiency of Von Willebrand factor.
- Factor VIII activity test (factor VIII coagulant assay) measures the level of factor VIII and tests its working in blood
- Von willebrand multimers test-helps to classify the type and subtype of Von willebrand disease
- Platelet function tests-checks the working ability of platelets in blood

Symptoms

- Bleeding lasts for more than 10 minutes
- Nose bleeding
- Heavy Menstrual Bleeding among women
- Heavy bleeding occurs during or after childbirth
- Blood in the stool (feces) from bleeding into the stomach or intestines

- Bleeding in the mucous membranes, such as the gums, nose, and lining of the gastrointestinal system

Types of VWD

- Type 1 VWD-Low levels of VWF in their blood is observed (20%-50%=normal condition). In this case, VWF have low levels of factor VIII and doesn't help the platelets attach together in order to form a clot. It affects around 60%-80% of patients.
- Type 2 VWD-It has some subtypes- In type-2 VWD, normal levels of VWF are present but the factor doesn't function as it should. Type 2 VWD can be classified into subtypes, namely: Type 2A, type 2B, type 2M and type 2N, depending on the specific way the VWF is defective.
- In Type 2A, the VWF doesn't help the platelets attach together in order to form a clot.
- In Type 2B, the VWF attaches to platelets when there is no injury. The body removes the platelets attached to VWF, causing a reduced amount of both platelets and VWF in the blood when necessary to form a clot.
- In Type 2M, the VWF does not attach to the platelets as it should, resulting in decreased platelets' ability to form a clot during an injury.
- In Type 2N, the VWF attaches to the platelets normally. But, the VWF does not attach to Factor VIII, which is required to form a clot.
- Type 3 VWD-It affects around 5%-10% of patients. In this case, patient has a very low level or no VWF in their blood.
- Acquired VWD- This type of VWD in adults after a diagnosis of an autoimmune disease, such as lupus, or from heart disease or some types of cancer. It can also occur after taking certain medications.

Treating aids

Treatment prescribed for VWD depends on the type and severity of the disease. Treatment might not be needed for minor bleeds. Most commonly followed treatments have been listed:

- Desmopressin Acetate Injection
- Desmopressin Acetate Nasal Spray
- Factor Replacement Therapy

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- Use of Antifibrinolytic Drugs
- Birth Control Pills

Desmopressin acetate injection and desmopressin acetate nasal spray are used against Type 1 patients, to force the body release more VWF into the blood. It also helps increase the level of factor VIII in the blood. Factor Replacement Therapy is used to treat people with more severe forms of VWD or people with milder forms of VWD who do not respond well to the nasal spray. In this therapy, medicines rich in VWF and factor VIII are used to replace the missing factor in the blood.

Antifibrinolytic Drugs are used to help slow or prevent the breakdown of blood clots. Birth Control Pills increase the levels

of VWF and factor VIII in the blood and reduce menstrual blood loss

An accurate diagnosing is very vital for girls and women so as to avoid inessential and/or invasive treatments, like ablation, hysterectomy. Additionally, genetic counseling is important as this gene may be carried to the off springs. Currently, there is no way to prevent Erik Adolf Von Willebrand Disease, but research in gene therapy may provide a cure for those with severe disease in the future.