Patient Education

UNDERSTANDING VON WILLEBRAND DISEASE

Von Willebrand disease (vWD) is a genetic bleeding disorder caused by a deficient or defective von Willebrand factor (vWF). vWF is a clotting protein in the blood that binds factor VIII (an essential blood-clotting protein) and platelets in blood vessel walls, which help form a platelet plug during the clotting process. vWD is the most common inherited bleeding disorder in the United States, affecting approximately 1 percent of the U.S. population. It is carried on chromosome 12 and occurs equally in men and women. Learn more about the diagnosis, symptoms, and treatment of vWD below.

This Patient Education tear sheet was produced in collaboration with the National Hemophilia Foundation *(www.hemophilia.org)*.

What Are the Symptoms of vWD?

People with vWD experience frequent nosebleeds, easy bruising, and excessive bleeding during and after invasive procedures, such as tooth extractions and surgery. Women often experience menorrhagia, heavy menstrual periods that last longer than average, and hemorrhaging after childbirth.

There are three main types of vWD, based on qualitative or quantitative defects in vWF. A fourth type, acquired vWD, is not hereditary.

- **Type 1 vWD** is found in 60% to 80% of patients. People with type 1 vWD have a quantitative deficiency of vWF. Levels of vWF in the blood range from 20% to 50% of normal. The bleeding symptoms are usually mild.

- **Type 2 vWD** is found in 15% to 30% of patients. People with type 2 vWD have a qualitative deficiency in their vWF. Type 2 is broken down into four subtypes: type 2A, type 2B, type 2M, and type 2N, depending on the presence and behavior of multimers (molecular chains of vWF). Symptoms are mild to moderate.

- **Type 3 vWD** is found in 5% to 10% of patients. People with type 3 vWD have a quantitative deficiency of vWF. Symptoms are typically severe and include spontaneous bleeding episodes, often into joints and muscles.

- **Acquired vWD** in adults is associated with 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.

How is vWD Diagnosed?

To diagnose vWD, a hematologist or other doctor will perform various specialized tests to evaluate clotting capability, platelet function, and factor protein levels. A medical health history is important to help support diagnosis and determine if other relatives have experienced symptoms and might need to be tested for vWD.

A clotting factor test, called an assay, and tests measuring platelet function also may be performed. The vWF antigen test measures the amount of vWF in blood plasma. Patients with vWD typically have <50 percent of normal vWF in their plasma.

After vWD is confirmed, a test to determine the exact type of disease is performed.

It should be noted that diagnostic testing to confirm vWD may have to be repeated because fluctuating levels of vWF can obscure the results. vWF levels can also be elevated due to stress, exercise, the use of oral contraceptives, pregnancy, and hyperthyroidism.

How is vWD Treated?

Treatment for vWD depends on the specific diagnosis subtype and severity of a patient's disease.

Desmopressin Acetate

The mainstay of treatment is desmopressin acetate (DDAVP), the synthetic version of a natural hormone vasopressin. DDAVP stimulates the release of vWF from cells, which also increases FVIII levels.

DDAVP comes in two forms: injectable and nasal spray. Because DDAVP is an antidiuretic, causing the body to retain water, fluid restrictions are important so patients do not develop hyponatremia (reduced sodium in the bloodstream).

Clotting Factor Concentrates

There are a few clotting factor concentrates that are rich in vWF and are recommended for patients with vWD. These therapies are given by intravenous infusion.

Antifibrinolytic Agents

Aminocaproic acid and tranexamic acid are antifibrinolytic agents that prevent the breakdown of blood clots. These drugs are often recommended before dental procedures, to treat nose and mouth bleeds, and for menorrhagia in patients with vWD.

Antifibrinolytics are taken orally, as a tablet or liquid. Consultation on the appropriate vWD treatment options is best obtained by a pediatric or adult hematologist who specializes in the management of individuals with bleeding disorders.

NHF Resources

The National Hemophilia Foundation (NHF) is dedicated to finding better treatments and cures for inheritable bleeding disorders and to preventing the complications of these disorders through education, advocacy, and research.

- Visit the NHF website (*www.hemophilia.org*) or their Steps for Living website (*stepsforliving.hemophilia.org*) for more information on living with vWD or other bleeding disorders.

- Patients or caregivers can also contact HANDI, NHF's information resource center on hemophilia and other bleeding disorders, by emailing *handi@hemophilia.org* or calling 1.800.42.HANDI.

- You can also find articles on bleeding disorders in *HemAware*, the NHF's award-winning magazine, available at *www.hemaware.org*.

Cut out and give to a patient