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Von Willebrand Disease (VWD) Fact Sheet

What It Is

Von Willebrand disease (VWD) is the most common hereditary bleeding disorder.[1] It is caused by a deficiency or abnormality of the blood protein von Willebrand factor. People with VWD bruise easily[2] and experience prolonged bleeding.[1] VWD is classified by types, ranging from Type 1 (the most common and mild), to Type 3 (the most severe).[3]

Causes and Symptoms

Some patients with VWD have low or nonexistent levels of the von Willebrand factor (VWF), a protein in the blood that is necessary for normal blood clotting. In other cases, VWD presents with normal levels of VWF but the factor does not work as it should. In either case, platelets do not bind properly at the site of vessel injury. [5]

The signs and symptoms of the VWD depend on the type and severity of the disease. Women with VWD are more likely to experience heavy, prolonged menstruation. Other common symptoms of VWD include frequent nosebleeds, easy bruising and prolonged bleeding during and/or after dental procedures. More serious symptoms include bleeding into joints and internal organs (such as the stomach or intestines), and prolonged bleeding after surgery or childbirth.

Incidence and Prevalence

According to the Canadian Hemophilia Society, physicians estimate that VWD may affect as many as 1 in 1,000 people, or about 30,000 Canadians.[4]

VWD is typically passed from parent to child. A man or woman with the disease has a 50 percent chance of passing the gene on to his or her child. Parents who carry the disease but do not have bleeding symptoms can pass the gene to their children.

A family history of a bleeding disorder is the primary risk factor. [5]

Living with VWD

Bleeding can be mild or serious and can occur as a result of injury, or without any obvious cause. Individuals with more severe VWD should avoid unnecessary trauma, including contact sports.

VWD can be more complicated in women due to heavy and prolonged menstrual bleeding, which is often misdiagnosed as a gynecological problem.[6]

Special care may be required for the VWD patient during childbirth, surgery and dental procedures.

Diagnosis and Treatment

Because VWD is usually mild, many people with this disorder either are asymptomatic or fail to report any symptoms; thus, in most cases, the condition is undiagnosed. VWD can be easily diagnosed by blood tests. Establishing a comprehensive family medical history is also essential but not always adequate to identify VWD.[7]

The treatment of VWD depends on the type of VWD and its severity and is aimed at raising the levels of VWF circulating in the blood stream.

Desmopressin acetate: For patients with Type 1 and some Type 2a, this medicine is available through either an injection or a nasal spray to help the body release stored VWF into the bloodstream.

Replacement therapy: For patients with Type 1 – 3, factor concentrate medicines can be used to replace the missing VWF proteins.[8] Factor concentrates, such as Humate-P®, are licensed in Canada for use in treating adult and pediatric patients who experience spontaneous and trauma-induced bleeding episodes in severe VWD, and in mild or moderate VWD where the use of desmopressin is known or suspected to be inadequate.

Oral contraceptives: For female patients with mild VWD, oral contraceptives are often used to control heavy menstrual bleeding in women.

References

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