Von Willebrand disease

Little known bleeding disorder

Von Willebrand disease (VWD), the most common inherited bleeding disorder, affects up to 1% of the population. Research has shown, however, that as many as 99 of 100 people with VWD remain undiagnosed. The mission of the Canadian Hemophilia Society (CHS) is to improve the quality of life of all Canadians with inherited bleeding disorders. Hence, it has embarked on a 3-year campaign to raise awareness about VWD among health care providers, the general public, and people diagnosed with the disease.

Like hemophilia, VWD is a hereditary bleeding disorder. The disease is named after the Finnish physician, Erik Von Willebrand, who first described it in 1925. He realized that VWD was different from hemophilia, which in its severe form affects only men, whereas VWD affects both sexes equally. People with VWD either do not produce enough VWD factor (a gluelike protein essential for normal blood coagulation) or they produce it but it does not work properly. The most common type of VWD, type I, is a mild form characterized by moderately low levels of VWD factor.

Symptoms and consequences

The four warning signs of VWD are easy bruising; heavy menstrual periods; frequent nosebleeds; and prolonged bleeding after dental work, surgery, and childbirth. The disease affects both men and women but, because VWD can cause heavy menstrual bleeding and prolonged bleeding after childbirth, more women than men have noticeable symptoms. Unfortunately, too many women with undiagnosed VWD undergo surgical procedures and even hysterectomies to relieve symptoms.

Diagnosis

There is no cure for VWD, but it can be easily treated once diagnosis is established. Obtaining an accurate diagnosis is often challenging: routine blood tests cannot provide enough information for such a diagnosis. Specialized blood tests, including factor VIII coagulant activity, VWD factor antigen, and ristocetin cofactor and multimer analysis have to be ordered, usually by a hematologist.

Treatment

Minor bleeding episodes associated with type I VWD often do not require treatment. For more serious bleeding (such as very heavy menstrual periods), effective treatments are available, including desmopressin acetate, which is not a blood product and can be administered as a nasal spray or injected to boost the level of VWD factor to promote blood clotting. An antifibrinolytic agent, such as aminocaproic acid, can also be prescribed. These agents prevent breakdown of blood clots, which might also help in treating bleeds. People with more severe type II or III VWD might require treatment with factor VIII–VWD factor concentrate.

Canadian Hemophilia Society

The CHS, which was founded in 1953 by people with hemophilia, has a mandate to improve the quality of life of people with hemophilia or other inherited bleeding disorders in Canada and to find a cure. As a result of CHS advocacy efforts, a network of 23 bleeding disorder comprehensive care clinics have been established across the country. The CHS National Office is located in Montreal, Que, and there are 10 provincial chapters operated by volunteers as well as provincial and regional offices in Manitoba, Ontario, and Quebec.

For more information about VWD or the CHS, please contact Nathalie Byk, Coordinator of the VWD Program, by telephone at (514) 831-2838 or by e-mail at nbyk@hemophilia.ca.