Von Willebrand Disease (vWD)

What is von Willebrand disease?

Von Willebrand disease (vWD) is the most common inherited bleeding disorder. It is caused by a problem with a blood protein that is necessary to stop bleeding. The protein is called "von Willebrand factor" and helps form clots. When there is a problem with von Willebrand factor, blood takes longer to clot.

How common is von Willebrand disease?

vWD is the most common type of bleeding disorder. It affects 1% of the world's population.

How did my child get von Willebrand disease?

vWD is an inherited disease, which means it is passed from parent to child. It is caused by a defective gene on chromosome 12. Male and female children have an equal chance of inheriting vWD.

There are 2 ways of getting vWD:

- A parent has a defective gene that is passed onto the child. The parent may or may not have been diagnosed with vWD. The child's siblings have a chance of also having vWD.
- One of the child's genes undergoes a change (called '*spontaneous mutation*'). In this case the parents have normal genes and there is no risk to the child's siblings.

Children are born with vWD, but sometimes the diagnosis isn't made until they are older. One can often trace vWD through a family tree, but there are many reasons why there is sometimes no history of bleeding in the family.

Are there different types of von Willebrand disease?

Yes – vWD is divided into 3 categories: Type 1, 2 & 3.

In **Type 1** vWD there is a low amount of von Willebrand factor. This is the most common form and accounts for 75% of all cases of vWD. Bleeding can be mild to severe.

In **Type 3** vWD there is almost no von Willebrand factor. This is very rare and only affects 1/500,000 people. Bleeding is generally frequent and severe.

In **Type 2** vWD the von Willebrand factor does not work properly, even though there is a normal amount. This is the second most common form of vWD. There are several sub-types of Type 2 vWD, including 2A, 2B, 2N and 2M. These are all treated differently.

How is von Willebrand disease diagnosed?

Diagnosing vWD is not easy. The results of blood tests can vary from day to day, many factors change the level of vWF in the blood, and routine blood tests are often normal in people with vWD.

What are the symptoms of von Willebrand disease?

Common symptoms of vWD are:

- Easy bruising
- Bleeding from nose and gums – usually frequent and prolonged (>30 minutes)
- Prolonged bleeding from cuts
- Prolonged bleeding from minor surgical procedures, such as circumcision, tooth extraction, or when baby teeth fall out
- Heavy or prolonged bleeding during menstruation

However, some people with vWD have very few or no symptoms. In these cases a bleeding problem is only discovered when another person in the family is diagnosed with vWD or after they have serious injury or surgery.

How is von Willebrand disease treated?

There is no cure for vWD. Treatment is aimed at reducing the frequency and severity of bleeding, especially during planned surgical procedures. Treatment options depend on the type of vWD, and often require a "test dose" to see if a patient responds.

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Minor bleeding episodes often do not require medical treatment.

Treatments options include:

- Desmopressin (DDAVP)
- Tranexamic acid
- Fibrin glue
- Transfusions of von Willebrand factor concentrate or other blood products
- Women with heavy periods may benefit from hormonal treatment like oral contraceptives or an IUD

What should I do?

- Avoid medications like aspirin, ibuprofen, naproxyn, and blood thinners like warfarin and heparin
- Enjoy a healthy lifestyle with regular physical activity
- Have a plan for how to deal with mild, moderate and severe bleeding. This should include contact numbers for your hematology team and information on when to go to the Emergency Department.

For more information, please contact the Inherited Bleeding and Red Cell Disorders Program: (604) 875-2345 x 5335

Nurse Practitioners:
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After hours: Hematologist on-call: (604) 875-2161