Von Willebrand disease (VWD)

What is von Willebrand disease?

Von Willebrand disease (**VWD** – also known as *von Willebrand disorder*) is a bleeding disorder that affects both females and males. VWD is the most common hereditary bleeding disorder worldwide. It is estimated that 1 in 1,000 people have a form of VWD that needs medical treatment. More than 2,500 people have been diagnosed with VWD in Australia, but many more are thought to be undiagnosed.

People with VWD have a problem with a clotting protein in their blood called **von Willebrand factor** (**VWF**), which works with another protein **factor VIII (8)** to help control bleeding. They do not have enough VWF or it doesn't work the way it should. As a result it takes longer for blood to clot and for bleeding to stop.

What are the symptoms of VWD?

Bleeding problems can vary a lot between people with VWD, even in the same family. Some people experience little or no disruption to their lives unless they have serious injury or surgery, and others bleed quite often.

There can be bleeding problems with all forms of VWD. Any bleeding that occurs with VWD needs to be assessed and treated promptly.

Bleeding in people with VWD usually involves the mucous membranes, the delicate tissues that line body passages such as the:

- **Nose** (nose bleeds that occur often or are difficult to stop)
- **Mouth** (bleeding in the mouth)
- **Stomach and intestines** (gastrointestinal bleeding)
- **Uterus and vagina** (heavy/long menstrual periods; heavy bleeding for longer than usual after childbirth).

Other common symptoms are:

- **Easy bruising, bleeding for a long time** with **minor cuts**
- **Bleeding after injury, surgery or dental work** that continues for a long time
- **Anaemia/iron deficiency**, especially in females.

Severe VWD: a small number of people with VWD have the severe form. Without preventive treatment they may also have bleeding into joints and muscles that results in swelling and pain and 'spontaneous' bleeds (that occur for no obvious reason). Over time repeated bleeding can damage joints and muscles, causing arthritis and chronic pain.

How do you get VWD?

VWD is usually inherited. It occurs when there is a change (*mutation*) to the VWF gene. This gene change is passed down from parent to child. Some people with the VWF gene change have no bleeding symptoms while others do.

The VWD inheritance pattern is *autosomal* and affects both males and females in equal numbers. Most often there is a 1 in 2 chance of a parent passing the gene change onto their children. In some other cases, both parents can have the gene change, often without symptoms. If their child inherits the gene change from both of them (a 1 in 4 chance), the child will usually have the severe form of VWD.

Sometimes there is no family history of VWD. This is because there was a VWF gene change that occurred during reproduction before the baby was born. The person born with the VWF gene change can pass it onto their children.

How is VWD diagnosed?

Diagnosing VWD needs specialist medical and laboratory experience in VWD. A screening test may suggest a bleeding disorder but more specific tests and sometimes repeated testing will be needed to make an accurate diagnosis.

If you have questions about bleeding symptoms, speak to your doctor. If your doctor thinks your symptoms suggest VWD, you can be referred to a haematologist or Haemophilia Treatment Centre for blood tests.

Haemophilia Treatment Centres have a team of specialist doctors, nurses, social workers/psychologists, physiotherapists and laboratory services with expertise in bleeding disorders. There is at least one in a public hospital in every Australian state or territory and they provide both health care and support.

What are the different types of VWD?

There are three main types of VWD. Bleeding symptoms can vary from person to person within each type.

It is important to know which type of VWD a person has to make sure they receive appropriate treatment.

Type 1 VWD	The most common form. Usually low levels of VWF.
Type 2 VWD (sub-types 2A, 2B, 2M, 2N)	The level of VWF may be normal but the VWF doesn't work properly.
Type 3 VWD	Very rare. Very little or no VWF and low FVIII. Symptoms are more severe.

What is the treatment for VWD?

Treatment for VWD is individualised to each person, depending on their VWD type, the severity of their bleeding and the reason for the bleeding.

Some people with VWD who do not bleed often will only need treatment if they are having medical procedures, surgery, dentistry or have an injury or accident. Girls and women may need special treatment plans to manage heavy menstrual bleeding or to prepare for childbirth.

People with the severe form of VWD or who bleed often may need treatment to prevent bleeding called *prophylaxis*.

Types of treatment include:

- tranexamic acid
- synthetic hormones (eg, desmopressin/DDAVP)
- hormone treatments for females (eg, oral contraceptives or IUDs)
- VWF or VWF/FVIII clotting factor concentrates
- other new treatments, eg, that mimic the way factors work in the body.

Speak to your haematologist or pharmacist before taking medicines that may interfere with clotting, including aspirin, non-steroidal anti-inflammatory drugs (eg, ibuprofen), or other blood thinners.

For more information

Speak to your **doctor,** your local **Haemophilia Treatment Centre (HTC)** or visit **www.haemophilia.org.au.** In an emergency, seek medical attention promptly. Ask to speak to *the haematologist on call* at your local HTC hospital for specialist advice.

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Important Note:

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