Patient & Family Education Sheet
von Willebrand Disease (VWD)

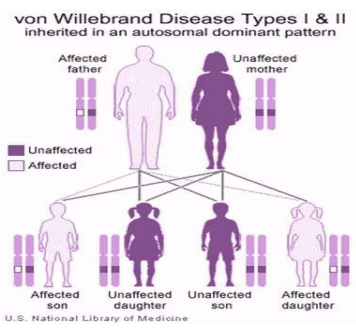
What is von Willebrand Disease? von Willebrand Disease (VWD) is the most common inherited bleeding disorder. It affects males and females equally, but females more commonly present with bleeding symptoms such as heavy periods. VWD is caused by low levels of a blood coagulation protein called von Willebrand Factor (VWF). This protein helps blood to clot (get thicker and stickier) by acting as glue for platelets and other coagulation proteins. Individuals with VWD often have symptoms of easy bleeding, such as frequent and large bruises, nosebleeds, or bleeding with dental work or surgery. For some individuals, symptoms are quite mild, but others may have more frequent or serious bleeding symptoms and need medications to control the bleeding. It can take time to diagnose VWD because VWF levels can vary with time and can be affected by many things, such as illness, physical activity, hormones and stress. Sometimes VWF testing needs to be repeated a few times to confirm your baseline levels.

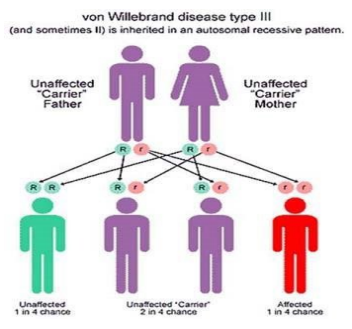
What are the symptoms of VWD?

- Frequent large bruises from minor accidents or injuries out of proportion to expected
- Frequent nosebleeds and/or nosebleeds that last longer than 20 minutes
- Teens and women can have long lasting (greater than 8 days) heavy periods
- Persistent or heavy bleeding from the gums while brushing teeth or having dental work
- Unexpected bleeding during or after surgical or dental procedures

What causes von Willebrand Disease?

- VWD is caused by a change in the VWF gene that gives the body instructions on how to make von Willebrand Factor (VWF). Each person has two copies of the VWF gene. An abnormal gene can be passed down from either parent to their children.
- Abnormality in just one of the VWF genes can cause VWD (typically causing type 1 or 2 VWD).
- Individuals with two abnormal VWF genes may have more severe types of VWD (type 3 or some type 2).
- Individuals with VWD (even in the same family) can have different types and severity of bleeding.





How is VWD diagnosed?

Understanding your bleeding symptoms and bleeding symptoms in other family members is important to help make a diagnosis of VWD (or other bleeding disorders). Special blood tests including a von Willebrand panel and potentially other blood coagulation tests are needed to learn why you may have abnormal bleeding.

There are 3 types of VWD

VWD Type	VWF Levels	Proportion on VWD patients	Symptoms and information
Type 1	<30%	About 75%	Common bleeding symptoms include: easy bruising, nosebleeds, heavy periods, bleeding with dental or surgical procedures.
Type 2	<50% and doesn't function normally	About 15%-25%	Bleeding symptoms are similar to those with Type 1 but may be more severe symptoms. There are multiple subtypes of type 2 VWD.
Type 3	Undetectable	Rare, <5%	Typically, a severe bleeding disorder. Bleeding symptoms include those noted above but can also include muscles and joints. This is there most rare and severe type of VWD.

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How is VWD Treated?

Treatment for VWD depends on the type of VWD and the location and severity of bleeding symptoms. For most people with VWD, treatments are only needed at times of bleeding or around the time of a dental or surgical procedure. For those with the most severe forms of VWD, sometimes regular bleed prevention medication (known as prophylaxis) is provided.
The most common VWD treatments include:

- ***Desmopressin (DDAVP):*** Desmopressin is a medication that causes cells to release more VWF and factor VIII into the blood. It can be administered through IV or as a nasal spray (150mg/mL). This treatment is used for most patients with Type 1 and some with Type 2 VWD.

- ***Antifibrinolytic drugs (aminocaproic acid [Amicar] or tranexamic acid [Lysteda]):*** These medicines slow down the body's breakdown of blood clots. They may be used alone or along with other treatments such as DDAVP and VWF concentrate. These medications can be taken by mouth or IV. Aminocaproic acid comes as a liquid; tranexamic acid only comes in pill form.

- ***VWF concentrate:*** The VWF clotting protein can be replaced with an intravenous (IV) infusion. This treatment is used for individuals who do not respond to DDAVP, need several days of treatment, or have more severe types of VWD. These are multiple VWF concentrate products available.

The DDAVP Challenge

Most individuals (~90%) with type 1 VWD respond to DDAVP, but not all. DDAVP challenge is performed to confirm an individual's response. This entails an infusion of DDAVP in a monitored setting (typically our CATCR). VWF panel is obtained prior to infusion, 1 hour after infusion and usually 4-hours post infusion. Symptoms experienced with DDAVP can include flushing. We have a detailed education sheet about the DDAVP challenge.

Are special precautions needed?

- If bleeding or injury occurs, start with basic first aid measures and apply pressure any bleeding areas.
- During nosebleeds, pinch the soft part of the nose and the child lean slightly forward to keep blood from the flowing down the throat.

Figure 1

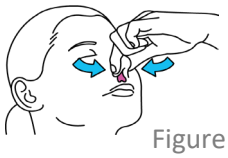

Figure 2

- People with VWD (and other bleeding disorders) should avoid unnecessary trauma, including contact sports with risk of head injury.
- Children with VWD should avoid NSAIDs (such as aspirin and ibuprofen). NSAID's interfere with platelet function and can increase the risk of bleeding. It is safe to take acetaminophen, which doesn't affect platelet function. Your child should also avoid blood-thinning medications such as Plavix or Coumadin.
- Girls with VWD may have heavy periods. Plan ahead! Access to extra pads or a change of clothes in case of accidents can help early on. There are many period focused treatments available to improve menstrual control. Collaboration with hematology and adolescent medicine or pediatric gynecology can be helpful in discussing the available options and providing the best plan for period management.

What does this mean for the future?

- You may need medicine to prevent or treat bleeding, especially around the time of surgical or dental procedures.
- Visit the hematology clinic at least once per year for ongoing education, evaluation, and bleed prevention and treatment updates.
- Always remember to tell the surgeon or dentist that you have VWD before any surgeries or procedures.
- Most individuals with VWD have a normal life expectancy and are expected to fully engage in school and work activities.

Helpful Websites & Educational Resources

Boston Bleeding Disorders Center https://www.childrenshospital.org/programs/boston-bleeding-disorders-center
National Bleeding Disorders Foundation www.hemophilia.org
New England Hemophilia Association www.newenglandhemophilia.org
World Foundation of Hemophilia www.wfh.org
HemAware www.hemaware.org
LA Kelley Communications www.kelleycom.com

BOSTON BLEEDING DISORDERS CENTER

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