**AANP poster submission on VWD guidelines DUE AUGUST 12 at 6 pm** by visiting <https://my.aanp.org/login.aspx?appUrl=https%3a%2f%2fspeakerabstracts.aanp.org%2f>

**AUTHORS:** Michelle L. Witkop, DNP, FNP-BC & Maria E. Santaella, RN-BC, MSN, CPHON

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**Title: Empowering Nurse Practitioners to Shorten Time to Diagnosis of von Willebrand Disease**

**Purpose**

Von Willebrand disease (VWD) is the most common inherited bleeding disorder known in humans, but there are numerous barriers to accurate and timely diagnosis. In January 2021, new evidence-based guidelines were released by the American Society of Hematology (ASH), the International Society on Thrombosis and Haemostasis (ISTH), the National Hemophilia Foundation (NHF), and the World Federation of Hemophilia (WFH) (James et al., 2021). The guidelines were designed to support patients, clinicians, and other health care professionals in promptly, accurately, and efficiently diagnosing VWD. Key recommendations in the guidelines include the use of bleeding-assessment tools when unusual bleeding is present and/or VWD is suspected; diagnostic assays; and the role of genetic vs. phenotypic testing for some types of VWD. Nurse practitioners can aid in identifying potential symptoms of VWD to help shorten the time from onset of symptoms to diagnosis.

**Background**

Von Willebrand disease is caused by deficiency or dysfunction of the multimeric glycoprotein von Willebrand factor (VWF), which plays key hemostatic roles in the circulation, including platelet adhesion and aggregation at sites of vascular injury, and acts as a chaperone for FVIII - another essential blood clotting protein (Sadler et al., 2000). It is inherited equally between men and women; however, women are more likely to present with symptoms because of gynecologic and obstetric bleeding.

In the primary-care setting, about 1% of the population is affected and requires medical attention for bleeding (Centers for Disease Control and Prevention [CDC], 2020). VWD patients experience excessive mucocutaneous bleeding, including heavy menstrual bleeding, epistaxis, easy bruising, prolonged bleeding from minor wounds and the oral cavity, and gastrointestinal bleeding, as well as bleeding after dental work, childbirth, and surgery, with musculoskeletal bleeding also seen in the most severe cases. (James et al., 2021).

Accurate and timely diagnosis of VWD remains a challenge, and patients may experience delays of 15 years or more from the onset of bleeding symptoms to a VWD diagnosis (Kirtava et al., 2004). Barriers to diagnosis include a lack of understanding of the difference between normal and abnormal bleeding symptoms, lack of clarity around an appropriate diagnostic approach, and limited availability and expertise of specialized laboratory testing. (James et al., 2021)

**Recognizing Signs and Symptoms of VWD: Role of the Nurse Practitioner**

For patients with a low probability of VWD (e.g., seen in the primary-care setting), the diagnostic guidelines recommend using a validated bleeding-assessment tool (BAT) as an initial screening test to determine the need for more specific blood testing vs. non-standardized clinical assessment.

During a patient’s appointment, if unusual bleeding is present and/or VWD is suspected, nurse practitioners can direct the patient to use the screening tools/BATs from Philipp and colleagues and Deforest and colleagues on the National Hemophilia Foundation’s “Better You Know” website for further assessment (Deforest et at., 2015; National Hemophilia Foundation, 2021; Phillip et al., 2011). Table 1 shows the questions included in the screening tools.

**Table 1: “Better You Know” Bleeding Assessment Tool Questions**

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| --- | --- |
| **Females** | **Males** |
| How many days did your period usually last, from the time bleeding began until it completely stopped? | Have you ever had a nosebleed? |
| How often did you experience a sensation of “flooding” or “gushing” during your period? | Have you ever had unexplained bruises or bruises larger than you think they should be? |
| During your period, did you ever have bleeding where you bled through a tampon or napkin in two hours or less? | Have you ever had bleeding from a small cut? (i.e. paper cut, nick from shaving) |
| Have you ever been treated for anemia? | Have you ever seen blood in your urine? |
| Has anyone in your family ever been diagnosed with a bleeding disorder? | Have you ever had bleeding from your intestines, stomach or bowel? (i.e. vomiting blood or had blood in your stools) |
| Have you ever had a tooth extracted or had dental surgery? | Have you ever had bleeding from the mouth? (i.e. bleeding after tooth brushing or flossing, or injury to the mouth). (This does NOT include tooth extraction at the dentist) |
| Did you have a problem with bleeding after tooth extraction or dental surgery? | Have you ever had a tooth/teeth pulled by the dentist? |
| Have you ever had surgery other than dental surgery? | Have you ever had surgery or major trauma (i.e. car accident)? |
| Did you have bleeding problems after surgery? | Have you ever had bleeding into a muscle? (a collection of blood in a muscle causes extreme pain and swelling) |
| Have you ever been pregnant? | Have you ever had bleeding into a joint? (a collection of blood in a joint that causes extreme pain and swelling) |
| Have you ever had a bleeding problem following delivery or after a miscarriage? | Have you ever had bleeding into the brain or spine? |
|  | Have you ever had bleeding in the whites of the eyes? |
|  | Have you ever had a nosebleed? |
|  | Have you ever had unexplained bruises or bruises larger than you think they should be? |

If a patient has an “At Risk” result from the screening tool, referral for laboratory testing is needed to determine if the patient has a bleeding disorder. In this case, referral to a federally funded hemophilia treatment center (HTC) is recommended for determination of required testing, a more definitive diagnosis, and proper treatment as appropriate. HTCs provide comprehensive care that addresses issues related to a variety of bleeding disorders, including diagnosis and education about the disorder. The interdisciplinary care team includes physicians (hematologists), nurses, social workers, physical therapists, and other health care providers who specialize in the treatment of (and care for) bleeding disorders (CDC, 2020). The publicly available CDC’s HTC directory can help NPs connect patients with the center nearest them as well as collaborate with the specialized team for guidance on care of these vulnerable patients with a high risk for unusual bleeding (CDC, 2020)

**Conclusions/Implications for Nurse Practitioners**

The new diagnostic guidelines for VWD were created to improve the identification of people with VWD while minimizing inappropriate testing. Nurse practitioners can play an important role early in the process by understanding the signs and symptoms of VWD, administering an initial BAT if appropriate, and knowing where to refer patients with possible VWD for more definitive testing and treatment. Sources such as the Better You Know website can assist in identifying at-risk individuals for referral to a specialized HTC. NPs who are equipped with this knowledge can contribute significantly to shortening the time to accurate diagnosis and prompt and effective treatment of this potentially debilitating bleeding disorder.

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