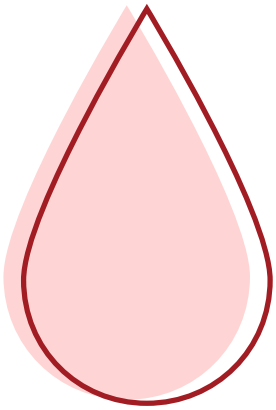
ASH ISTH NHF WFH Guideline Recommendations for the Diagnosis of von Willebrand Disease (VWD)



What it covers

- Evidence-based guidelines intending to improve accurate diagnosis of von Willebrand Disease (VWD), minimize inappropriate testing and avoid harms from over-diagnosis.

Why it matters

- VWD is the most common inherited bleeding disorder, yet accurate and timely diagnosis is challenging.
- Current barriers to accurate diagnosis of VWD include:
  - A lack of understanding of the difference between normal and abnormal bleeding symptoms.
  - A lack of/limited availability and expertise for specialized lab testing.
- It is important to improve accurate diagnosis to ensure access to care and minimize inappropriate testing and harms caused from over-diagnosis.

Who it affects

- **Hematologists, General Practitioners, Internists, Obstetricians, Gynecologists**
  - Health care professionals who provide screening for patients to accurately diagnose VWD.
- **Individuals who may be experiencing abnormal bleeding who should be evaluated for VWD**
  - Symptoms can disproportionately affect women, who may experience menstrual and postpartum hemorrhage.

What are the highlights

- Call to improve education around the value and use of bleeding assessment tools (BATs).
  - BATs are recommended as an initial screening tool for patients with a low probability of VWD (e.g., those seen in the primary care setting).
  - BATs are NOT recommended as a screening tool to decide whether to order specific blood testing for patients with intermediate/high probability of VWD (e.g., those referred to a hematologist or those with an affected first-degree relative).
- New recommendations suggest broadening the classification of VWD to be more inclusive of individuals who experience VWD-like bleeding but whose von Willebrand factor (VWF) levels used to confirm diagnosis of type 1 VWD do not meet the previously proposed diagnostic threshold of 30% or less.
- Suggestion to change the approach for a type 1 VWD patient with normalized VWF levels over time, specifically to reconsider diagnosis as opposed to removing diagnosis.
- Recommendations to use targeted genetic testing to diagnose type 2B VWD.

Total number of panel recommendations: 11

Reference: James PD, Connell NT, Ameer B, et al. ASH ISTH NHF WFH 2021 guidelines on the diagnosis of von Willebrand disease. Blood Adv. 2021;5(1):280-300.