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


ASH guidelines are reviewed annually by expert work groups convened by ASH. Resources derived from guidelines that require updating are removed from the ASH website.