National Hemophilia Foundation
for all bleeding disorders

Summary of the Guidelines on the Diagnosis of von Willebrand Disease (VWD)

(a collaborative effort of ASH ISTH NHF WFH)

1. If the chance of having VWD is low *(i.e. a person with no family history of VWD in the primary care setting)*, a validated bleeding-assessment tool *(BAT)* should be used to determine who needs specific blood testing.
2. If the chance of having VWD is intermediate *(i.e. person was referred to hematologist)* a BAT should not be used to decide whether to order specific blood testing.
3. If the chance of having VWD is high *(i.e. a person with a family history of VWD in a parent, sibling, or child)*, a BAT should not be used to decide whether to order specific blood testing.
4. For the diagnosis of VWD, newer tests that measure the platelet-binding activity of von Willebrand factor *(VWF) (i.e. VWF:GPIbM, VWF:GPIbR)* should be used in the laboratory rather than VWF ristocetin cofactor tests *(VWF:RCo)*.
5. For patients with previously confirmed type 1 VWD, who now have VWF levels that have normalized with age, a VWD diagnosis should be reconsidered based on the person's preferences rather than being removed.
6. To confirm a diagnosis of type 1 VWD, a person with bleeding symptoms needs a VWF level of 50% or less. A person with no bleeding symptoms needs a VWF level of 30% or less.
7. For people with suspected type 1C VWD, a desmopressin test with bloodwork drawn at 1- and 4- hours after the infusion should be completed to confirm increased VWF clearance.
8. Instead of using a platelet-dependent VWF activity/VWF antigen *(VWF:Ag)* ratio cutoff of less than 0.5, a higher cut off of less than 0.7 should be used to confirm type 2 VWD for patients with an abnormal initial VWD screen.
9. In patients with suspected types 2A, 2B, or 2M VWD, who are in need of additional testing, either a VWF multimer analysis or ratio of VWF collagen binding to antigen *(VWF:CB/VWF:Ag)* should be used in the laboratory.
10. In patients with suspected type 2A or 2B VWD, who are in need of additional testing, targeted genetic testing should be used over low-dose Ristocetin-induced platelet agglutination *(RIPA)* to identify type 2B.
11. In patients with suspected type 2N VWD, who are in need of additional testing, either VWF FVIII binding *(VWF:FVIIIB)* or targeted genetic testing *(if available)* should be used.

To learn more about specific lab tests, please go to:
NHF's Guide to Lab Tests, Screening Tools, and Health Exams

To read the VWD Guidelines in full, please go to:
https://ashpublications.org/bloodadvances/article/5/1/280/474888/ASH-ISTH-NHF-WFH-2021-guidelines-on-the-diagnosis

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