Von Willebrand Disease Testing

Click here for topics associated with this algorithm

Test Key

- VWF:Ag, von Willebrand Factor Antigen
- VWF:RCo, von Willebrand Factor Activity (Ristocetin Cofactor)
- FVIII, Factor VIII, Activity
- RCo:Ag ratio, ratio of von Willebrand Factor Activity (Ristocetin Cofactor) to von Willebrand Factor Antigen
- RIPA, Ristocetin-Induced Platelet Aggregation

INDICATIONS FOR TESTING
Excessive mucosal bleeding (eg, epistaxis, bleeding of gums, menorrhagia, postsurgical bleeding)

Initial Testing
- von Willebrand Panel, OR
- von Willebrand Panel with Reflex to von Willebrand Multimeric Analysis, OR
- von Willebrand Factor Antigen, von Willebrand Factor Activity (Ristocetin Cofactor), and Factor VIII, Activity

Results compatible with VWD

Results **not** compatible with VWD

Referral or repeat testing in 1-3 months if high suspicion for VWD

Additional Testing to Confirm Subtype
- Ratio of von Willebrand Factor Activity (Ristocetin Cofactor) to von Willebrand Factor Antigen
- von Willebrand Multimeric Panel
- Ristocetin-Induced Platelet Aggregation
- Genetic testing (refer to the Von Willebrand Disease ARUP Consult topic for more information)

Type 1
- VWF:Ag: <30 IU/dL
- VWF:RCo: <30 IU/dL
- FVIII: low or normal
- RCo:Ag ratio: >0.5-0.7
- Multimer pattern: normal but reduced intensity
- RIPA: usually normal

Type 2A
- VWF:Ag: <30-200 (commonly <50) IU/dL
- VWF:RCo: <30 IU/dL
- FVIII: low or normal
- RCo:Ag ratio: <0.5-0.7
- Multimer pattern: abnormal
- RIPA: often reduced at high ristocetin levels; no enhanced aggregation at low ristocetin levels
- Type 2A mutation present

Type 2B and Platelet Type
- VWF:Ag: <30-200 (commonly <50) IU/dL
- VWF:RCo: <30 IU/dL
- FVIII: low or normal
- RCo:Ag ratio: <0.5-0.7
- Multimer pattern: abnormal
- RIPA: abnormal; enhanced aggregation at low ristocetin concentrations
- Type 2B mutation present
- *GP1BA* variant present in platelet type

Type 2M
- VWF:Ag: <30-200 (commonly <50) IU/dL
- VWF:RCo: <30 IU/dL
- FVIII: low or normal
- RCo:Ag ratio: <0.5-0.7
- Multimer pattern: normal pattern, may have reduced intensity
- RIPA: reduced at high ristocetin concentrations
- Type 2M mutation present

Type 2N
- VWF:Ag: 30-200 IU/dL
- VWF:RCo: 30-200 IU/dL
- FVIII: mildly or markedly low
- RCo:Ag ratio: >0.5-0.7
- Multimer pattern: normal
- RIPA: normal
- Type 2N mutation present

Type 3
- VWF:Ag: Absent
- VWF:RCo: Absent
- FVIII: severely low (<10 IU/dL)
- RCo:Ag ratio: n/a
- Multimer pattern: no VWF present
- RIPA: absent

References
- 2012 Clinical Practice Guideline on the Evaluation and Management of von Willebrand Disease (VWD). American Society of Hematology. Washington DC [Revised: 2012; Accessed: Feb 2019]
- Nichols WL , Hultin MB , James AH, et al. von Willebrand disease (VWD): evidence-based diagnosis and management guidelines, the National Heart, Lung, and Blood Institute (NHLBI) Expert Panel report (USA). Haemophilia. 2008;14(2):171-232.
- Favaloro EJ, Bodó I, Israels SJ, Brown SA. von Willebrand disease and platelet disorders. Haemophilia. 2014; 20 Suppl 4: 59-64.

Content reviewed: June 2020 Last updated: June 2020