**Von Willebrand Disease Testing**

**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 (e.g., 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**
**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)

**Results not compatible with VWD**
- Referral or repeat testing in 1-3 months if high suspicion for VWD

**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
- RIPA: absent

**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**