BACKGROUND:
von Willebrand factor (VWF) propeptide is encoded on the gene for VWF and directs intracellular processing of VWF. Both VWF propeptide and mature VWF circulate in plasma and are present in endothelial cell Weibel-Palade bodies and platelet alpha granules. In normal individuals, VWF and VWF propeptide antigen levels are within the reference intervals; both levels may be low in type 1 von Willebrand disease. Patients with increased clearance of VWF will have normal VWF propeptide levels and a VWF antigen level that is decreased compared to the VWF propeptide antigen. This has been observed in some congenital von Willebrand disease variants (1C, 2A, 2B) and in a subset of individuals with acquired von Willebrand disease.

Unlike Factor VIII inhibitors in hemophilia A, VWF antibodies usually cause increased clearance of VWF from circulation rather than inhibiting VWF function. Therefore, most VWF antibodies are not detected in the standard mixing studies used for coagulation factor inhibitors. Measurement of VWF propeptide is helpful in diagnosing some cases of acquired von Willebrand disease.

METHOD:
ELISA with fluorescence detection

REASONS FOR REFERRAL:
• Diagnosis of acquired von Willebrand disease
• Evaluation of patients with increased clearance of von Willebrand factor

LIMITATIONS:
• VWF propeptide antigen should be correlated with VWF antigen, VWF ristocetin cofactor activity, VWF multimers and clinical history by the ordering physician to arrive at the appropriate diagnosis.
• VWF levels will be elevated by acute phase response and DDAVP.
• Serum is not an acceptable specimen type as results are significantly higher than in citrated plasma.

REFERENCE RANGE:
55 – 141 Iu/dL
SPECIMEN REQUIREMENTS:
0.5 ml citrated plasma, frozen in a plastic tube.

SHIPPING REQUIREMENTS:
Place the frozen specimen and the requisition into plastic bags, seal and place in an insulated container. Surround with at least 5 pounds of dry ice. Seal the insulated container, place into a sturdy cardboard box, and tape securely. Ship the package in compliance with your overnight carrier guidelines. Label with the following address:

    Client Services/Hemostasis Reference Laboratory
    BloodCenter of Wisconsin
    638 N. 18 St.
    Milwaukee, WI 53233-2121
    800-245-3117, ext. 6250

TURNAROUND TIME: 7 - 10 days

CPT CODES: 83520