Anti-von Willebrand Factor Antibody Assay

The Anti-von Willebrand factor (VWF) Antibody Assay quantifies anti-VWF antibodies (IgG, IgM) found in patient plasma to support diagnosis of acquired von Willebrand disease.

Background: Acquired von Willebrand disease (aVWD) generally presents as new or unexplained bleeding, attributed to deficiency of von Willebrand factor (VWF) function. There are 5 major pathogenic mechanisms that result in aVWD.[1,2] These include autoantibody mediated clearance (~ 2% of cases, associated with other autoimmune diseases, such as SLE, lymphoproliferative neoplasms, monoclonal gammopathies, or idiopathic), shear-induced VWF degradation (~ 21% of cases, associated with aortic stenosis, hypertrophic obstructive cardiomyopathy, left ventricular assist devices, and extracorporeal membrane oxygenation), adsorption onto cells (up to 68% of cases, such as myeloproliferative or lymphoproliferative neoplasm)[3], proteolysis (as may occur with essential thrombocythemia or disseminated intravascular coagulation),[4] or decreased synthesis (such as hypothyroidism or with valproic acid therapy). Laboratory evaluation may show abnormal blood counts if there is an associated disorder, elevated aPTT ± reduced FVIII if VWF antigen is sufficiently reduced, and abnormal von Willebrand profile (with reduced quantitative VWF levels and potentially abnormal ratio of VWF activity to antigen and abnormal multimers depending upon the mechanism of disease and severity of acquired VWF abnormality). In cases associated with increased VWF clearance, the ratio of VWF propeptide antigen to VWF antigen will be increased.[5] Therapy is aimed at controlling acute bleeding,[6-8] prevention of bleeding during risk procedures[1], and achieving stable remission by addressing the underlying mechanism causing aVWD.[1,2]

Indications for testing:

1. Generating supportive evidence for an autoimmune mechanism of aVWD.
2. Understanding the basis for poor response to VWF replacement therapy in a patient with Type 3 VWD.
3. Monitoring immunomodulatory therapy of a patient with either immune aVWD or those refractory to replacement therapy.

Test method:

ELISA

Assay sensitivity and limitations:

Anti-VWF antibodies other than IgG or IgM will not be detected by this assay.

A positive test result provides evidence supporting an immune mechanism of disease, however, a negative result does not necessarily exclude autoimmune disease. In a well-defined population, assay sensitivity and specificity >90%. Due to the rare incidence of autoimmune acquired VWD, a sufficient number of samples from well-defined patients was not available to generate estimates of clinical sensitivity and specificity.

Reporting of Results:

Positive, Negative, Borderline: results are reported in Arbitrary units (AU) and calibrated against a high titer recombinant humanized anti-VWF antibody.

Specimen requirements:

0.5 ml citrated plasma aliquot, frozen in a plastic tube

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SHIP

Shipping requirements:

Place the frozen specimen and the completed test requisition form 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:

Send to:
Versiti Diagnostic Laboratories
638 N. 18th Street
Milwaukee, WI 53233

800-245-3117, ext. 6250

ORDER

Required forms:

Please complete all pages of the requisition form, including patient history on page 2 for optimal results interpretation.

CPT Codes/Billing/Turnaround time:

Order Code: 1056

CPT Codes: For suggested CPT codes, visit Versiti.org/test-catalog

Turnaround Time: 14 days

References

1. Tiede, A., et al., How I treat the acquired von Willebrand syndrome. Blood, 2011. 117(25): p. 6777-6785.
2. Federici, A.B., et al., Current diagnostic and therapeutic approaches to patients with acquired von Willebrand syndrome: a 2013 update. Semin Thromb Hemost, 2013. 39(2): p. 191-201.
3. Rottenstreich, A., et al., Factors related to the development of acquired von Willebrand syndrome in patients with essential thrombocythemia and polycythemia vera. Eur J Intern Med, 2017. 41: p. 49-54.
4. Vedachalam, S., et al., Treatment of gastrointestinal bleeding in left ventricular assist devices: A comprehensive review. World J Gastroenterol, 2020. 26(20): p. 2550-2558.
5. Haberichter, S.L., et al., Assay of the von Willebrand factor (VWF) propeptide to identify patients with type 1 von Willebrand disease with decreased VWF survival. Blood, 2006. 108(10): p. 3344-51.
6. Biguzzi, E., S.M. Siboni, and F. Peyvandi, How I treat gastrointestinal bleeding in congenital and acquired von Willebrand disease. Blood, 2020. 136(10): p. 1125-1133.
7. Ahsan, I., et al., Clinical Approach to Manage Gastrointestinal Bleeding with a Left Ventricular Assist Device (LVAD). Cureus, 2019. 11(12): p. e6341.
8. Littlefield, A.J., et al., A reappraisal of the pharmacologic management of gastrointestinal bleeding in patients with continuous flow left ventricular assist devices. Heart Fail Rev, 2021. 26(2): p. 277-288.

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