BLOODCENTER of WISCONSIN™

VON WILLEBRAND DISEASE TYPE 2B EVALUATION

The purpose of the platelet-VWF binding assay is to allow identification of type 2B von Willebrand Disease, and to distinguish it from platelet-type von Willebrand disease.

BACKGROUND:

von Willebrand disease (VWD) is a collection of hemorrhagic disorders that are characterized by either quantitative or qualitative defects of von Willebrand factor (VWF).[1] Type 2B von Willebrand disease is an inherited disorder caused by a qualitative abnormality of VWF which leads to increased binding of high molecular weight VWF multimers to normal platelets.[2] Clearance of plasma VWF and circulating platelets results in a hemorrhagic tendency. In a somewhat similar manner, a qualitative platelet disorder with increased affinity of platelet glycoprotein Ib for normal VWF results in platelet-type VWD.[3] Screening laboratory studies in both disorders are non-specific, revealing variably depressed levels of factor VIII coagulant, VWF antigen, VWF ristocetin cofactor activity and platelet count. With more specific testing, multimer analysis may show loss of the largest multimers, and the increased association of patient VWF for patient platelets is revealed by abnormal low-dose ristocetin-induced platelet aggregation in both conditions.[1] The platelet-VWF binding assay can differentiate 2B VWD from platelet-type VWD.

REASONS FOR REFERRAL:

Differentiation of type 2B VWD from platelet-type VWD is essential in order to provide appropriate replacement therapy in bleeding patients.[4] Replacement therapy with VWF containing concentrate is appropriate in type 2B VWD, while platelet transfusion is more appropriate in platelet von Willebrand disease. The role of DDAVP in the treatment of these conditions remains controversial.

METHOD:

In the platelet-VWF binding assay, a monoclonal antibody to VWF is used to monitor the ability of patient plasma VWF to bind to formalin fixed platelets in the presence of low dose ristocetin.[5] To assist in the interpretation of the platelet-VWF binding assay, VWF multimer analysis is also performed.

LIMITATIONS:

The binding assay may be less sensitive in patients with VWF antigen levels below 10 u/dl. Other types of VWD are not detected by the platelet-VWF binding assay, and low-dose ristocetin induced platelet aggregation may be helpful in patient evaluation. Samples taken after transfusion therapy may reflect the characteristics of transfused VWF.

PO Box 2178
Milwaukee, WI 53201-2178
Location/Sample Deliveries:
638 N. 18th St. Milwaukee, WI 53233-2121
p800-245-3117 | f414-937-6202 | www.bcw.edu

NORMAL VALUES:
No increased binding of patient's VWF to normal platelets at low concentration of ristocetin. This result is not consistent with type 2B VWD. (Abnormal result is increased binding of VWF to normal platelets at low concentration of ristocetin, consistent with 2B VWD.)

SPECIMEN REQUIREMENTS:
One 1.0 ml and one 0.5 ml aliquots of citrated plasma frozen in plastic tubes, shipped frozen on dry ice.

SHIPPING REQUIREMENTS:
Place the frozen specimen and the test requisition form in plastic bags, seal and surround with at least 5 pounds of dry ice in a Styrofoam container. Place the sealed Styrofoam container in 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. 18th St.
Milwaukee, WI 53233
800-245-3117, ext. 6129

TURNAROUND TIME: 7-10 days

CPT CODES:
83519, 85247 (Includes 2B VWD binding test and VWF multimer analysis.)

REFERENCES:
1. Montgomery RR, Coller BS: von Willebrand Disease, in Colman RW, Hirsh J, Marder VJ, Salzman EW (eds): Hemostasis and Thrombosis: Basic Principles and Clinical Practice. Philadelphia PA, Lippincott 1994.

2. Ruggeri ZM, Pareti FI, Mannucci PM, Ciaverella N, Zimmerman TS: Heightened interaction between platelets and factor VIII/von Willebrand factor in a new subtype of von Willebrand=s disease. New England Journal of Medicine 1980; 302:1047.

3. Weiss HJ, Meyer D, Rabinowitz R, et al.: Pseudo-von Willebrand disease: an intrinsic platelet defect with aggregation by unmodified human FVIII/von Willebrand factor and enhanced adsorption of its high molecular weight multimers. New England Journal of Medicine 1982; 306:326.

4. Scott JP, Montgomery RR: Therapy of von Willebrand disease. Seminars in Thrombosis and Hemostasis. 1993; 19:37.

5. Scott JP, Montgomery RR: The rapid differentiation of type IIb von Willebrands disease from platelet-type (pseudo-) von Willebrands disease by the Aneutral monoclonal antibody binding assay. American Journal of Clinical Pathology 1991; 96:723.

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