

VWF (GMA-022): sc-65973

BACKGROUND

Von Willebrand disease is a congenital bleeding disorder caused by defects in the von Willebrand factor protein (VWF). VWF is a multimeric glycoprotein that is found in endothelial cells, plasma and platelets, and it is involved in the coagulation of blood at injury sites. VWF acts as a carrier protein for Factor VIII, a cofactor required for coagulation, and it promotes platelet adhesion and aggregation. Several factors are known to stimulate the binding of VWF to platelets, including glycoprotein 1b, ristocetin, botrocetin, collagen, sulphatides and heparin. Of the several domains contained within VWF, the A1, A2 and A3 domains have been shown to mediate this activation. VWF is thought to undergo a variety of posttranslational modifications that influence the affinity and availability for Factor VII, including cleavage of the propeptide and formation of N-terminal intersubunit disulfide bonds.

REFERENCES

1. Naiem, M., Gerdes, J., Abdulaziz, Z., Sunderland, C., Allington, M., Stein, H. and Mason, D.Y. 1982. The value of immunohistological screening in the production of monoclonal antibodies. *J. Immunol. Methods* 50: 145-160.
2. Wise, R.J., Dorner, A.J., Krane, M., Pittman, D.D. and Kaufman, R.J. 1991. The role of von Willebrand factor multimers and propeptide cleavage in binding and stabilization of Factor VIII. *J. Biol. Chem.* 266: 21948-21955.
3. Fischer, B.E., Kramer, G., Mitterer, A., Grillberger, L., Reiter, M., Mundt, W., Dorner, F. and Eibl, J. 1996. Effect of multimerization of human and recombinant von Willebrand factor on platelet aggregation, binding to collagen and binding of coagulation Factor VIII. *Thromb. Res.* 84: 55-66.
4. Ward, C.M., Tetaz, T.J., Andrews, R.K. and Berndt, M.C. 1997. Binding of the von Willebrand factor A1 domain to histone. *Thromb. Res.* 86: 469-477.
5. Jenkins, P.V., Pasi, K.J. and Perkins, S.J. 1998. Molecular modeling of ligand and mutation sites of the type A domains of human von Willebrand factor and their relevance to von Willebrand's disease. *Blood* 91: 2032-2044.
6. Bendetowicz, A.V., Morris, J.A., Wise, R.J., Gilbert, G.E. and Kaufman, R.J. 1998. Binding of Factor VIII to von Willebrand factor is enabled by cleavage of the von Willebrand factor propeptide and enhanced by formation of disulfide-linked multimers. *Blood* 92: 529-538.
7. Mazurier, C., Ribba, A.S., Gaucher, C. and Meyer, D. 1998. Molecular genetics of von Willebrand disease. *Ann. Genet.* 41: 34-43.

CHROMOSOMAL LOCATION

Genetic locus: VWF (human) mapping to 12p13.31.

SOURCE

VWF (GMA-022) is a mouse monoclonal antibody raised against VWF of human origin.

PRODUCT

Each vial contains 100 µg IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

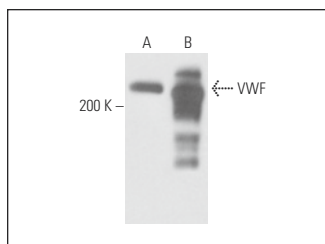
VWF (GMA-022) is recommended for detection of VWF of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)].

Suitable for use as control antibody for VWF siRNA (h): sc-36828, VWF shRNA Plasmid (h): sc-36828-SH and VWF shRNA (h) Lentiviral Particles: sc-36828-V.

Molecular Weight of VWF: 250 kDa.

Positive Controls: human platelet extract: sc-363773 or HUV-EC-C whole cell lysate: sc-364180.

DATA



VWF (GMA-022): sc-65973. Western blot analysis of VWF expression in human platelet extract (A) and HUV-EC-C whole cell lysate (B).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.



See **VWF (C-12): sc-365712** for VWF antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor® 488, 546, 594, 647, 680 and 790.