

## DESCRIPTION

|                           |                                                                                                                                                                                                                                                                             |
|---------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Species Reactivity</b> | Human                                                                                                                                                                                                                                                                       |
| <b>Specificity</b>        | Detects human vWF-A2 (ADAMTS13-cleaved) in Western blots. This antibody detects a nascent epitope that is generated upon cleavage of recombinant human (rh) vWF-A2 domain with ADAMTS13. It does not detect intact rhvWF-A2 domain.                                         |
| <b>Source</b>             | Monoclonal Mouse IgG <sub>1</sub> Clone # 490628                                                                                                                                                                                                                            |
| <b>Purification</b>       | Protein A or G purified from hybridoma culture supernatant                                                                                                                                                                                                                  |
| <b>Immunogen</b>          | KLH-conjugated human vWF synthetic peptide<br>Asp1596-Tyr1605<br>Accession # NP_000543                                                                                                                                                                                      |
| <b>Conjugate</b>          | Alexa Fluor 594<br>Excitation Wavelength: 590 nm<br>Emission Wavelength: 617 nm                                                                                                                                                                                             |
| <b>Formulation</b>        | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide<br><br>*Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions. |

## APPLICATIONS

**Please Note:** Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

**Western Blot** Optimal dilution of this antibody should be experimentally determined.

## PREPARATION AND STORAGE

**Shipping** The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.

**Stability & Storage** Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

## BACKGROUND

von Willebrand Factor (vWF) is a large, multimeric glycoprotein made by endothelial cells and megakaryocytes. The pre-pro-vWF protein contains 2813 amino acids (aa), which consists of 22 aa signal peptide, 741 aa propeptide, and mature vWF monomer of 2050 aa (1-4). The pro-vWF undergoes dimerization in the endoplasmic reticulum (ER) through C-terminal "cysteine-knot" (CK) domain. The pro-vWF dimers are transported to Golgi and form multimers by forming disulfide bond in amino-terminal region of the mature form. The proteolytic processing of pro-region also occurs in Golgi. The matured vWF is stored in Weibel-Pallade bodies in endothelial cells and granules in megakaryocytes and platelets. The unusually-large vWF (ulvWF) multimers released from cells are very efficient in binding to platelets to form thrombus. The population of these highly active ulvWF multimers is controlled by a specific protease, ADAMTS13, which cleaves between residues Tyr1605 and Met1606 in the A2 domain of vWF. In the plasma, vWF appears as a series of large and intermediate multimers with molecular masses from several thousand to 500 kDa. vWF also performs hemostatic functions (3-5). In a high shear-stressed environment, vWF undergoes conformational change to expose a binding site for glycoprotein Iba. As a result, vWF facilitates aggregation of platelets. In addition to platelet binding, vWF binds coagulation factor VIII to increase the lifetime of FVIII in plasma. The purified rhvWF-A2 contains the A2 domain of vWF.

## PRODUCT SPECIFIC NOTICES

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