SunLong Biotech Co.,LTD
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# Rabbit Anti-VWF antibody

SLM-52775R

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|---|---|
| **Product Name** | VWF |
| **Chinese Name** | 血管假性血友病因子 Recombinant rabbit monoclonal anti |
| **Alias** | Von Willebrand Factor; Coagulation factor VIII; F8VWF; Factor VIII related antigen; von Willebrand antigen 2; Von Willebrand antigen II; Von Willebrand disease; VWD; VWF_HUMAN |
| **Research Area** | Cardiovascular Cell biology immunology |
| **Immunogen Species** | Rabbit |
| **Clonality** | Monoclonal |
| **Clone NO.** | 9C2 |
| **React Species** | Human,Mouse |
| **Applications** | WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 （Paraffin sections need antigen repair）<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Theoretical molecular weight** | 309kDa |
| **Cellular localization** | Extracellular matrix Secretory protein |
| **Form** | Liquid |
| **Concentration** | 1mg/ml |
| **immunogen** | Recombinant human VWF protein |
| **Lsotype** | IgG |
| **Purification** | affinity purified by Protein A |
| **Buffer Solution** | 1*TBS (pH7.4), Human,Mouse5% BSA, 40% Glycerol. Preservative: Human,Mouse5% Sodium Azide. |
| **Storage** | Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. |
| **Attention** | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |

**PubMed**

PubMed

Von Willebrand Factor (VWF) was previously known as Factor VIII related antigen. VWF is synthesized exclusively by endothelial cells and megakaryocytes, and stored in the intracellular granules or constitutively secreted into plasma. This glycoprotein functions as both an antihemophilic factor carrier and a platelet vessel wall mediator in the blood coagulation system. Important in the maintenance of homeostasis, it participates in platelet vessel wall interactions by forming a noncovalent complex with coagulation factor VIII at the site of vascular injury. The Von Willebrand factor has functional binding domains to platelet glycoprotein Ib, glycoprotein IIb/IIIa, collagen and heparin. Mutations in this gene or deficiencies in this protein result in Von Willebrand's disease. VWD is characterized by frequent bleeding (gingival, minor skin quantitative lacerations, menorrhagia, etc.).

**Product Detail**

**Function:**
Important in the maintenance of hemostasis, it promotes adhesion of platelets to the sites of vascular injury by forming a molecular bridge between sub-endothelial collagen matrix and platelet-surface receptor complex GPIb-IX-V. Also acts as a chaperone for coagulation factor VIII, delivering it to the site of injury, stabilizing its heterodimeric structure and protecting it from premature clearance from plasma.

**Subunit:**
Multimeric. Interacts with F8.

**Subcellular Location:**
Secreted. Secreted, extracellular space, extracellular matrix. Note=Localized to storage granules.

**Tissue Specificity:**
Plasma.

**Post-translational modifications:**
All cysteine residues are involved in intrachain or interchain disulfide bonds.
N- and O-glycosylated.

**DISEASE:**
Defects in VWF are the cause of von Willebrand disease type 1 (VWD1) [MIM:193400]. A common hemorrhagic disorder due to defects in von Willebrand factor protein and resulting in impaired platelet aggregation. Von Willebrand disease type 1 is characterized by partial quantitative deficiency of circulating von Willebrand factor, that is otherwise structurally and functionally normal. Clinical manifestations are mucocutaneous bleeding, such as epistaxis and menorrhagia, and prolonged bleeding after surgery or trauma.
Defects in VWF are the cause of von Willebrand disease type 2 (VWD2) [MIM:613554]. A hemorrhagic disorder due to defects in von Willebrand factor

protein and resulting in impaired platelet aggregation. Von Willebrand disease type 2 is characterized by qualitative deficiency and functional anomalies of von Willebrand factor. It is divided in different subtypes including 2A, 2B, 2M and 2N (Normandy variant). The mutant VWF protein in types 2A, 2B and 2M are defective in their platelet-dependent function, whereas the mutant protein in type 2N is defective in its ability to bind factor VIII. Clinical manifestations are mucocutaneous bleeding, such as epistaxis and menorrhagia, and prolonged bleeding after surgery or trauma. Defects in VWF are the cause of von Willebrand disease type 3 (VWD3) [MIM:277480]. A severe hemorrhagic disorder due to a total or near total absence of von Willebrand factor in the plasma and cellular compartments, also leading to a profound deficiency of plasmatic factor VIII. Bleeding usually starts in infancy and can include epistaxis, recurrent mucocutaneous bleeding, excessive bleeding after minor trauma, and hemarthroses.

**Similarity:**
Contains 1 CTCK (C-terminal cystine knot-like) domain.
Contains 4 TIL (trypsin inhibitory-like) domains.
Contains 3 VWFA domains.

**SWISS:**
P04275

**Gene ID:**
7450

**Database links:**

Entrez Gene: 280958 Cow

Entrez Gene: 399544 Dog

Entrez Gene: 7450 Human

Entrez Gene: 399543 Pig

Entrez Gene: 116669 Rat

Omim: 613160 Human

SwissProt: Q28295 Dog

SwissProt: P04275 Human

SwissProt: Q28833 Pig

SwissProt: Q62935 Rat

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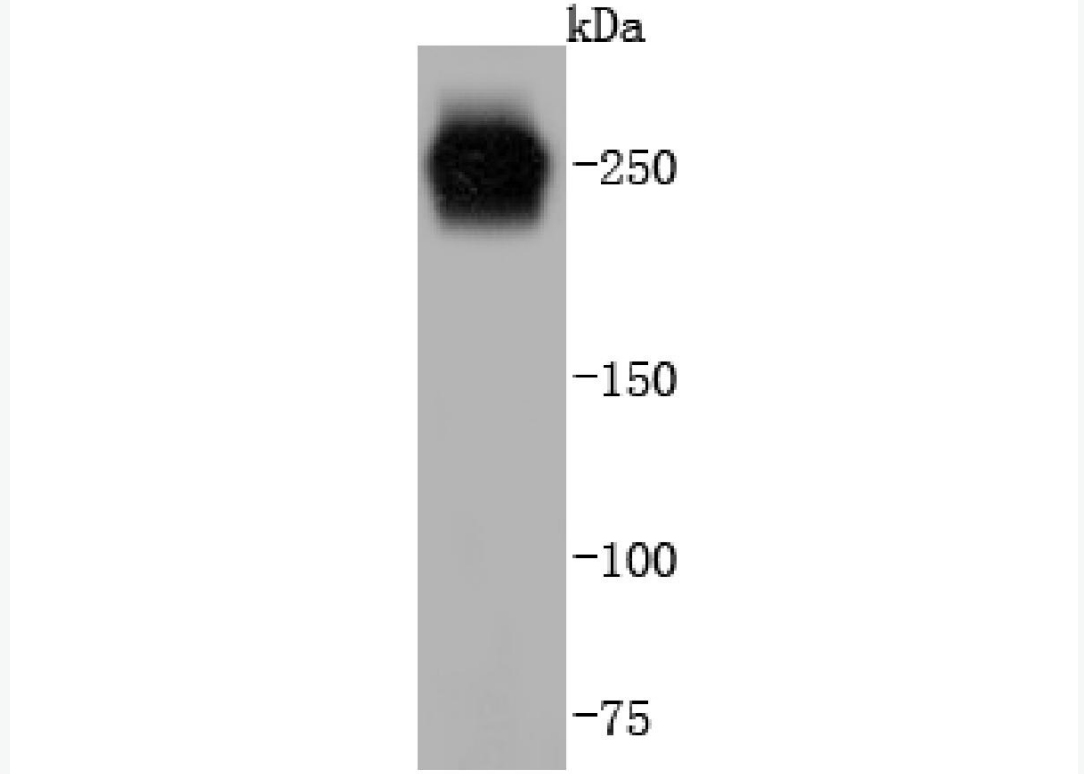
Unigene: 440848 Human

Unigene: 35561 Rat

血管性血友病因子(vWF)是 vascular endothelial cell 和骨髓巨核细胞合成的一种 glycoprotein，在 1 期和 2 期止血中都起着重要作用，如缺乏将导致患者出现血管性血友病(vWD)。vWF 可被 ADAMTS13 裂解以失去活性，血小板反应蛋白/凝血酶敏感蛋白-1(Thrombospondin,TSP-1))可参与了这个调节过程。vWF 水平受多种遗传和环境因素影响，其中 ABO 血型影响较大。vWF 主要通过 A1 和 A3 区与血小板 GP 1b 和胶原结合，在止血和血栓形成过程中起重要作用，并与心、脑血管疾病及血管新生密切相关，因此研究 vWF 的生物学特性和功能具有重要的意义。

**Product Picture**




Western blot analysis of Von Willebrand Factor on human lung lysates with Rabbit anti-Von Willebrand Factor antibody (SLM-52775R) at 1/1000 dilution.

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Lysates/proteins at 20 µg/Lane.

Predicted band size: 309 kDa

Observed band size: 250 kDa

Proteins were transferred to a PVDF membrane and blocked with 5% NFDM/TBST for 1 hour at room temperature. The primary antibody (SLM-52775R) at 1/500 dilution was used in 5% NFDM/TBST at room temperature for 2 hours. Goat Anti-Rabbit IgG - HRP Secondary Antibody (HA1001) at 1:300,000 dilution was used for 1 hour at room temperature.

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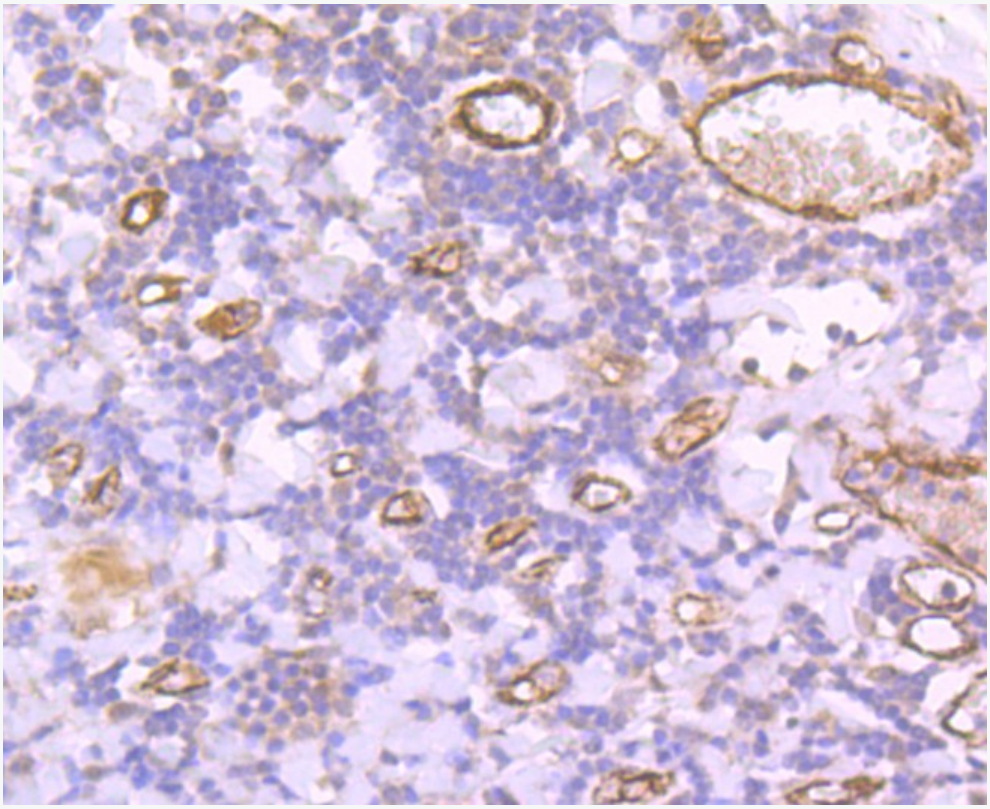

Immunohistochemical analysis of paraffin-embedded human tonsil tissue with Rabbit anti-Von Willebrand Factor antibody (SLM-52775R) at 1/400 dilution. The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH2O and PBS, and then probed with the primary antibody (SLM-52775R) at 1/400 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.