**Product datasheet**

**Anti-Von Willebrand Factor antibody ab6994**

★★★★★ 55 Abreviews  165 References  5 Images

### Overview

**Product name**  
Anti-Von Willebrand Factor antibody

**Description**  
Rabbit polyclonal to Von Willebrand Factor

**Host species**  
Rabbit

**Tested applications**  
Suitable for: ICC/IF, IHC-FrFl, IHC-P, IHC-Fr, WB, Flow Cyt, IHC-FoFr

**Species reactivity**  
Reacts with: Rat, Sheep, Guinea pig, Cow, Dog, Human, Pig  
Does not react with: Chicken

**Immunogen**  
Full length native protein (purified) corresponding to Human Von Willebrand Factor. Purified from plasma.

**Positive control**  
Human tongue.

### Properties

**Form**  
Liquid

**Storage instructions**  
Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.

**Storage buffer**  
pH: 7.4  
Preservative: 0.097% Sodium azide  
Constituent: PBS

**Purity**  
IgG fraction

**Purification notes**  
Whole antiserum is fractionated and then further purified by ion exchange chromatography to provide the IgG fraction of antiserum. This fraction is essentially free of other rabbit serum proteins.

**Clonality**  
Polyclonal

**Isotype**  
IgG

### Applications

Our [Abpromise guarantee](#) covers the use of ab6994 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.
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.

Tissue specificity

Plasma.

Involvement in disease

Defects in VWF are the cause of von Willebrand disease (VWD) [MIM:277480]. VWD defines a group of hemorrhagic disorders in which the von Willebrand factor is either quantitatively or qualitatively abnormal resulting in altered platelet function. Symptoms vary depending on severity and disease type but may include prolonged bleeding time, deficiency of factor VIII and impaired platelet adhesion. Type I von Willebrand disease is the most common form and is characterized by partial quantitative plasmatic deficiency of an otherwise structurally and functionally normal Willebrand factor; type II is associated with a qualitative deficiency and functional anomalies of the Willebrand factor; type III is the most severe form and is characterized by total or near-total absence of Willebrand factor in the plasma and cellular compartments, also leading to a profound deficiency of plasmatic factor VIII.

Sequence similarities

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

Domain

The von Willebrand antigen 2 is required for multimerization of vWF and for its targeting to storage granules.

Post-translational modifications

All cysteine residues are involved in intrachain or interchain disulfide bonds.

N- and O-glycosylated.

**Application** | **Abreviews** | **Notes**
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ICC/IF | ★★★★★ | 1/400.
IHC-FrFli | ★★★★★ | 1/200. (see Abreview)
IHC-P | ★★★★★ | 1/200 - 1/400. for IF and 1/1000-1/2000 for ABC methods with HRP conjugates. Perform enzymatic antigen retrieval with 0.1% pronase for 10 min at 35 °C before commencing with IHC protocol. Indirect Immunofluorescence: minimum working dilution of 1:200 was determined using FFPE sections of human tongue with FITC-conjugated secondary. Indirect Immunoperoxidase Labeling: minimum working dilution of 1:800 was determined using FFPE sections of human tongue with biotinylated secondary and signal amplification.
IHC-Fr | ★★★★★ | 1/1000.
WB | ★★★★★ | 1/500.
Flow Cyt | ★★★★★ | Use a concentration of 5 µg/ml.
            |  | ab171870 - Rabbit polyclonal IgG, is suitable for use as an isotype control with this antibody.
IHC-FoFr | ★★★★★ | Use at an assay dependent concentration. PubMed: 19622235

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**Target**

**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.

**Tissue specificity**

Plasma.

**Involvement in disease**

Defects in VWF are the cause of von Willebrand disease (VWD) [MIM:277480]. VWD defines a group of hemorrhagic disorders in which the von Willebrand factor is either quantitatively or qualitatively abnormal resulting in altered platelet function. Symptoms vary depending on severity and disease type but may include prolonged bleeding time, deficiency of factor VIII and impaired platelet adhesion. Type I von Willebrand disease is the most common form and is characterized by partial quantitative plasmatic deficiency of an otherwise structurally and functionally normal Willebrand factor; type II is associated with a qualitative deficiency and functional anomalies of the Willebrand factor; type III is the most severe form and is characterized by total or near-total absence of Willebrand factor in the plasma and cellular compartments, also leading to a profound deficiency of plasmatic factor VIII.

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**Domain**

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**Post-translational modifications**

All cysteine residues are involved in intrachain or interchain disulfide bonds.

N- and O-glycosylated.
Cellular localization

Secreted. Secreted > extracellular space > extracellular matrix. Localized to storage granules.

Images

Ab6994 positively staining formaldehyde fixed paraffin embedded mouse tumour sections (1/1000). Mice were subcutaneously injected with 3T3 cells which over expressed HER2.

Secondary: Biotin conjugated horse anti rabbit (1/300). Detection was achieved using DAB and the sections were counterstained with Hematoxylin.

This image is courtesy of an Abreview submitted by Jiqiang Zhang on 16 September 2005. For further details relating to the reviewers protocol please refer to the Abreviews section of the data sheet.

Confocal extended focus photograph of von Willebrand factor staining (red) and non-specific nuclear counterstain (green) in the adult mouse olfactory bulb. This picture shows a single major vessel within the glomerular layer of the olfactory bulb. For more details see the review of this antibody by Adam Puche.
Immunocytochemical analysis labeling Von Willebrand Factor with ab6994 at 1/100 dilution.

The nuclear counterstain is DAPI (blue)

ab6994 staining Von Willebrand Factor in pig skin tissue sections by Immunohistochemistry (IHC-P - paraformaldehyde-fixed, paraffin-embedded sections). Tissue was fixed with formaldehyde and blocked with 3% serum for 30 minutes at 20°C; antigen retrieval was enzymatic using pronase, 1mg/ml. Samples were incubated with primary antibody (1/500 in PBS) for 12 hours at 4°C. A Biotin-conjugated goat anti-rabbit IgG polyclonal (1/200) was used as the secondary antibody.

This picture shows Factor VII Immunohistochemical localization in BCG infected Guinea Pig Lung. The image was kindly supplied as part of the review submitted by Elizabeth Chlipala.
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