abcam

Product datasheet

Native Human Von Willebrand Factor protein ab88533

3 References 1 Image

Description

Product name	Native Human Von Willebrand Factor protein
Purity	> 95 % SDS-PAGE.
Expression system	Native
Accession	P04275
Protein length	Full length protein
Animal free	No
Nature	Native
Species	Human
Additional sequence information	Amino acid sequence is not determined.

Specifications

Our **Abpromise guarantee** covers the use of **ab88533** in the following tested applications.

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

Applications	SDS-PAGE
Form	Liquid

Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 6.80 Constituents: 0.735% Sodium citrate, 0.75% Glycine, 0.58% Sodium chloride The percentages are based on the constituent weight/volume

General Info

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

Images

SDS-PAGE - Native Human Von Willebrand Factor protein (ab88533)

4-12% SDS-PAGE analysis of Reduced and Non-reduced samples of ab88533 (1µg).
Molecular weight markers: Myosin (191 kDa), Phosphorylase B (97 kDa), BSA (64 kDa), Glutamic Dehydrogenase (51 kDa), Alcohol Dehydrogenase (39 kDa), Carbonic Anhydrase (28 kDa), Myoglobin Red (19 kDa), Lysozyme (14 kDa)

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