abcam

Product datasheet

Native Human Von Willebrand Factor protein ab88533

3 References 画像数 1

製品の詳細

製品名	Native Human Von Willebrand Factor protein
精製度	> 95 % SDS-PAGE.
発現系	Native
アクセッション番号	**P04275**
タンパク質長	Full length protein
Animal free	No
由来	Native
生物種	Human
配列の追加情報	Amino acid sequence is not determined.

特性

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.

アプリケーション	SDS-PAGE
製品の状態	Liquid

前処理および保存

保存方法および安定性	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

関連情報

機能	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.
組織特異性	Plasma.
関連疾患	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.
配列類似性	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.
ドメイン	The von Willebrand antigen 2 is required for multimerization of vWF and for its targeting to storage granules.
翻訳後修飾	All cysteine residues are involved in intrachain or interchain disulfide bonds. N- and O-glycosylated.
細胞内局在	Secreted. Secreted > extracellular space > extracellular matrix. Localized to storage granules.

画像

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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