abcam

Product datasheet

Native Human Von Willebrand Factor protein ab88533

3 References 1 图像

描述

产品名称	Native人Von Willebrand Factor蛋白
纯度	> 95 % SDS-PAGE.
表达系统	Native
Accession	**P04275**
蛋白长度	Full length protein
无动物成分	No
性质	Native
种属	Human
额外的序列信息	Amino acid sequence is not determined.

技术指标

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.

应用	SDS-PAGE
形式	Liquid

制备和贮存

稳定性和存储	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

常规信息

功能	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.
组织特异性	Plasma.
疾病相关	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.
序列相似性	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.
结构域	The von Willebrand antigen 2 is required for multimerization of vWF and for its targeting to storage granules.
翻译后修饰	All cysteine residues are involved in intrachain or interchain disulfide bonds. N- and O-glycosylated.
细胞定位	Secreted. Secreted > extracellular space > extracellular matrix. Localized to storage granules.

图片

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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