

Anti-human VWF (48.26) VHH



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Properties

Product type:	VHH
Catalog number:	W-001
Clone number:	48.26
Immunogen:	Purified recombinant human von Willebrand Factor (VWF)
Reacts with:	Human von Willebrand factor (VWF)
Tested applications:	ELISA, Flow cytometry
Source:	Monoclonal VHH (<i>Llama glama</i>), purified from HEK293-E 253 cells using affinity chromatography
Clonality:	Monoclonal
Purity:	IMAC purified with Nickel excel Sepharose, >98%
Storage buffer:	PBS
Form:	Liquid
Concentration:	4.4 mg/ml
Storage:	Store at -80°C

Products

Cat. No.	Target	Clone	Form	Applications	Size
W-001	Human VWF	48.26	Purified	ELISA, FC	250 µg

Description

Von Willebrand Factor (VWF) is a multimeric adhesive plasma glycoprotein that is important in the maintenance of hemostasis.¹ It promotes adhesion of platelets to the sites of vessel injury by forming a bridge between subendothelial collagen and the platelet GPIb-IX-V receptor complex. VWF also acts as a chaperone for coagulation factor VIII, by delivering it to the site of injury, stabilizing its heterodimeric structure and protecting it from premature clearance from plasma.^{1,2}

Defects in VWF cause von Willebrand disease (VWD), which defines a group of hemorrhagic disorders in which VWF is either quantitatively or qualitatively abnormal, resulting in altered platelet function (i.e. impaired platelet adhesion), deficiency of factor VIII, and a prolonged bleeding time. Symptoms vary depending on severity and type of disease. Type I VWD is the most common form and is characterized by a partial quantitative deficiency of a structurally and functionally normal VWF; type II VWD is caused by a qualitative deficiency and functional abnormalities of VWF; type III VWD is the most severe form and is associated with a total or near-total absence of VWF in plasma and cells, which also causes the profound deficiency of coagulation factor VIII in plasma.³

Use, storage and stability

Anti-human VWF (48.26) VHH is suitable for use in ELISA and flow cytometry studies. 48.26 recognizes human VWF in solution.

After thawing, 48.26 should be stored in appropriate small aliquots at -20°C or -80°C (stable for at least 6 months) or at 4°C for short time storage.

References

1. Lenting PJ, Christophe OD, Denis CV. von Willebrand factor biosynthesis, secretion, and clearance: connecting the far ends. *Blood*. 2015; 125: 2019-2028. doi: 10.1182/blood-2014-06-528406.
2. De Groot PG, Urbanus RT, Roest M. Platelet interaction with the vessel wall. *Handb Exp Pharmacol*. 2012; (210): 87-110. doi: 10.1007/978-3-642-29423-5_4.
3. Leebeek FWG, Susen S. Von Willebrand disease: Clinical conundrums. *Haemophilia*. 2018; 24 Suppl 6: 37-43. doi: 10.1111/hae.13508.

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