

# Product Sheet



# QVQ

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Note:

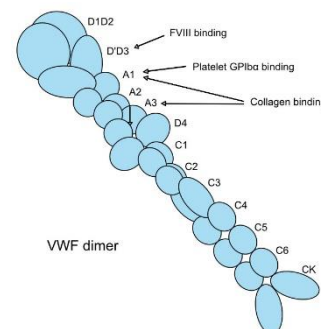
This product has been generated  
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## von Willebrand Factor

**Catalogue no.:** Q118  
**Clone name:** 48.26

**Product:** VHH directed against human von Willebrand Factor (VWF)

**Target:** Von Willebrand Factor (VWF) is a multimeric adhesive plasma glycoprotein that is important in the maintenance of hemostasis.<sup>1</sup> 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.<sup>2</sup> 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.<sup>3</sup> Defects in VWF cause von Willebrand disease (VWD), a common inherited bleeding disorder characterized by excessive mucocutaneous bleeding. 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.<sup>4</sup>



**Source:** Immunization with and phage-display selection on purified recombinant human VWF.  
Recombinant monoclonal VHH (Llama glama), purified from HEK293-E 253 cells using Nickel excel Sepharose affinity chromatography.

**Specificity:** Human VWF.  
Clone 48.26 recognizes human VWF in solution.

**Formulation:** Myc-tagged PBS.

**Mol. Weight:** 14.9 kDa  
**Ext. Coeff. (ε):** 26373 M<sup>-1</sup> cm<sup>-1</sup>  
**A<sub>280</sub> at 1g/L:** 1.8

**Storage:** Shipped on blue ice. Store at 4 °C or -20 °C (aliquots). Addition of 0.02% sodiumazide is optional.

**Applications:** ELISA, flow cytometry (FC)

## References:

- 1 De Meyer et al., (2009) Blood 113(21):5049-57
- 2 Denorme et al., (2019) Front Immunol 10:2884
- 3 Pipe et al., (2016) 128(16): 2007-2016
- 4 James and Goodeve, (2011) Genet Med 13(5):365-76