

HA723431H



Product Type:	Recombinant Rabbit monoclonal IgG, primary antibodies
Species reactivity:	Human
Applications:	ELISA(Det)
Clone number:	PSH09-04

Description: 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. A common hemorrhagic disorder due to defects in von Willebrand factor protein and resulting in impaired platelet aggregation. Von Willebrand disease type 1 is characterized by partial quantitative deficiency of circulating von Willebrand factor, that is otherwise structurally and functionally normal. Clinical manifestations are mucocutaneous bleeding, such as epistaxis and menorrhagia, and prolonged bleeding after surgery or trauma. A hemorrhagic disorder due to defects in von Willebrand factor protein and resulting in altered platelet aggregation. Von Willebrand disease type 2 is characterized by qualitative deficiency and functional anomalies of von Willebrand factor. It is divided in different subtypes including 2A, 2B, 2M and 2N (Normandy variant). The mutant VWF protein in types 2A, 2B and 2M are defective in their platelet-dependent function, whereas the mutant protein in type 2N is defective in its ability to bind factor VIII. Clinical manifestations are mucocutaneous bleeding, such as epistaxis and menorrhagia, and prolonged bleeding after surgery or trauma.

Conjugate:	HRP-conjugated
Immunogen:	Recombinant protein within human von Willebrand factor protein aa 764-1263 (HA210992).
Positive control:	Recombinant human von Willebrand factor protein (HA210992).
Subcellular location:	Secreted.
Database links:	SwissProt: P04275 Human
Recommended Dilutions:	
ELISA(Det)	Use at an assay dependent concentration. Can be paired for Sandwich ELISA with Rabbit monoclonal [PSH09-03] to Human Von Willebrand Factor antibody (Capture) (HA723058) and Recombinant Human Von Willebrand Factor protein (HA210992) as the standard.
Storage Buffer:	PBS (pH7.4), 0.1% BSA, 40% Glycerol.
Storage Instruction:	Shipped at 4°C. Store at +4°C short term (1-2 weeks). It is recommended to aliquot into single-use upon delivery. Store at -20°C long term.
Purity:	Protein A affinity purified.

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Orders:0086-571-88062880

Technical:0086-571-89986345

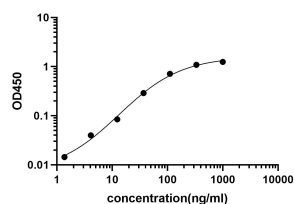
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Images

Fig1: Sandwich ELISA analysis of human Von Willebrand Factor matched pair antibodies

Standard curve of human Von Willebrand Factor matched pair antibodies



Elisa assay was performed by coating wells of a 96-well plate with 100 μ l per well of capture antibody (HA723058) diluted in carbonate/bicarbonate buffer, at a concentration of 2 μ g/ml overnight at 4 $^{\circ}$ C. Wells of the plate were washed, blocked with 150 μ l 0.05% tween-20 1% BSA blocking buffer, and incubated with serial diluted Human Von Willebrand Factor protein (HA210992) starting from 1,000 ng/ml to 0 ng/ml and detect antibody (HA723431H, HRP, 0.5 μ g/ml) for 1 hour at 30 $^{\circ}$ C with shaking. Detection was performed using an Ultra TMB Substrate for 10 minutes at room temperature in the dark. The reaction was stopped with sulfuric acid and absorbances were read on a spectrophotometer at 450 nm.

Note: All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE".

Background References

1. Allen S., Abuzenadah A.M., Hinks J., Blagg J.L., Gursel T., Ingerslev J., Goodeve A.C., Peake I.R., Daly M.E. A novel von Willebrand disease-causing mutation (Arg273Trp) in the von Willebrand factor propeptide that results in defective multimerization and secretion. *Blood* 96:560-568 (2000)
2. Bodo I., Katsumi A., Tuley E.A., Eikenboom J.C., Dong Z., Sadler J.E. Type 1 von Willebrand disease mutation Cys1149Arg causes intracellular retention and degradation of heterodimers: a possible general mechanism for dominant mutations of oligomeric proteins. *Blood* 98:2973-2979 (2001)

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Applications:WB=Western blot IHC-P=Immunohistochemistry (paraffin) IF-Cell=Immunofluorescence (Cell) IF-Tissue=Immunofluorescence (Tissue) FC=Flow cytometry IP=Immunoprecipitation