VWF Plasma – Von Willebrand Factor

PRODUCT OVERVIEW

Product Name	von Willebrand Factor (VWF), Plasma
Full Product Name	von Willebrand Factor Deficient Plasma
Product Gene Name	VWF plasma
Molecular Weight	38,745 Da
OMIM	X04385 mRNA
3D Structure	ModBase 3D Structure for P04275
Product Categories/Family for VWF plasma	Neutralizing Antibodies & Control IgG
Related Product Information for VWF plasma	MBS514025 is a plasma targeting VWF. Also known as von Willebrand Factor Deficient Plasma.
Other Notes	Small volumes of VWF plasma vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for VWF. It may not necessarily be applicable to this product.

NCBI GI #	89191868
NCBI GeneID	7450
NCBI Accession #	NP_000543.2
UniProt Primary Accession #	P04275
UniProt Secondary Accession #	Q8TCE8; Q99806
UniProt Related Accession #	P04275
GenBank Nucleotide #	NM_000552.4
NCBI Official Full Name	von Willebrand factor preproprotein
NCBI Official Synonym Full Names	von Willebrand factor
NCBI Official Symbol	VWF
NCBI Official Synonym Symbols	VWD; F8VWF
UniProt Protein Name	von Willebrand factor
UniProt Synonym Protein Names	von Willebrand antigen 2Alternative name(s):von Willebrand antigen II
Protein Family	von Willebrand factor
UniProt Gene Name	VWF
UniProt Synonym Gene Names	F8VWF; vWF
NCBI Protein Information	von Willebrand factor

NCBI Summary for VWF	This gene encodes a glycoprotein involved in hemostasis. The encoded preproprotein is proteolytically processed following assembly into large multimeric complexes. These complexes function in the adhesion of platelets to sites of vascular injury and the transport of various proteins in the blood. Mutations in this gene result in von Willebrand disease, an inherited bleeding disorder. An unprocessed pseudogene has been found on chromosome 22. [provided by RefSeq, Oct 2015]
UniProt Comments for VWF	VWF: 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. Defects in VWF are the cause of von Willebrand disease type 1 (VWD1). 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. Defects in VWF are the cause of von Willebrand disease type 2 (VWD2). A hemorrhagic disorder due to defects in von Willebrand factor protein and resulting in impaired 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. Defects in VWF are the cause of von Willebrand disease type 3 (VWD3). A severe hemorrhagic disorder due to a total or near total absence of von Willebrand factor in the plasma and cellular compartments, also leading to a profound deficiency of plasmatic factor VIII. Bleeding usually starts in infancy and can include epistaxis, recurrent mucocutaneous bleeding, excessive bleeding after minor trauma, and hemarthroses. Protein type: Cell adhesion; Extracellular matrix; Motility/polarity/chemotaxis; Secreted; Secreted, signal peptide Chromosomal Location of Human Ortholog: 12p13.31 Cellular Component: endoplasmic reticulum; extracellular matrix; extracellular region Molecular Function: chaperone binding; collagen binding; glycoprotein binding; identical protein binding; immunoglobulin binding; integrin binding; protease binding; protein binding; protein homodimerization activity; protein N-terminus binding Biological Process: blood coagulation; blood coagulation, intrinsic pathway; cell adhesion; cell-substrate adhesion; extracellular matrix organization and biogenesis; hemostasis; platelet activation; platelet degranulation; protein homooligomerization; response to wounding Disease: Von Willebrand Disease, Type 1; Von Willebrand Disease, Type 2; Von Willebrand Disease, Type 3
Research Articles on VWF	**1.** von Willebrand factor binds to the surface of dendritic cells and modulates peptide presentation of factor VIII.
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