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ADAMTS13 Rabbit pAb

Catalog # AP98743

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q76LX8
Other Accession	Q76LX8
Reactivity	Human
Predicted	Rat, Pig, Mouse, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	153604
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ADAMTS13
Epitope Specificity	401-500/1427
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 2 CUB domains. Contains 1 disintegrin domain. Contains 1 peptidase M12B domain. Contains 8 TSP type-1 domains.
Post-translational modifications	Glycosylated. O-fucosylated by POFUT2 on a serine or a threonine residue found within the consensus sequence C1-X(2)-(S/T)-C2-G of the TSP type-1 repeat domains where C1 and C2 are the first and second cysteine residue of the repeat, respectively. Fucosylated repeats can then be further glycosylated by the addition of a beta-1,3-glucose residue by the glucosyltransferase, B3GALTL. Fucosylation mediates the efficient secretion of ADAMTS13. May also be C-glycosylated on tryptophan residues within the consensus sequence W-X-X-W of the TPRs, and also N-glycosylated. These other glycosylations can also facilitate secretion. The precursor is processed by a furin endopeptidase which cleaves off the pro-domain.
DISEASE	Defects in ADAMTS13 are the cause of thrombotic thrombocytopenic purpura congenital (TTP) [MIM:274150]; also known as Upshaw-Schulman syndrome (USS). A hematologic disease characterized by hemolytic anemia with fragmentation of erythrocytes, thrombocytopenia, diffuse and non-focal neurologic findings, decreased renal function and fever.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a member of the ADAMTS (a disintegrin and metalloproteinase with thrombospondin motif) protein family. Members of the family share several distinct protein modules, including a propeptide region, a metalloproteinase domain, a disintegrin-like domain, and a thrombospondin type 1 (TS) motif. Individual members of this family differ in the number of C-terminal TS motifs, and some have unique C-terminal domains. The enzyme encoded by this gene is the von Willebrand Factor (vWF)-cleaving protease, which is responsible for cleaving at the site of Tyr842-Met843 of the vWF molecule. A deficiency of this enzyme is associated with thrombotic thrombocytopenic purpura. Alternative splicing of this gene generates multiple transcript variants encoding different isoforms. [provided by RefSeq, Nov 2008].

Additional Information

Gene ID	11093
Other Names	ADAM-TS13; ADAMTS-13; C9orf8; VWFCP; vWF-CP; Gm710; ATS13_HUMAN; ADAMTS13; ADAM-TS 13; von Willebrand factor-cleaving protease (vWF-CP | vWF-cleaving protease); 3.4.24.87; ATS13_MOUSE; ADAM metallopeptidase with thrombospondin type 1 motif 13; a disintegrin-like and metalloprotease (reprolysin type) with thrombospondin type 1 motif, 13
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	ADAMTS13
Synonyms	C9orf8
Function	Cleaves the vWF multimers in plasma into smaller forms thereby controlling vWF-mediated platelet thrombus formation.
Cellular Location	Secreted. Note=Secretion enhanced by O-fucosylation of TSP type-1 repeats
Tissue Location	Plasma. Expressed primarily in liver.

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