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Rabbit Anti-ADAMTS13 antibody

SL5856R

Product Name:	ADAMTS13
Chinese Name:	整合素样金属蛋白酶与凝血酶13型抗体
Alias:	Cleaves the vWF multimers in plasma into smaller forms. Von Willebrand factor cleaving protease; A disintegrin and metalloproteinase with thrombospondin motifs 13; A disintegrin like and metalloprotease (reprolysin type) with thrombospondin type 1 motif 13; A disintegrin like and metalloprotease with thrombospondin type 1 motif 13; ADAM metallopeptidase with thrombospondin type 1 motif 13; ADAM TS 13; ADAM TS13; ADAM-TS 13; ADAM-TS13; ADAMTS 13; ADAMTS-13; ADAMTS13; ADAMTS13 protein; ATS13_HUMAN; C9orf8; TTP; von Willebrand factor-cleaving protease; vWF cleaving protease; vWF CP; vWF-cleaving protease; vWF-CP; vWFCP.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	145kDa
Cellular localization:	Secretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ADAMTS13:401-500/1427
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed

Product Detail:	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]. **Function:** Cleaves the vWF multimers in plasma into smaller forms. **Subcellular Location:** Secreted. Note=Secretion enhanced by O-fucosylation of TSP type-1 repeats. **Tissue Specificity:** Plasma. Expressed primarily in liver. **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. **Similarity:** Contains 2 CUB domains. Contains 1 disintegrin domain. Contains 1 peptidase M12B domain. Contains 8 TSP type-1 domains. **SWISS:** Q76LX8

Gene ID:
11093

Database links:

Entrez Gene: 11093Human

Entrez Gene: 279028Mouse

Entrez Gene: 362091Rat

Omim: 604134Human

SwissProt: Q76LX8Human

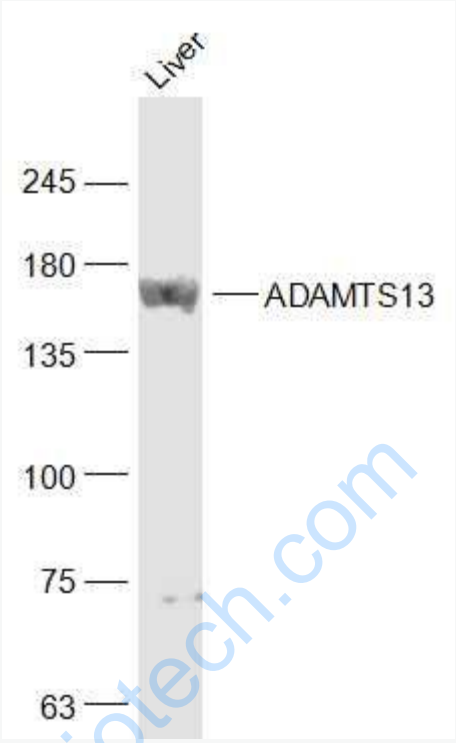
SwissProt: Q769J6Mouse

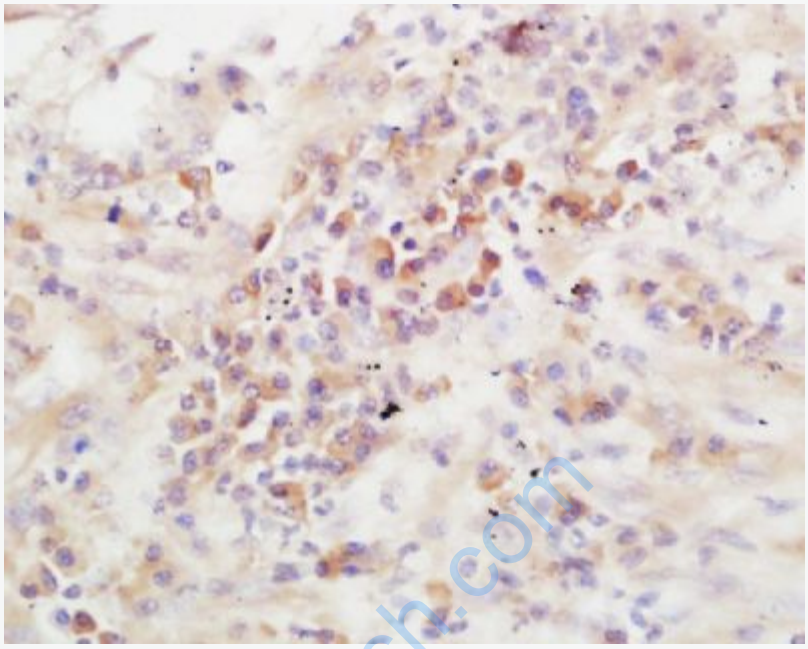
Unigene: 131433Human

Unigene: 330084Mouse

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Extracellular matrix蛋白

Picture:	 Sample: Liver (Mouse) Lysate at 40 ug Primary: Anti-ADAMTS13 (SL5856R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 145 kD Observed band size: 155 kD



Tissue/cell: human lung carcinoma; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-ADAMTS13 Polyclonal Antibody, Unconjugated(SL5856R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining