



ATS13 Polyclonal Antibody

Catalog No	YP-Ab-05287
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB;ELISA
Gene Name	ADAMTS13 C9orf8 UNQ6102/PRO20085
Immunogen	Synthesized peptide derived from human protein . at AA range: 940-1020
Specificity	ATS13 Polyclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	ATS13;ADAMTS13 C9orf8 UNQ6102/PRO20085;A disintegrin and metalloproteinase with thrombospondin motifs 13 (ADAM-TS 13) (ADAM-TS13) (ADAMTS-13) (EC 3.4.24.87) (von Willebrand factor-cleaving protease) (vWF-CP) (vWF-cleaving protease)
Observed Band	156kD
Calculated Molecular Weight	156kD
Cell Pathway	Secreted . Secretion enhanced by O-fucosylation of TSP type-1 repeats.
Tissue Specificity	Plasma. Expressed primarily in liver.
Function	catalytic activity: Cleaves the vWF at the 842-Tyr- -Met-843 in the A2 domain of the vWF subunit., cofactor: Binds 1 zinc ion per subunit., cofactor: Binds 4 calcium ions ., disease: Defects in ADAMTS13 are the cause of congenital thrombotic thrombocytopenic purpura (TTP) [MIM:274150]; also known as Upshaw-Schulman syndrome (USS). Congenital TTP is a life-threatening systemic disorder due to constitutional deficiency of vWF-cleaving protease. Typical features are hemolytic anemia with fragmentation of erythrocytes, thrombocytopenia, diffuse and nonfocal neurologic findings, decreased renal function and fever. Congenital TTP is characterized by neonatal onset, response to fresh plasma infusion and frequent relapses. Inheritance pattern is autosomal



recessive. In sporadic cases, TTP is associated with deficiency of vWF-cleaving protease due to the presence of inhibiting autoantibodies (acquired T

Background

This gene encodes a member of a family of proteins containing several distinct regions, including a metalloproteinase domain, a disintegrin-like domain, and a thrombospondin type 1 (TS) motif. The enzyme encoded by this gene specifically cleaves von Willebrand Factor (vWF). Defects in this gene are associated with thrombotic thrombocytopenic purpura. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jul 2013],

matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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