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Product datasheet for AP55393SU-N

ADAMTS13 Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	ELISA, IHC, WB
Recommended Dilution:	**ELISA.** **Western Blot:** 1/500-1/2000. **Immunohistochemistry:** 1/50-1/500.
Reactivity:	Human, Mouse
Host:	Rabbit
Clonality:	Polyclonal
Immunogen:	Synthetic peptide derived from N-terminal domain of ADAM-TS13 protein
Specificity:	Reacts specifically with Human 153 kDa ADAMTS13 protein. Cross reacts with Mouse protein.
Formulation:	State: Serum State: Lyophilized Serum Preservative: None
Reconstitution Method:	Restore in distilled water to initial volume.
Conjugation:	Unconjugated
Storage:	Store lyophilized at 2-8°C for 6 months or at -20°C long term. After reconstitution store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C long term. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Predicted Protein Size:	154 kDa
Gene Name:	ADAM metallopeptidase with thrombospondin type 1 motif 13
Database Link:	Entrez Gene 11093 Human Q76LX8

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Background:

ADAMTS-13 is produced by hepatic stellate cells and in smaller amounts by human endothelial cells, and is present in plasma at a concentration of approximately 1 µg/ml. ADAMTS-13 is a zinc-containing metalloprotease belonging to the ADAMTS family characterized by a protease domain, an adjacent disintegrin-like domain, one or more thrombospondin type 1 repeats, a cystein-rich domain and a typical spacer region. ADAMTS-13 is composed of a series of domains (amino to carboxy terminal): metalloprotease, disintegrin-like, central thrombospondin-1 (TSP-1), cysteine-rich, spacer, seven additional TSP-1 domains and two unique CUB domains. ADAMTS-13 has no hydrophobic transmembrane domain, and hence it is not anchored in the cell membrane. The apparent molecular weight is 170 or 190 kDa on non-reducing or reducing SDS-PAGE, respectively.
ADAMTS-13 has an important function in haemostasis, where it catalyzes the cleavage of the peptide bond between tyrosine-1605 and methionine-1606 in the A2 domain of von Willebrand Factor (VWF), resulting in 2 electrophoretic reduced fragments of 176 and 140 kDa, respectively. This process renders large multimers less adhesive and hence less reactive in the setting of thrombus formation. ADAMTS-13 is therefore said to be a natural anti-thrombotic agent.
Severe ADAMTS-13 deficiency is associated with systemic microvascular thrombosis in familial or acquired thrombotic thrombocytopenic purpura (TTP). The accumulation of non-cleaved large VWF multimers causes spontaneous systemic platelet aggregation blocking oxygen supply to vital organs. This life-threatening disorder can lead to ischemic disease with (multiple) organ failure.

Synonyms:

ADAMTS-13, ADAM-TS13, C9orf8, vWF-cleaving protease