OriGene Technologies, Inc.
9620 Medical Center Drive, Ste 200
Rockville, MD 20850, US
Phone: +1-888-267-4436
https://www.origene.com
techsupport@origene.com
EU: info-de@origene.com
CN: techsupport@origene.cn

Product datasheet for AM26194PU-N

ADAMTS13 (686-894) Mouse Monoclonal Antibody [Clone ID: 20A5]

Product data:

Product Type:	Primary Antibodies
Clone Name:	20A5
Applications:	ELISA, IF, WB
Recommended Dilution:	**Immunoassay.** **Immunoflourescence:** The typical starting working dilution is 1/50. **Western blot:** The typical starting working dilution is 1/50.
Reactivity:	Human
Host:	Mouse
Isotype:	IgG1
Clonality:	Monoclonal
Specificity:	The monoclonal antibody 20A5 recognizes human ADAMTS-13, A Disintegrin And Metalloprotease with ThromboSpondin type 1 domain 13. It recognizes the central to C-terminal TSP-1 repeats 2 to 5 of ADAMTS-13 (amino acid 686-894).
Formulation:	PBS State: Purified State: Liquid 0.2 µm filtered Ig fraction Stabilizer: 0.1% BSA Preservative: 0.02% sodium azide
Concentration:	lot specific
Purification:	Protein G Chromatography
Conjugation:	Unconjugated
Storage:	Store undiluted at 2-8°C.
Stability:	Shelf life: one year from despatch.
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