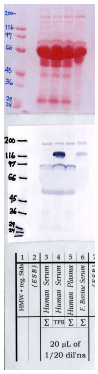
Anti-ADAMTS13 Rabbit Polyclonal Antibody

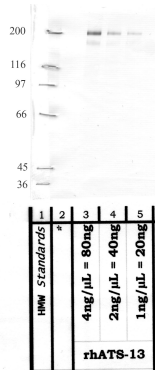
A disintegrin and metalloproteinase with thrombospondin motifs 13, ADAM-TS 13, ADAM-TS13, ADAMTS-13, EC 3.4.24.87, von Willebrand factor-cleaving protease, vWF-CP, vWF-cleaving protease,

Gene: ADAMTS13 **UniProt:** Q76LX8 **Host:** Rabbit **Clonality:** Polyclonal (pAb)

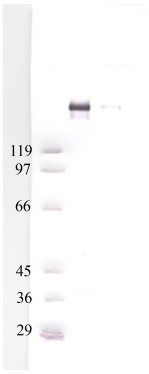
Product Image



propeptide
RP1ADAMTS13



Propeptide domain
RP1ADAMTS13



Carboxyterminal end long form
RP3ADAMTS13

— Western Blot (4-panel validation across domain-specific antibody clones)

Actual Western blot validation for anti-ADAMTS13 rabbit polyclonal antibody. 4-panel validation across domain-specific antibody clones — each panel labeled with its catalog number and target domain. Reducing conditions; 10–20 µg/lane on 7.5–10% SDS-PAGE.

Key Product Facts

Supplier	Triple Point Biologics, Inc. (Sudbury, Ontario, Canada)
Product Name	Anti-ADAMTS13 Rabbit Polyclonal Antibody
Gene Symbol	ADAMTS13
Alternative Names	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,
UniProt ID	Q76LX8
Host Species	Rabbit
Clonality	Polyclonal (pAb)
Isotype	IgG
Concentration	1mg/ml
Purity	Affinity purified
Pack Size	100ug
Validated Application	WB
Recommended Dilution	WB-1/1000
Species Reactivity	Validated- Human Potential-
Immunogen Type	Synthetic Peptide from Uniprot Sequence
Conjugation	none
Buffer / Preservatives	Sodium Chloride 500 mM, Sodium Phosphate 10 mM, Potassium Phosphate 1.8 mM, Potassium Chloride 2.7 mM, Glycerol 50% (v/v)
Website	triplepointbiologics.com/anti-adamts-13-rabbit-polyclonal-antibody

Available Domain-Specific Variants

Triple Point Biologics produces antibodies to defined structural domains of ADAMTS13. Each variant is directed at a specific structural region of the protein. Exact peptide sequences are proprietary; each variant is validated for the applications listed.

Domain / Variant	Epitope Region (approx. residues)	Cat. No.	Availability

Propeptide domain	**30–74** Region within the propeptide of human ADAMTS13, located between the signal peptide cleavage site and the furin-recognition motif that precedes the mature metalloproteinase domain; this segment is cleaved intracellularly and is not present in the circulating active enzyme	RP1ADAMTS13	In catalog
Metalloproteinase domain	**80–286** Region within the catalytic metalloproteinase domain of human ADAMTS13, which harbours the conserved HEXXHXXGXXH zinc-binding motif responsible for peptide bond hydrolysis and constitutes the core proteolytic unit of the enzyme	RP2ADAMTS13	In catalog
Carboxyterminal end long form	**1377–1427** Region spanning the carboxy-terminal CUB domains (CUB1 and CUB2) of human ADAMTS13, which are unique among ADAMTS family members and mediate autoinhibitory interactions as well as binding to the D4 and CK regions of vWF, contributing to substrate recognition under flow conditions	RP3ADAMTS13	In catalog
—	**75–1427** Antigen spanning multiple structural domains of human ADAMTS13, encompassing regions from the propeptide through the carboxy-terminal CUB domains, providing broad epitope coverage across the full-length precursor protein architecture	RP-ADAMTS13	In catalog
Metalloproteinase domain	Metalloproteinase domain	RP4ADAMTS13	In catalog

Applications & Recommended Dilutions

App.	Species	Dilution Range	Notes
WB	Validated- Human Potential-	1:1000	Optimal dilution should be determined per laboratory and lot.

Optimal dilutions should be determined by each laboratory for each application. Triple Point Biologics antibodies include a peptide-neutralized negative control aliquot for specific-signal confirmation.

Storage & Handling

Short-term Storage	+4°C (1–2 weeks)
Long-term Storage	Aliquot and store at -20°C

Shipping Conditions	Shipped in styrofoam cooler on blue ice
Hazard Warning	Product may contain Sodium Azide — POISONOUS AND HAZARDOUS. Handle by trained personnel only.
Shelf Life	12 months from date of receipt under recommended conditions

Target Background

Biology & Family

ADAMTS13 (A Disintegrin And Metalloproteinase with ThromboSpondin motifs 13, EC 3.4.24.87) is a member of the ADAMTS family of secreted, multidomain zinc-dependent metalloproteinases. Among the 19 human ADAMTS family members, ADAMTS13 is unique in its dedicated physiological role: it is the sole enzyme responsible for proteolytic regulation of von Willebrand factor (vWF) multimer size in the circulation. Its domain architecture—signal peptide, propeptide, metalloproteinase domain, disintegrin-like domain, thrombospondin type-1 repeat (TSP1), cysteine-rich domain, spacer domain, additional TSP1 repeats, and two carboxy-terminal CUB domains—reflects a highly evolved substrate-recognition apparatus.

Expression & Localisation

ADAMTS13 is synthesized predominantly by hepatic stellate cells, which are considered the principal source of circulating plasma ADAMTS13. Hepatocytes, sinusoidal endothelial cells, and megakaryocytes also contribute to production, and the protein has been detected in platelet α-granules, suggesting a local hemostatic role at sites of vascular injury. Endothelial cells lining the microvasculature express ADAMTS13 and may provide a locally acting pool of the protease. The mature enzyme circulates in plasma at a concentration of approximately 1 μg/mL as a constitutively active, single-chain glycoprotein.

Activation Mechanism & Substrate Recognition

Unlike many metalloproteinases that are secreted as latent zymogens requiring extracellular activation, ADAMTS13 is processed intracellularly—its propeptide is cleaved in the trans-Golgi network by furin-like proprotein convertases—and is secreted in a constitutively active form. Cleavage of the Tyr1605–Met1606 bond within the vWF A2 domain is the canonical catalytic event. Substrate recognition is exosite-dependent: the spacer and disintegrin-like domains engage the vWF A2 domain, while the carboxy-terminal TSP1 repeats and CUB domains mediate interactions with the D4 and CK regions of vWF, enabling cleavage under high-shear conditions that unfold the cryptic A2 site.

Key Substrates & Inhibitor Profile

The primary physiological substrate of ADAMTS13 is vWF, specifically the Tyr1605–Met1606 scissile bond in the mechanically unfolded A2 domain. Additional reported substrates include COMP (cartilage oligomeric matrix protein) and osteopontin, though their in vivo relevance remains under investigation. ADAMTS13 is not regulated by canonical tissue inhibitors of metalloproteinases (TIMPs). Its activity is modulated by PCSK6 (furin) during activation, and is inhibited in plasma by autoantibodies—particularly IgG directed against the spacer domain—as well as by a2-macroglobulin under certain conditions. Platelet factor 4 (PF4) and certain acute-phase proteins have also been reported to attenuate ADAMTS13 activity.

Disease Relevance

Severe deficiency of ADAMTS13 activity (typically <10% of normal) leads to accumulation of ultra-large vWF multimers and is the molecular basis of thrombotic thrombocytopenic purpura (TTP). Congenital TTP (Upshaw-Schulman syndrome) results from biallelic loss-of-function mutations in the ADAMTS13 gene. Acquired TTP, the more common form, is caused by IgG autoantibodies that inhibit protease activity or accelerate its clearance. Beyond TTP, reduced ADAMTS13 activity has been associated with ischemic stroke, myocardial infarction, pre-eclampsia, and sepsis-associated coagulopathy, making quantification of ADAMTS13 antigen and activity an important diagnostic and research tool.

Research Applications

Anti-ADAMTS13 antibodies are employed to quantify protease antigen levels in patient plasma by ELISA and Western blot, to localise ADAMTS13 in tissue sections by immunohistochemistry, and to characterise the epitope landscape targeted by autoantibodies in acquired TTP. Antibodies raised against discrete structural domains—propeptide, metalloproteinase domain, spacer, or carboxy-terminal regions—are particularly useful for mapping inhibitory autoantibody binding sites and for studying domain-specific interactions with vWF. Immunofluorescence applications have been used to study ADAMTS13 localisation within hepatic stellate cells, sinusoids, and endothelial compartments.

Gene & Protein Reference

Gene Symbol	ADAMTS13
HGNC	HGNC:1366
NCBI Gene ID (Human)	11093
NCBI Gene ID (Mouse)	72821
Ensembl	ENSG00000160323
OMIM	604134
UniProt (Human)	Q76LX8
UniProt (Mouse)	Q7TSN7
Protein Family	ADAMTS family; peptidase M12B subfamily
Predicted MW	~145 kDa (unglycosylated); ~190 kDa with glycosylation
Observed MW	~190 kDa on SDS-PAGE (mature secreted form)
Chromosome	9q34.2
Paralogue	ADAMTS1, ADAMTS4, ADAMTS5 (broader ADAMTS family); no close functional paralogue for vWF cleavage
Key Substrates	von Willebrand factor (Tyr1605–Met1606 bond, vWF A2 domain); COMP; osteopontin
Activated by	Furin/PCSK6 (intracellular propeptide cleavage); shear stress-induced unfolding of vWF A2 domain exposes scissile bond
Inhibited by	Autoimmune IgG (spacer and other domain epitopes); α2-macroglobulin; platelet factor 4 (PF4); acute-phase reactants

About Triple Point Biologics

With over 30 years dedicated to antibody production, Triple Point Biologics is a Canadian company and the premier resource for proteinase and proteinase inhibitor research. Products have been cited in thousands of peer-reviewed publications.

The domain-specific antibody strategy — available in standard, ATK, SPA, and GSK kit formats — delivers predefined epitopes, built-in peptide-neutralized negative controls, and superior specificity for Western blot.

Website: www.triplepointbiologics.com