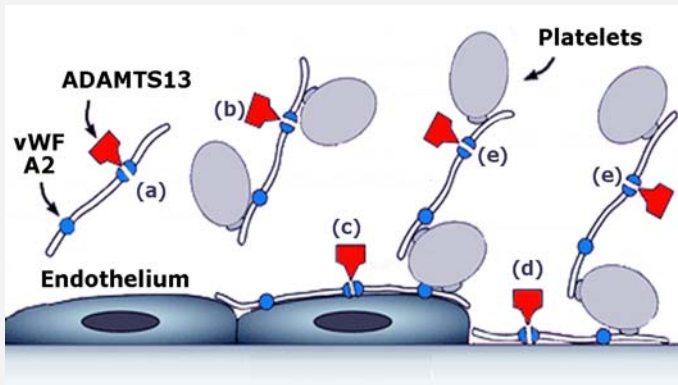


ADAMTS13 and Thrombotic Thrombocytopenic Purpura



ADAMTS13 (A Disintegrin-like And Metalloprotease with ThromboSpondin-1 repeats), also called von Willebrand Factor cleaving protease, is a zinc metalloprotease that cleaves von Willebrand Factor (vWF) multimers.



VWF is released from endothelial cells as unusually large multimers that may diffuse into the circulation (a and b) or adhere to the endothelial cell surface (c). VWF also binds to connective tissue exposed at sites of vascular injury (d). Under conditions of high fluid shear stress, platelets can adhere to VWF in solution (b) or on surfaces (c and d). VWF also may recruit platelets to previously adhering platelets (e). ADAMTS13 cleaves a Tyr-Met bond in the A2 domain of the VWF subunit, severing the multimer. This reaction is slow for VWF in solution (a) but occurs rapidly when platelets adhere to VWF under high fluid shear conditions in suspension (b) or on surfaces (c-e). Failure of this mechanism appears to cause Thrombotic Thrombocytopenic Purpura (TTP).

Thrombotic Thrombocytopenic Purpura (TTP) is a life-threatening disorder characterized by formation of thrombi in micro vessels, red cell fragmentation, CNS, and renal complications. The accumulation of vWF multimers seen in TTP is strongly associated with low ADAMTS13 activity which may be caused by:

- 1) genetic alterations in the ADAMTS13 gene leading to non-functional ADAMTS13 (congenital TTP) or
- 2) development of blocking auto-antibodies against ADAMTS13 (acquired TTP). The induction of ADAMTS13 auto-antibodies in some cases has been linked to drug therapy (e.g., clopidogrel, ticlopidine, quinine).

However, the biological role ADAMTS13 plays in the etiology of TTP is not fully understood, as clinical studies have shown that there is not a strict correlation between ADAMTS13 activity and the clinical expression of TTP symptoms. Many patients with TTP have normal levels of ADAMTS13 activity. Additionally, patients with low levels of ADAMTS13 activity do not exhibit clinical manifestations of TTP. Research at ADI has also found that ADAMTS13 is complexed with Factor XI in plasma. The measurement of ADAMTS13 antigen, auto-antibodies, and ADAMTS13/FXI complexes should provide a greater understanding of the role ADAMTS13 plays in TTP, vWF processing, and hemostasis.

ADAMTS©280313

Complete Product Portfolio on ADAMTS13over

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ADAMTS13

Assays and Reagents



ASSAYS	Assay Type	Format	Product #
IMUBIND® ADAMTS13 kit	ELISA	96 tests	813
IMUBIND® ADAMTS13 Autoantibody kit	ELISA	96 tests	814
ACTIFLUOR™ ADAMTS13 Activity kit	Fluorometric assay	48 tests	812
ANTIBODIES	Known applications	Amount	Product #
anti-human ADAMTS13 mAb	ELISA	250 µg	ADG3305
anti-human ADAMTS13 mAb	ELISA, Western blot, Immuno-precipitation, functional inhibition	250 µg	ADG3312

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ACTIFLUOR™ ADAMTS13

Activity assay, # 812

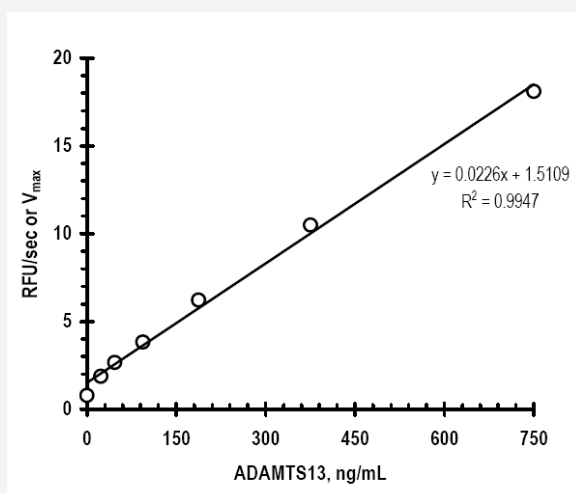


Principle of the assay

ACTIFLUOR ADAMTS13 is a FRET assay that measures the amount of ADAMTS13 activity in human plasma. A citrated plasma sample is assayed for ADAMTS13 protease activity using recombinant VWF86-ALEXA FRET substrate. Proteolytic cleavage of the VWF86-ALEXA FRET substrate by ADAMTS13 uncouples the ALEXA fluorochromes resulting in an increase in fluorescence. The increase in fluorescence over time (V_{max}) due to cleavage of the substrate by ADAMTS13 is monitored at 37°C using a spectrofluorometer (Ex=485 nm; Em=535 nm).

A standard curve is constructed using normal plasma with a known concentration of ADAMTS13. The ADAMTS13 activity in the plasma is determined by interpolation of the V_{max} values from the standard curve.

Standard curve



Related products

- IMUBIND® ADAMTS13 ELISA
- IMUBIND® ADAMTS13 Autoantibody ELISA
- anti-ADAMTS13 monoclonal antibodies

Key Features

- **Format:** 48-well format
- **Assay Time:** 20 minutes
- **Sample Type:** citrated human plasma
- **Sample Volume:** 20 μ L to perform a duplicate analysis
- **Assay Range:** 0 - 750 ng/ml
- **Sensitivity:** 3 ng/ml (< 5% normal)
- **Values:** results expressed as ADAMTS13 conc. [ng/ml]
- **Precision:** intra-assay CV = 4.1 %, inter-assay CV= 4.4 %

REFER TO APPROPRIATE PACKAGE INSERT FOR ADDITIONAL TECHNICAL INFORMATION