ADAMTS13

Thrombo-Guide
vom Labor zur Diagnose

Description and Clinical Significance:

ADAMTS13 is an enzyme that physiologically cleaves von Willebrand factor (VWF). Depending on blood flow shear forces, ADAMTS13 binds to VWF and cleaves it at a highly specific site. This regulates the size of VWF multimers and generates the normal VWF multimer pattern. ADAMTS13 is synthesized in the liver.
A severe ADAMTS13 deficiency leads to the persistence of ultra-large VWF multimers in the circulation. Depending on blood-flow shear forces, platelets increasingly bind to these multimers, resulting in the formation of VWF-platelet-rich microthrombi that impair the function of vulnerable organs. This clinical picture, thrombotic thrombocytopenic purpura (TTP), is life-threatening and requires immediate therapy. For details, see the TTP information text.

- ADAMTS13 activity is determined from citrated plasma using an ELISA or automated with a FRETS-VWF73-based method.
- Based on the Bethesda method (serial dilutions), the presence of ADAMTS13-inhibiting substances in the blood can be detected and quantified.
- Immunologically, ADAMTS13-binding (IgG) antibodies can be detected and quantified.
- Genetic analyses (ADAMTS13 gene sequencing) are the standard method for detecting congenital ADAMTS13 deficiency.

Reference Ranges:

ADAMTS13-Activity:	40-120%
ADAMTS13-Inhibitor:	0 BU/L
Anti-ADAMTS13 IgG:	<12 U/mL

Elevated Values:

Of no clinical relevance

Decreased Values:

Slightly decreased ADAMTS13 values often occur during acute-phase reactions (excess of VWF – reduction of ADAMTS13) and are of no clinical significance. Only a severe ADAMTS13 deficiency (<10%) leads to TTP and requires further investigation.

Pre-analytics:

ADAMTS13 is determined from citrated plasma. Attention must be paid to precise blood collection, avoidance of contamination, correct filling of the blood collection tube, and thorough mixing with the citrate. EDTA inhibits ADAMTS13; therefore, EDTA blood is not suitable for analysis.

References:

Thomas L, Labor und Diagnose, 2023, Release 5: https://www.labor-und-diagnose.de/index.html
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