Test Definition: AADAM

ADAMTS13 Profile Interpretation

Overview

Useful For

Technical interpretation of a profile to assist with the diagnosis of congenital, immune, or acquired thrombotic thrombocytopenic purpura including inhibitor studies

Method Name

Only orderable as a reflex. For more information see ADAMP / ADAMTS13 Activity with Reflex Inhibitor Profile, Plasma

Technical Interpretation

NY State Available

Yes

Specimen

Specimen Type

Plasma Na Cit

Specimen Required

Only orderable as a reflex. For more information see ADAMP / ADAMTS13 Activity with Reflex Inhibitor Profile, Plasma

Reject Due To

All specimens will be evaluated at Mayo Clinic Laboratories for test suitability.

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Plasma Na Cit	Frozen		

Clinical & Interpretive

Clinical Information

Thrombotic thrombocytopenic purpura (TTP), a rare (estimated incidence of 3.7 cases per million) and potentially fatal thrombotic microangiopathy (TMA) syndrome, is characterized by a pentad of symptoms: thrombocytopenia, microangiopathic hemolytic anemia (intravascular hemolysis and presence of peripheral blood schistocytes), neurological symptoms, fever, and kidney dysfunction. A large majority of patients initially present with thrombocytopenia and peripheral blood evidence of microangiopathy, and in the absence of any other potential explanation for such findings, satisfy criteria for early initiation of plasma exchange, which is critical for patient survival.

TTP may rarely be congenital (Upshaw-Shulman syndrome) but, far more commonly, is acquired. Acquired TTP may be considered primary or idiopathic (the most frequent type) or associated with distinctive clinical conditions (secondary TTP) such as medications, hematopoietic stem cell or solid organ transplantation, sepsis, and malignancy.

The isolation and characterization of an IgG autoantibody frequently found in patients with idiopathic TTP, clarified the basis of this entity and led to the isolation and characterization of a metalloprotease called ADAMTS13 (a disintegrin and metalloprotease with thrombospondin type 1 motif 13 repeats), which is the target for the IgG autoantibody, leading to a functional deficiency of ADAMTS13. ADAMTS13 cleaves the ultra-high-molecular-weight multimers of von Willebrand factor (VWF) at the peptide bond Tyr1605-Met1606 to disrupt VWF-induced platelet aggregation. The IgG antibody prevents this cleavage and leads to TTP. Although the diagnosis of TTP may be confirmed with ADAMTS13 activity and inhibition studies, the decision to initiate plasma exchange should not be delayed pending results of this assay.

ADAMTS13 and inhibitor Bethesda titer results can have an impact on overall survival, ultimate clinical outcome, responsiveness to plasma exchange, and relapse are still controversial in recent literature. Therefore, clinical correlation is essential.

Reference Values

Only orderable as a reflex. For more information see ADAMP / ADAMTS13 Activity with Reflex Inhibitor Profile, Plasma

ADAMTS13 ACTIVITY ASSAY
> or =70%
ADMTS13 Inhibitor Titer
<0.5 BU

Interpretation

Interpretive comments are provided.

Less than 10% ADAMTS13 activity is highly indicative of thrombotic thrombocytopenic purpura (TTP) in an appropriate clinical setting. The presence of ADAMTS-13 measurable Bethesda titer is most consistent with an acquired (autoimmune) TTP.

Cautions

No significant cautionary statements

Clinical Reference

1. Sadler JE. Von Willebrand factor, ADAMTS13, and thrombotic thrombocytopenic purpura. Blood. 2008;112(1):11-18. doi:10.1182/blood-2008-02-078170
2. George JN. How I treat patients with thrombotic thrombocytopenic purpura: 2010. Blood. 2010;116(20):4060-4069. doi:10.1182/blood-2010-07-271445
3. Upshaw JD. Congenital deficiency of a factor in normal plasma that reverses microangiopathic hemolysis and thrombocytopenia. N Engl J Med. 1978;298(24):1350-1352. doi:10.1056/NEJM197806152982407
4. Chiasakul T, Cuker A. Clinical and laboratory diagnosis of TTP: an integrated approach. Hematology Am Soc Hematol Educ Program. 2018;2018(1):530-538. doi:10.1182/asheducation-2018.1.530
5. Mackie I, Mancini I, Muia J, et al. International Council for Standardization in Haematology (ICSH) recommendations for laboratory measurement of ADAMTS13. Int J Lab Hematol. 2020;42(6):685-696. doi:10.1111/ijlh.13295

Performance

Method Description
Computer-generated interpretive comments will be provided based on the profile results.

PDF Report
No

Day(s) Performed
Monday through Sunday

Report Available
3 to 5 days

Specimen Retention Time
14 days

Performing Laboratory Location
Mayo Clinic Laboratories - Rochester Main Campus

Fees & Codes

Fees
- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification
Not Applicable

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
AADAM	ADAMTS13 Profile Interpretation	No LOINC Needed

Result ID	Test Result Name	Result LOINC® Value
620818	ADAMTS13 Profile Interpretation	69049-5
620820	ADAMTS13 Profile Interpretation	No LOINC Needed