Overview

Useful For
Assisting with the diagnosis of congenital or acquired thrombotic thrombocytopenic purpura

Method Name
Only orderable as part of a profile. For more information see ADM13 / ADAMTS13 Activity and Inhibitor Profile.

NY State Available
Yes

Specimen

Specimen Type
Plasma Na Cit

Specimen Required
Only orderable as part of a profile. For more information see ADM13 / ADAMTS13 Activity and Inhibitor Profile.

Specimen Minimum Volume
2 mL

Reject Due To

<table>
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<th>Gross hemolysis</th>
<th>Reject</th>
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<td>Gross lipemia</td>
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<tr>
<td>Gross icterus</td>
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Specimen Stability Information

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Clinical and 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 renal dysfunction. The 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 to be 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.

**Reference Values**

Negative

Only orderable as part of a profile. For more information see ADM13 / ADAMTS13 Activity and Inhibitor Profile.

**Interpretation**

Less than 10% ADAMTS13 activity is highly indicative of thrombotic thrombocytopenic purpura (TTP) in an appropriate clinical setting. The presence of ADAMTS13 inhibition (positive inhibitor screen) with a measurable antibody titer is most consistent with an acquired TTP.

**Cautions**

The ADAMTS13 activity assay is an in vitro assay using a synthetic substrate peptide in a static liquid environment. The measured ADAMTS13 activity may not reflect the true in vivo biological ADAMTS13 activity.

Not all patients with a clinical diagnosis of idiopathic thrombotic thrombocytopenic purpura (TTP) have a severe ADAMTS13 deficiency. Conversely, patients with other non-TTP conditions may have a severe ADAMTS13 deficiency (< or =10%). These conditions include hemolytic uremic syndrome, hematopoietic stem cell and solid organ transplantation, liver disease, disseminated intravascular coagulation, sepsis, pregnancy, and certain medication. Therefore, TTP remains a clinical diagnosis.

Interferences of ADAMTS13 activity assay include high levels of endogenous von Willebrand factor, hyperlipidemia, hemolysis with plasma free hemoglobin greater than 2 g/L, hyperbilirubinemia (bilirubin concentration >100 micromolar), and cleavage by other protease.

Recent plasma exchange or transfusion may falsely normalize ADAMTS13 levels, thus potentially masking the diagnosis of TTP.

The impact of ADAMTS13 levels and presence of inhibitors on overall survival, ultimate clinical outcome, responsiveness to plasma exchange, and relapse are still controversial. Therefore, clinical correlation is recommended.

**Clinical Reference**


Test Definition: ADMIS
ADAMTS13 Inhibitor Screen

Performance

Method Description
The ADAMTS13 activity is measured by a fluorescence resonance energy transfer (FRET)-based assay using a 73 amino-acid peptide (FRETS-VWF73) of von Willebrand factor (VWF) as substrate. The inhibitor screen and titer assay are performed by using mixing studies that are similar to the Bethesda assay. One inhibitor (Bethesda) unit is defined as the concentration of an inhibitor that is able to reduce ADAMTS13 activity of normal pooled plasma by 50%.(Kokame K, Nobe Y, Kokubo Y, et al: FRETS-VWF73, a first fluorogenic substrate for ADAMTS13 assay. Br J Haematol 2005 Apr;129[1]:93-100)

PDF Report
No

Specimen Retention Time
14 Days

Performing Laboratory Location
Rochester

Fees and 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 Regional Manager. For assistance, contact Customer Service.

Test Classification
This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the U.S. Food and Drug Administration.

CPT Code Information
85335

LOINC® Information

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