
Lab Dept: Coagulation

Test Name: ADAMTS13 ACTIVITY AND INHIBITOR PROFILE

General Information

Lab Order Codes: ADM13

Synonyms: vonWillebrand Factor Cleaving Protease

CPT Codes: 85397 – ADAMTS13 activity assay
85335 – ADAMTS13 inhibitor screen assay (if appropriate)
85335 – ADAMTS13 Bethesda titer (if appropriate)

Test Includes: Testing begins with ADAMTS13 activity assay to evaluate the percent activity. If the ADAMTS13 activity assay is <30%, an inhibitor screen will be performed to look for specific ADAMTS13 inhibition. If specific inhibition is apparent, the titer of the inhibitor will be determined.

Logistics

Test Indications: Assisting with the diagnosis of congenital or acquired thrombotic thrombocytopenic purpura.

Lab Testing Sections: Coagulation

Phone Numbers: MIN Lab: 612-813-6280

STP Lab: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: ADAMTS13 Activity Assay: 24 hours; ADAMTS13 Inhibitor Assay: 3-5 days;
ADAMTS13 Bethesda Titer: 3-5 days

Special Instructions: Specimen must be drawn prior to replacement therapy.

Specimen

Specimen Type: Blood

Container: Light Blue top (Buffered Na citrate 3.2%) tube

Draw Volume: 5.4 mL blood into TWO- 3 mL tubes

Processed Volume: 2 mL platelet poor plasma

Collection:

A clean venipuncture is essential.

If the patients hematocrit is >55%, call the laboratory for a special tube. Fill tube completely.

Mix thoroughly by gentle inversion.

Special Processing:

Lab Staff: Spin down blue tubes, REMOVE plasma into a new plastic tube, RESPIN plasma to create "platelet poor plasma". Aliquot respun plasma, MINIMUM of 1 mL into TWO plastic tubes each properly labeled with patient information.

Freeze specimens immediately at -40 degrees Centigrade. Forward promptly.

Patient Preparation:

Specimen must be drawn prior to replacement therapy.

Sample Rejection:

Improper tube; clotted sample; underfilled tube; mislabeled or unlabeled specimens; gross hemolysis; gross lipemia; grossly icteric

Interpretive**Reference Range:**

ADAMTS13 Activity Assay	>or = 70%
ADAMTS13 Inhibitor Screen	Negative
ADAMTS13 Bethesda Titer	<0.4 BU
<p><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.</p>	

Critical Values:

N/A

Limitations:

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 thrombocytopenia 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 >2g/L, hyperbilirubinemia (bilirubin concentration >100 micromolar), and cleavage by 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.

Methodology:

ADAMTS13 Activity Assay: Fluorescence Resonance Energy Transfer (FRET)

ADAMTS13 Inhibitor Screen: Mixing Studies

ADAMTS13 Inhibitor Bethesda Titer: Mixing Studies

References:

[Mayo Medical Laboratories](#) March 2013