**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 patient's 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

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### Interpretive

<table>
<thead>
<tr>
<th>Reference Range:</th>
<th>ADAMTS13 Activity Assay</th>
<th>&gt;or = 70%</th>
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<tbody>
<tr>
<td></td>
<td>ADAMTS13 Inhibitor Screen</td>
<td>Negative</td>
</tr>
<tr>
<td></td>
<td>ADAMTS13 Bethesda Titer</td>
<td>&lt;0.4 BU</td>
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<10% adams13 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.

**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