Lab Dept: Coagulation

Test Name: ADAMTS13 ACTIVITY AND REFLEX INHIBITOR PROFILE

General Information

Lab Order Codes:	ADAMP
Synonyms:	vonWillebrand Factor Cleaving Protease; VWF protease activity and inhibitor; ADAMTS13
CPT Codes:	85397 – ADAMTS13 activity assay 85335 – ADAMTS13 inhibitor titer (if appropriate)
Test Includes:	Testing begins with ADAMTS13 activity assay to evaluate the percent activity. The inhibitor titer will reflex if appropriate.

Logistics

Test Indications:	Order the profile when considering a diagnosis of immune thrombotic thrombocytopenic purpura (iTTP). This diagnosis should be considered in a patient presenting with hemolytic anemia and thrombocytopenia. In patients with known iTTP/aTTP, the profile (rather than the single order ADAMTS13 activity) should be ordered if wanting to monitor presence of ADAMTS13 inhibitor.
Lab Testing Sections:	Coagulation - Sendouts
Referred to:	Mayo Clinic Laboratories (MML Test Code: ADAMP)
Phone Numbers:	MIN Lab: 612-813-6280 STP Lab: 651-220-6550
Test Availability:	Daily, 24 hours
Turnaround Time:	1-3 days
Special Instructions:	Specimen must be drawn prior to replacement therapy. Complete and Send Mayo's Coagulation Patient Information form T675. See Mayo's test catalog for link to form.

Specimen

Specimen Type:	Blood

Container: Light Blue top (sodium citrate 3.2%) tube

Draw Volume: 5.4 mL blood into TWO 3 mL tubes

Processed Volume: 2 mL platelet poor plasma

Collection: Follow standard venipuncture practice for coagulation tubes, filling to line on the tube.

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: Centrifuge tubes, REMOVE plasma from both containers into one new plastic tube, RESPIN plasma to create "platelet poor plasma." Aliquot respun plasma, MINIMUM of 1 mL into TWO plastic tubes, leaving 0.25 mL in the bottom of the centrifuged vial to avoid cell pellet. Ensure that each tube is properly labeled with patient information.

Freeze specimens immediately (within four hours of collection) at -20°C or preferably below -40 °C. Forward promptly.

Frozen plasma is stable for 2 years.

Patient Preparation: Specimen must be drawn prior to replacement therapy. Fasting is preferred.

Sample Rejection: Improper tube; clotted sample; underfilled or overfilled tube; mislabeled or unlabeled specimens; gross hemolysis; gross lipemia; grossly icteric.

Interpretive

Reference Range:

ADAMTS13 Activity Assay	>or = 70% Although not verified, the pediatric (<1 year old) reference range could be similar to or lower than that of adults.
ADAMTS13 Inhibitor Titer	<0.5 BU
<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.	

Critical Values: N/A*

*The reference lab defines a first time ADAMTS13 Activity result of ≤ 10% as semi-urgent. Per Mayo Clinical Lab policy, semi-urgent results are called and documented. Critical Values and Results - Mayo Clinic Laboratories (mayocliniclabs.com)

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:	Fluorescence Resonance Energy Transfer (FRET) with technical interpretation
References:	Mayo Clinic Laboratories May 2024
Updates	5/24/2024: Initial entry. Replaces obsolete ADM13