

## ADAMTS13 Activity with Reflex Inhibitor Profile, Plasma

**Test ID:** ADAMP

### Ordering Guidance:

Order this profile when considering a diagnosis of immune thrombotic thrombocytopenic purpura (iTTP), also known as acquired thrombotic thrombocytopenic purpura (aTTP). 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.

### Useful for:

Assisting with the diagnosis of immune or acquired thrombotic thrombocytopenic purpura.

### Profile Information:

Test ID	Reporting Name	Available Separately	Always Performed
ADAMS	ADAMTS13 Activity Assay, P	Yes	Yes
ADMI	ADAMTS13 Interpretation	No	Yes

### Reflex Tests:

Test ID	Reporting Name	Available Separately	Always Performed
ADMB	ADAMTS13 Inhibitor Titer, P	No	No
AADAM	ADAMTS13 Profile Interpretation	No	No

### Methods:

ADAMS, ADMB: Fluorescence Resonance Energy Transfer (FRET)

ADMI, AADAM: Technical Interpretation

### Reference Values:

ADAMTS13 Activity Assay

> or =70%

Although not verified, the pediatric (<1 years old) reference range could be similar to or lower than that of adults.

ADAMTS13 Inhibitor Titer

<0.5 BU

## Specimen Requirements:

**Patient Preparation:** Fasting preferred

**Collection Container/Tube:** Light-blue top (3.2% sodium citrate)

**Submission Container/Tube:** Plastic vials

**Specimen Volume:** 2 mL in 2 plastic vials each containing 1 mL

**Collection Instructions:**

1. Specimen must be collected prior to replacement therapy.
2. For complete instructions, see [Coagulation Guidelines for Specimen Handling and Processing](#)
3. Centrifuge, transfer all plasma into a plastic vial, and centrifuge plasma again.
4. Aliquot plasma (1 mL per aliquot) into 2 separate plastic vials, leaving 0.25 mL in the bottom of centrifuged vial.
5. Freeze plasma immediately (no longer than 4 hours after collection) at -20 degrees C or, ideally, below -40 degrees C.

**Minimum Volume:** 2 mL

## Addition Information:

1. Double-centrifuged specimen is critical for accurate results as platelet contamination may cause spurious results.
2. Each coagulation assay requested should have its own vial.

Specimen Stability information: Frozen 2 years

## Specimen Stability Information:

Specimen Type	Temperature	Time
Plasma Na Cit	Frozen	

## Cautions:

This ADAMTS13 activity assay is an in vitro assay using a synthetic substrate peptide in a static liquid environment. The measure ADAMTS13 activity may not reflect the true in vivo biological ADAMT13 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.

Samples collected in EDTA instead of 3.2% sodium citrate will result in artificially reduced ADAMTS13 activity.

Interferences of the ADAMTS13 activity assay include high levels of endogenous von Willebrand factor, hyperlipidemia, hyperbilirubinemia (bilirubin concentration >30 mg/dL), and cleavage by other proteases.

Recent plasma exchange or plasma 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.

**CPT Code:**

85397

85335 (if appropriate)

**Day(s) Performed:** Monday through Friday, Sunday **Report Available:** 1 to 3 days

**Questions**

Contact Bonnie Meyers, Laboratory Resource Coordinator at 800-533-1710.