Diagnostic algorithm¹

Differential diagnosis for primary TMAs (including ADAMTS13)¹

1. Evidence of TMA

Thrombocytopenia

Platelet Count <150,000/mm³ or >25% Decrease From Baseline

Microangiopathic Hemolysis

AND

Schistocytes

and/or Elevated LDH

and/or Decreased Haptoglobin

and/or Decreased Hemoglobin

PLUS ONE OR MORE OF THE FOLLOWING

2. Symptoms

Cardiovascular Symptoms

Myocardial Infarction

and/or Cardiomyopathy

and/or Heart Failure

and/or Peripheral Gangrene

Neurological Symptoms

Confusion

and/or Seizures

and/or Other Cerebral

Abnormalities

Renal Impairment

Elevated Creatinine

and/or Decreased eGFR

and/or Elevated Blood Pressure

and/or Abnormal Urinalysis

Gastrointestinal Symptoms

Diarrhea ± Blood and/or Nausea/Vomiting and/or Abdominal Pain and/or Gastroenteritis

Pulmonary Symptoms

Dyspnea

and/or Pulmonary Hemorrhage

and/or Pulmonary Edema

Visual Symptoms

Pain and Blurred Vision Retinal Vessel Occlusion Ocular Hemorrhage

3. Please choose below to show results for:

✓ Differential diagnosis for primary TMAs (including ADAMTS13)¹



Watch this short video to see how presentations can overlap

OR

Patient approach to a TMA associated with a complement-amplifying condition¹



Watch this short video to learn about the complement-amplifying conditions associated with aHUS

4. Evaluate ADAMTS13 Activity and Shiga-toxin/EHEC* Test

In the absence of ADAMTS13 results, a serum creatinine level >150–200 μ mol/L (>1.7–2.3 mg/dL) or a platelet count of >30,000/mm³ almost eliminates a diagnosis of severe ADAMTS13 deficiency (TTP).

≤10% ADAMTS13 Activity

>10% ADAMTS13 Activity

Shiga-toxin/EHEC Positive

STEC-HUS

TTP

aHUS

- Genetic mutations are not identified in 30–50% of patients with aHUS.
- A diagnosis of aHUS does not require identification of a mutation.

ADAMTS13, A Disintegrin and Metalloproteinase with a ThromboSpondin type 1 motif, member 13; eGFR, estimated Glomerular Filtration Rate; EHEC, Enterohemorrhagic *E. coli*; LDH, Lactate Dehydrogenase; STEC-HUS, Shiga-Toxin-Producing *E. coli* Hemolytic Uremic Syndrome; TMA, Thrombotic Microangiopathy; TTP, Thrombotic Thrombocytopenic Purpura.

1. Laurence J, et al. Clin Adv Hematol Oncol. 2016 14 Suppl 11(11):2-15.



^{*} Shiga-toxin/EHEC test is warranted in history/presence of GI symptoms.

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Patient approach to a TMA associated with a complement-amplifying condition¹

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4. Perform panel of TMA tests (to include ADAMTS13 and Shiga-toxin/EHEC tests)*

AND

If CAC is identified (e.g., pregnancy, SLE, MHT, drug-induced)

Treat condition to resolve TMA

If TMA unresolved or reoccurs



ADAMTS13 >10%

Coexisting aHUS

* Shiga-toxin/EHEC test is warranted in history/presence of GI symptoms.

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1. Laurence J, et al. Clin Adv Hematol Oncol. 2016 14 Suppl 11(11):2-15.

