Differential diagnosis for TMAs: aHUS, TTP, and STEC-HUS

Thrombocytopenia\(^1,2\)
- Platelet Count <150,000
  - or >25% Decrease
  - From Baseline\(^1\)

AND

Microangiopathic Hemolysis\(^2,3\)
- Schistocytes\(^2,3\) and/or
- Elevated LDH\(^2\) and/or
- Decreased Haptoglobin\(^2\) and/or
- Decreased Hemoglobin\(^2\)

Plus one or more of the following:

Neurological Symptoms\(^4-7\)
- Confusion\(^4,5\) and/or
- Seizures\(^6,7\) and/or
- Other Cerebral Abnormalities\(^5\)

Renal Impairment\(^2,8-10\)
- Elevated Creatinine\(^9\) and/or
- Decreased eGFR\(^2,9\) and/or
- Elevated Blood Pressure\(^10\) and/or
- Abnormal Urinalysis\(^8\)

Gastrointestinal Symptoms\(^2,6,11\)
- Diarrhea +/- Blood\(^11\) and/or
- Nausea/Vomiting\(^6\) and/or
- Abdominal Pain\(^6\) and/or
- Gastroenteritis\(^2,11\)

Evaluate ADAMTS13 Activity and Shiga-toxin/EHEC\(^*\) Test\(^12-15\)

While waiting for ADAMTS13 results, a platelet count of >30,000/mm\(^3\) or a serum creatinine level >150-200 \(\mu\)mol/L (1.7-2.3 mg/dL) almost eliminates a diagnosis of severe ADAMTS13 deficiency (TTP)\(^16\)

\(\leq5\%\) ADAMTS13 Activity\(^12,14,15\)

TTP

\(>5\%\) ADAMTS13 Activity\(^15\)

aHUS

Shiga-toxin/EHEC Positive\(^13\)

STEC-HUS\(^*\)

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

The information on this page is intended as educational information for healthcare providers. It does not replace a healthcare professional’s judgment or clinical diagnosis.

High clinical suspicion of aHUS is required in all patients presenting with any sign or symptom of systemic, complement-mediated thrombotic microangiopathy\(^17\)
## Diagnostic tests to aid differentiation of aHUS from TTP or STEC-HUS

<table>
<thead>
<tr>
<th>Specimen source</th>
<th>Peripheral blood</th>
<th>Stool/Rectal swab</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sample volume</td>
<td>1-3 mL</td>
<td>1-2 g, 1-2 mL, 1 swab</td>
</tr>
<tr>
<td>Collection time</td>
<td>Prior to any plasma intervention</td>
<td>Immediately upon suspicion; prior to antibiotic therapy</td>
</tr>
<tr>
<td>Sample requirement</td>
<td>Citrated plasma, frozen</td>
<td>Refrigerated/Cold pack</td>
</tr>
<tr>
<td>Anticoagulant</td>
<td>Sodium citrate (light blue-top tube) preferred; EDTA is not acceptable</td>
<td></td>
</tr>
<tr>
<td>Expected turnaround time (TAT)</td>
<td>24-48 hrs</td>
<td>24-48 hrs</td>
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</tbody>
</table>

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The catastrophic consequences of aHUS make rapid diagnosis critical

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