**T1** Summary of testing used to evaluate suspected TTP

| TEST | Indication/Utility/Limitations |
|---|---|
| Routine hematology and plasma chemistry testing | • Widely available essential testing for initial evaluation of MAHA and thrombocytopenia<br>• In TTP, results indicate intravascular hemolytic anemia with frequent schistocytes, marked thrombocytopenia, strikingly high LDH, and minimal renal dysfunction<br>• Clinical prediction scores using routine tests facilitate initiation of appropriate therapy<br>• Used to monitor treatment efficacy and help determine length of TPE (continued to recovery of hemolysis and thrombocytopenia and resolution of neurologic symptoms) |
| Routine coagulation testing | • Widely available<br>• Used to exclude DIC as a cause of MAHA and thrombocytopenia<br>• Normal or only mild derangements in most TTP cases |
| ADAMTS13 activity | • Evaluate suspected TTP, defined as MAHA and thrombocytopenia without alternative cause<br>• Limited availability, treatment decisions made before results available but are frequently modified when results are available<br>• Diagnostic testing must be performed on pre-treatment specimens<br>• Severe deficiency (<10% of normal activity) compatible with TTP (high sensitivity and specificity)<br>• Severe deficiency correlates with response to therapy and risk of relapse |
| ADAMTS13 antibody testing | • Can be used to confirm acquired TTP in cases with severe ADAMTS13 deficiency<br>• Differentiates acquired from hereditary TTP<br>• Limited availability, Bethesda assays are technically complex LDTs<br>• Diagnostic testing must be performed on pre-treatment specimens<br>• Presence of antibodies correlates with response to therapy and risk of relapse<br>• Bethesda assays are more specific but less sensitive than ELISA assays, algorithmic testing panels are useful<br>• Currently available methods do not detect the causative antibodies in every case of acquired TTP |
| ADAMTS13 gene sequencing | • Expensive and with limited availability<br>• Identifies the mutations responsible for hereditary TTP (Upshaw-Schulman syndrome), which confirms the diagnosis and simplifies testing for family members<br>• May assist in differentiation of acquired and inherited TTP in complex cases |

Legend: MAHA = Microangiopathic hemolytic anemia;
TTP = thrombotic thrombocytopenic purpura;
DIC = disseminated intravascular coagulation;
LDT = laboratory-developed tests