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