ADAMTS13 testing algorithm for evaluation of patients with suspected TTP. Diagnostic testing should be performed on pre-treatment specimens. The algorithm depicts the typical case, but note that rare TTP cases with only mild or moderately decreased ADAMTS13 have been reported, as have non-TTP cases with severe ADAMTS13 deficiency.

**ADAMTS13 ACTIVITY**

- Normal
  - TTP excluded, pursue other diagnoses
- Mild to moderate decrease
  - TTP unlikely, apply close clinicopathologic correlation to explain decrease
- Severe deficiency, TTP confirmed, pursue ADAMTS13 antibody testing if desired
  - Bethesda assay
    - Not detected
    - ELISA assay
      - Not detected
      - ADAMTS13 Sequencing if hereditary TTP suspected
        - Positive
          - Hereditary TTP
        - Negative
          - Requires clinicopathologic correlation
    - Detected
      - Acquired TTP