Table 1. Primary TMA syndromes

Disorder	Mechanism
TTP	Homozygous or compound heterozygous mutations ADAMTS13 mutations (hereditary or Upshaw-Schulman syndrome) or autoantibodies against ADAMTS13 (acquired)
ST-HUS	Enteric infection with ST-producing *Escherichia coli* or *Shigella dysenteriae*
Complement-mediated or atypical HUS	Mutations in the complement genes causing uninhibited activity of the alternative pathway or antibodies against complement factor H
Drug-induced	Immune reaction, possibly through drug-dependent antibodies. Observed with ticlopidine/clopidogrel (2), quinine (3), and gemcitabine (4) or toxic, dose-related reaction through various mechanisms
Metabolism-mediated	Homozygous mutations in *MMACHC*
Coagulation-mediated	Homozygous mutations in *DGKE, PLG,* and *THBD*

Based on reference 1. Numbers in parentheses are supporting references. HUS= hemolytic uremic syndrome; ST= Shiga toxin; TMA= thrombotic microangiopathy; TTP= thrombotic thrombocytopenic purpura.