Thrombotic Microangiopathy (TMA) - Classification

TMA defined by:
- MAHA
- Thrombocytopenia
- Organ dysfunction (from occlusive thrombi)

Primary TMA

TTP
Defined by low ADAMTS13 < 10%

- **Congenital**
  - aka hereditary TTP, Upshaw-Schulman syndrome
- **Acquired**
  - aka immune TTP
  - **Primary**
  - **Secondary**
    - Infection
    - Autoimmune disease
    - Cancer
    - Transplantation
    - HIV infection
    - Drugs

HUS
ADAMTS13 > 10-20%

- STEC-HUS
- SP-HUS
- aHUS

Secondary TMA

ADAMTS13 > 10-20%

- Malignant HTN
- HIV
- Other viruses
- CTD
  - SRC
  - SLE
  - CAPS
  - Vasculitis
- Cancer
- Transplant
- Drugs
- Pregnancy
- Pancreatitis
- Cobalamin C

Some consider DIC a type of TMA (but differs in that thrombi in DIC are fibrin rich, not platelet rich)

TTP, thrombotic thrombocytopenic purpura; HUS, hemolytic uremic syndrome; STEC, Shiga toxin-producing Escherichia coli; SP, Streptococcus pneumonia; aHUS, atypical HUS; CTD, connective tissue disease; SCR, scleroderma renal crisis; SLE, systemic lupus erythematosus; CAPS, catastrophic antiphospholipid syndrome