Plasma Exchange (PEX)-Free Regimen for Immune-Mediated TTP (iTTP) (*Posted 4/15/2020*)

1. Indications for this approach (A+B):

A. Patients with clinical features of iTTP (microangiopathic hemolytic anemia, severe thrombocytopenia [platelets ≤ 30 x 10^9/L] and mild renal involvement [serum creatinine ≤ 2.27 mg/dL; 200 µmol/L) that are predictive of severe ADAMTS13 deficiency[1];
AND

B. Unavailability/limited access to plasma exchange (context of SARS-CoV-2 pandemic); without possibility of transfer to a site with capabilities

2. Treatment regimen:

A. **Plasma infusion:** 10-15 cc/kg daily until normal platelet count (>150 x10^9/L for 2 days and normalized LDH as tolerated, based on volume status and ability to manage the volume of plasma infused **(Omit if plasma refused for religious or other reasons[2,3])**

B. **Immunosuppression**:
- Prednisone or Prednisolone 1.0-1.5 mg/kg/day for 3 weeks, then taper over 5 days;
- Rituximab 375 mg/m2 IV one dose between days 1-4-and then weekly for a total of 4 doses; Premedicate with antihistamines and acetaminophen before each rituximab infusion to prevent allergic/anaphylactic reactions

C. **Caplacizumab** 10 or 11 mg IV before the first plasma infusion, then 10 or 11 mg subcutaneously daily following each plasma administration, until ADAMTS13 activity stable at ≥20% on at least 2 measurements, obtained at least 24 hours after plasma infusion;

3. Supportive care:

A. Transfusion of RBC if hemoglobin < 7 g/dL or anemia is poorly tolerated;

B. Prevention of fluid overload with diuretics;

C. Administration of folic acid 5 mg/day orally until anemia and hemolysis resolve

4. Follow-up:

A. Daily:
- Clinical: Blood pressure, pulse, weight, diuresis, respiratory rate, SaO2, temperature;
- Laboratory: CBC, reticulocyte count, LDH, serum creatinine, standard blood chemistry, troponin if elevated, until normalizes;

B. Weekly: beginning the last day of plasma infusion (or after normalization of platelet count if no plasma administered):
- ADAMTS13 activity, until ADAMTS13 activity stable at ≥ 20% on 2 occasions, obtained at least 24 hours after plasma infusion

References:

1. Coppo P, Schwarzinger M, Buffet M, et al. Predictive features of severe acquired ADAMTS13 deficiency in idiopathic thrombotic microangiopathies: the French TMA reference center experience. *PLoS One*. 2010;5(4):e10208.

2. Chander DP, Loch MM, Cataland SR, George JN. Caplacizumab Therapy without Plasma Exchange for Acquired Thrombotic Thrombocytopenic Purpura. *N Engl J Med*. 2019;381(1):92-94.

3. Sukumar S, George JN, Cataland SR. Shared decision making, thrombotic thrombocytopenic purpura, and caplacizumab. *Am J Hematol*. 2020.