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General Hematology Protocols

Acquired Hemophilia
- High aPTT, new onset of bleeding/ecchymosis
- Test: Factor 8 level with reflex inhibitor
- Tx:
  - Bleeding: rFVIIa 90 ug/kg
  - Inhibitor:
    - Prednisone 1mg/kg
    - Cyclophosphamide 100mg po
    - Rituximab 1000mg IV day 1 and 14

aHUS
- Worsening renal function, schistocytes, thrombocytopenia, HTN, high LDH
- Idiopathic or provoked by transplant and antineoplastic
- Test: sC5-9/MAC, aHUS genetic panel, ADMATS13 > 10%
- Tx: Eculizumab 900mg iv weekly x 4 then 1200mg every other week
  - Goal to keep CH50 < 3

Autoimmune Hemolytic Anemia
- Anemia, low haptoglobin, high LDH, spherocytes,
- Testing: spherocytes, positive DAT (consider "super Coombs' if DAT negative)
- Tx
  - Warm: Prednisone 1mg/kg, Rituximab 1000mg IV day 1 and 14
    - Splenectomy for refractory cases
  - Cold: Rituximab 1000mg IV day 1 and 14

Cancer related Thrombosis
- Proximal DVT or PE in the setting of active cancer
- Tx: Apixaban 10mg bid x 7 days then 5mg bid
- Duration – as long as active disease
  - Continue 3 months after end of adjuvant chemotherapy if patient is NED

Hemophilia
- Factor 8 replacement dose:
  - (desired Factor 8 concentration - current level) * weight (kg)/2
  - 50 units/kg emergency dosing
- Factor 9 replacement dose:
  - (desired Factor 9 concentration - current level) * weight (kg)
  - 100 units/kg Emergency dosing
- Continuous infusion of products
  - Factor 8: Bolus of 50 units/kg followed by a continuous infusion of 3.5-5 units/hour guided by levels.
  - Factor 9: load with 100 units/kg and then use a continuous infusion of 3.5-4 units/hour guided by levels.

Heparin Induced Thrombocytopenia
- Suspect if platelets fall by 50% or new clots on heparin
- DX: 4T score > 3: check Anti-PF4, if + then send serotonin release
- Tx: need to treat empirically while waiting for PF4
  - Argatroban: Dose at 2 ug/kg/min infusion with dose adjustments to keep aPTT 1.5 – 3 x control
    - Liver disease 0.5 ug/kg/min
    - ICU 1.0 ug/kg/min
  - DOAC: increasing data – good option for not critical ill patients or fresh arterial clots
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- Bivalirudin: Limited data - most useful in HIT patients needing PCI

**Intravenous Iron**
- Options
  - INFeD (iron dextran) 1 gram over one hour
  - Feraheme (ferumoxytol) 1020mg over 15 minutes
  - Injectifer (iron carboxymaltose) 750 mg x 2 1 week apart

**ITP**
- Pretreatment labs: Hep B and C screening, HIV
- TX:
  - Dexamethasone 40mg x 4 days every 14 days x 4
  - If bleeding add IVIG 1 gram/kg x 1
  - Platelet transfusion ONLY if life threatening bleeding
- Refractory to initial therapy
  - Platelet boilermaker: continuous IVIG 1 gm/kg over 24 hours along with continuous platelets (one platelet pheresis unit over 6 hours’ x 4)
  - Vincristine 1.4mg/m2 weekly
  - Rituximab 1000mg x 2 day 1 and 14
  - Eltrombopag 50 mg daily

**Plasmapheresis/Exchange**
- TTP:
  - 1.5 plasma volume daily
  - Replacement: FFP
- Autoimmune (neuro, rheum crisis)
  - 1.0 plasma volume every other day x 5
  - Replacement: Albumin
  - Follow INR/PTT/Fibrinogen
  - If low the day of exchange then replace with 50% Albumin and 50% FFP

**Preferred Drug Dosing**
- Cyclophosphamide
  - 1000mg IV monthly
- Dexamethasone (ITP)
  - 40mg x 4 days every 14 days x 4
- Eculizumab
  - Meningoccal vaccine before
  - PNH: 900mg weekly x 4 then 900mg every other week
  - aHUS: 900mg weekly x 4 then 1200mg every other week
- IVIG
  - Obtain Hep B screening before (can cause false positive HepB core)
  - 1 gram/kg x 1
- Mycophenolate
  - 500mg bid then raise in 1000mg bid if ANC > 1500
- Rituximab (immunosuppressive)
  - Obtain Chronic Hep B panel before giving
  - Rituximab 1000 mg IV day 1 and 14
- Vincristine (max at 2mg/m2)
  - ITP: Vincristine 1.4mg/m2 weekly
  - TTP: Vincristine 1.4mg/m2 days 1, 4, 7, 10

**Reversal of anticoagulation in ICH/life threatening bleeding**
- Xa inhibitors: 50 units/kg Kcentra
- Dabigatran: 5 grams Idarucizumab
• Warfarin: 2000 units Kcentra + 10 mg IV vitamin K

TTP
• Schistocytes, thrombocytopenia, HTN, high LDH
• Tests: ADAMTS13
• TX:
  o Plasma exchange 1.5 plasma volumes replace with FFP
    ▪ Continue at least 5 days until LDH and platelet normal
  o Prednisone 1 mg/kg daily
  o Rituximab 1000 mg q 14 days if + ADAMTS13 antibodies
  o Refractory: Vincristine 1.4mg/m2 days 1, 4, 7, 10

Splenectomy
• Asplenic Recommendations:
  o Pneumococcal vaccination q5 years
  o Haemophilus influenzae and meningitis vaccination
  o Yearly flu vaccines
  o PRN antibiotic available (amoxicillin/clavulanic acid) if she shows rapid onset fever, rigors, etc. plus she was told to report to the Emergency Department immediately with any fever and or rigor
  o Prophylactic antibiotics for dog bites (amoxicillin/clavulanic acid) (can see overwhelming Capnocytophaga canimorsus infections)
  o Med alert bracelet

Visceral Vein Thrombosis
• Portal Vein
  o Cirrhosis: incidental and not in SMV: observe
  o Cirrhosis: symptomatic and/or in SMV: Anticoagulate
  o No Cirrhosis
    ▪ Provoked: 3 months’ therapy
    ▪ Idiopathic: indefinite therapy, screen for MPN/PNH/APLA
• Other
  o Provoked: 3 months
  o Idiopathic: indefinite therapy, screen for MPN/PNH/APLA

Von Willebrand Disease
• Treatment by type
  o Type 1: Desmopressin
  o Type 2A: Desmopressin (Only Effective in 10%), Humate-P
  o Type 2B: Humate-P
  o Type 2N: Desmopressin (mild) Factor 8 concentrates
  o Type 2M: Desmopressin (mild)/Humate-P
  o Type 3: Humate-P
  o Platelet-Type: Platelets+Humate-P, rFVIIa
• Treatments
  o Intravenous Desmopressin 0.3 ug/kg can be repeated daily (3 days limit)
  o Intranasal Desmopressin 300 ug (150 ug/nostril) (3 days limit)
  o Humate-P:
    ▪ Levels below 30%: 40-50 IU/kg Followed by 20 IU/kg Every 12 Hours
    ▪ Levels above 30%: 20-40 IU/kg Every Day