Deloughery

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

- 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
  - Injecitfer (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, rhem 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:
  - Plasma exchange 1.5 plasma volumes replace with FFP
    - Continue at least 5 days until LDH and platelet normal
  - Prednisone 1 mg/kg daily
  - Rituximab 1000 mg q 14 days if + ADAMTS13 antibodies
  - Refractory: Vincristine 1.4mg/m2 days 1, 4, 7, 10

Splenectomy

- Asplenic Recommendations:
  - Pneumococcal vaccination q5 years
  - Haemophilus influenzae and meningitis vaccination
  - Yearly flu vaccines
  - 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
  - Prophylactic antibiotics for dog bites (amoxicillin/clavulanic acid) (can see overwhelming Capnocytophaga canimorsus infections)
  - Med alert bracelet

Visceral Vein Thrombosis

- Portal Vein
  - Cirrhosis: incidental and not in SMV: observe
  - Cirrhosis: symptomatic and/or in SMV: Anticoagulate
  - No Cirrhosis
    - Provoked: 3 months' therapy
    - Idiopathic: indefinite therapy, screen for MPN/PNH/APLA
- Other
  - Provoked: 3 months
  - Idiopathic: indefinite therapy, screen for MPN/PNH/APLA

Von Willebrand Disease

- Treatment by type
  - Type 1: Desmopressin
  - Type 2A: Desmopressin (Only Effective in 10%), Humate-P
  - Type 2B: Humate-P
  - Type 2N: Desmopressin (mild) Factor 8 concentrates
  - Type 2M: Desmopressin (mild)/Humate-P
  - Type 3: Humate-P
  - Platelet-Type: Platelets+Humate-P, rFVIIa

- Treatments
  - Intravenous Desmopressin 0.3 ug/kg can be repeated daily (3 days limit)
  - Intranasal Desmopressin 300 ug (150 ug/nostril) (3 days limit)
  - 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