Dr. Jacob Pendergrast, Sickle Cell Disease

- **PRINCIPLES**
  - Decr HgbS%, generally more important than increasing total Hgb
  - Benefit only with high-shear vasculature
  - Ceiling of Hgb ~100 g/L
- **WEAK EVIDENCE WITH PREGNANCY**
  - Available evidence suggests more benefit for mom than developing fetus
  - There may be exceptions (eg., signs of placental insufficiency, prev IUGR)
- **CAUTION WITH SEVERE ANEMIA**
  - Aplastic crisis: *volume overload*
  - Sequestration: *autotransfusion*
  - Hyperhemolysis: *worsening anemia*
- **GOOD EVIDENCE FOR STROKE PREVENTION**
  - Transfusion indicated for all children with high-risk dopplers and history of stroke
  - Smaller value for children with SCIs
  - Limited evidence in adults; look for other causes, caution with hemorrhagic stroke
- **NUANCED APPROACH FOR SURGERY**
  - Usually not needed for low-risk patient with low risk procedure
  - Indicated for everyone else, top-up vs exchange depends on comorbidity, procedure risk, baseline hemoglobin
- **THERAPEUTIC TRANSFUSION IF ACUTE ORGAN COMPROMISE**
  - Limited evidence, but consensus supports transfusion for acute stroke, acute chest syndrome, sickle hepatopathy
  - Other situations: "if all else fails"
- **SELECTION OF RBCs MUST BE DONE WITH CARE!**
  - Tell your blood bank early that your patient has sickle cell, provide detailed transfusion history

Congential Coag – VWD, Hemophilia, Dr. Michelle Sholzberg

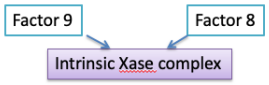
- The bleeding history is the most important test of hemostasis

Von Willebrand Disease

- Diagnosis: 3 Clinical Components
  - Bleeding Symptoms
  - +/- Family History
  - Laboratory results

Bleeding Symptoms	Treatment
Mucocutaneous • Menorrhagia • Epistaxis • Bruising • Excessive bleeding from minor wounds • GI bleeding • Oral cavity/post-dental procedure • Post-operative • Post-partum Musculoskeletal (Type 3 VWD) • Hemarthrosis • Soft tissue, muscle hematomas	• **Call Hematology/Transfusion Medicine** • DDAVP (Desmopressin) • Anti-fibrinolytic agents (TXA) • Transfusion based – VWF:FVIII concentrate – Humate-P or Wilate

Hemophilias:



Hemophilia B
- Factor IX deficiency
- 1 in 30,000 males

Hemophilia A
- Factor VIII deficiency
- 1 in 5,000 males

Bleeding Symptoms	Treatment
- Musculoskeletal bleeding o Hemarthrosis o Intra-muscular hematoma - Mouth bleeding, epistaxis - Intracranial bleeding - Bleeding with trauma, procedures, surgery - Menorrhagia (symptomatic carriers)	• **Call Hematology/Transfusion Medicine** • Anti-fibrinolytic agents (TXA) • Transfusion based – specific factor concentrate • – Factor VIII: Xyntha, Kovaltry, Nuwiq, Adynovate – Factor IX: Benefix, Rebinyn • DDAVP/Desmopressin – for mild hemophilia A (FVIII >10%)

Resources

- "Principles of Management of Urgent Bleeding in Hemophilia" - developed by Dr. Jerry Teitel
- Blood Easy: Coagulation Simplified – developed by ORBCoN
- Illustrated Review of Bleeding Assessment Tools and Coagulation tests (Elbaz, Sholzberg)