



Dr. Jacob Pendergrast, Sickle Cell Disease

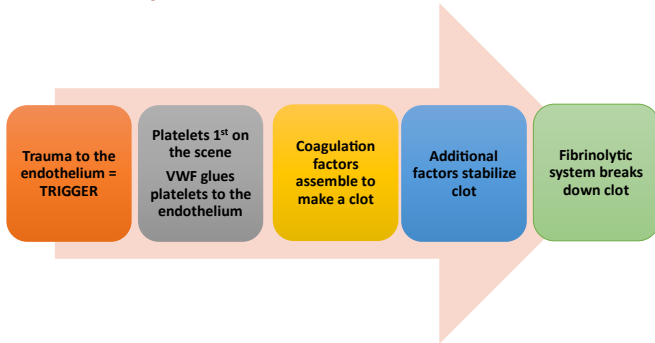
QUESTIONS/COMMENTS?

<p>▶ PRINCIPLES</p> <ul style="list-style-type: none"> ▶ Decr HgbS%, generally more important than increasing total Hgb ▶ Benefit only with high-shear vasculature ▶ Ceiling of Hgb ~100 g/L 	<p>▶ WEAK EVIDENCE WITH PREGNANCY</p> <ul style="list-style-type: none"> ▶ Available evidence suggests more benefit for mom than developing fetus ▶ There may be exceptions (eg., signs of placental insufficiency, prev IUGR)
<p>▶ CAUTION WITH SEVERE ANEMIA</p> <ul style="list-style-type: none"> ▶ Aplastic crisis: <i>volume overload</i> ▶ Sequestration: <i>autotransfusion</i> ▶ Hyperhemolysis: <i>worsening anemia</i> 	<p>▶ GOOD EVIDENCE FOR STROKE PREVENTION</p> <ul style="list-style-type: none"> ▶ 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
<p>▶ NUANCED APPROACH FOR SURGERY</p> <ul style="list-style-type: none"> ▶ 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 	<p>▶ THERAPEUTIC TRANSFUSION IF ACUTE ORGAN COMPROMISE</p> <ul style="list-style-type: none"> ▶ Limited evidence, but consensus supports transfusion for acute stroke, acute chest syndrome, sickle hepatopathy ▶ Other situations: "if all else fails"
<p>▶ SELECTION OF RBCs MUST BE DONE WITH CARE!</p> <ul style="list-style-type: none"> ▶ Tell your blood bank early that your patient has sickle cell, provide detailed transfusion history 	



Dr. Natasha Rupani, Dr. Michelle Sholzberg - Congenital Coag – VWD, Hemophilia

Hemostasis Simplified



The bleeding history is the most important TEST of hemostasis, using a validated bleeding assessment tool (BAT).

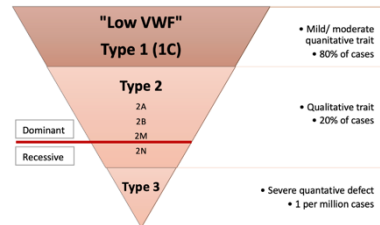
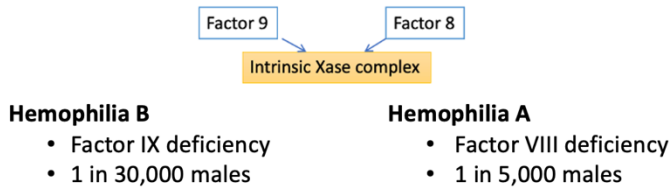
A normal PT and aPTT does not rule out a bleeding disorder.

Von Willebrand Disease

Diagnosis: 1) Bleeding Symptoms, 2) Family History, 3) Laboratory Results

Bleeding Symptoms		Treatment
Mucocutaneous <ul style="list-style-type: none"> Heavy menstrual bleeding Epistaxis Bruising Excessive bleeding from minor wounds GI bleeding Oral cavity/post-dental procedure Post-operative Post-partum 	Musculoskeletal (Type 2N, 3) <ul style="list-style-type: none"> Hemarthrosis Soft tissue, muscle hematomas 	Call Hematology/Transfusion Medicine Principle of treatment: Increase or replace VWF <ul style="list-style-type: none"> DDAVP (Desmopressin) VWF:FVIII Concentrate (Humate P, Wilate) Adjunctive anti-fibrinolytic agent (TXA)

Hemophilias:



Bleeding Symptoms	Treatment
<ul style="list-style-type: none"> Musculoskeletal bleeding <ul style="list-style-type: none"> Hemarthrosis Intra-muscular hematoma Mouth bleeding, epistaxis Intracranial bleeding Bleeding with trauma, procedures, surgery Heavy menstrual bleeding (symptomatic carriers) 	Call Hematology/Transfusion Medicine Principle of treatment: Replace deficient factor <ul style="list-style-type: none"> Factor VIII: Xyntha, Kovaltry, Nuwiq, Adynovate, Jivi Factor IX: Benefix, Rebinyn DDAVP (Desmopressin) – mild hemophilia (FVIII>10%) Non-factor therapies: Emicizumab <ul style="list-style-type: none"> Avoid PCC – risk of thrombosis Inhibitor present - rVIIa No inhibitor – FVIII concentrate Adjunctive anti-fibrinolytic agent (TXA)

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)