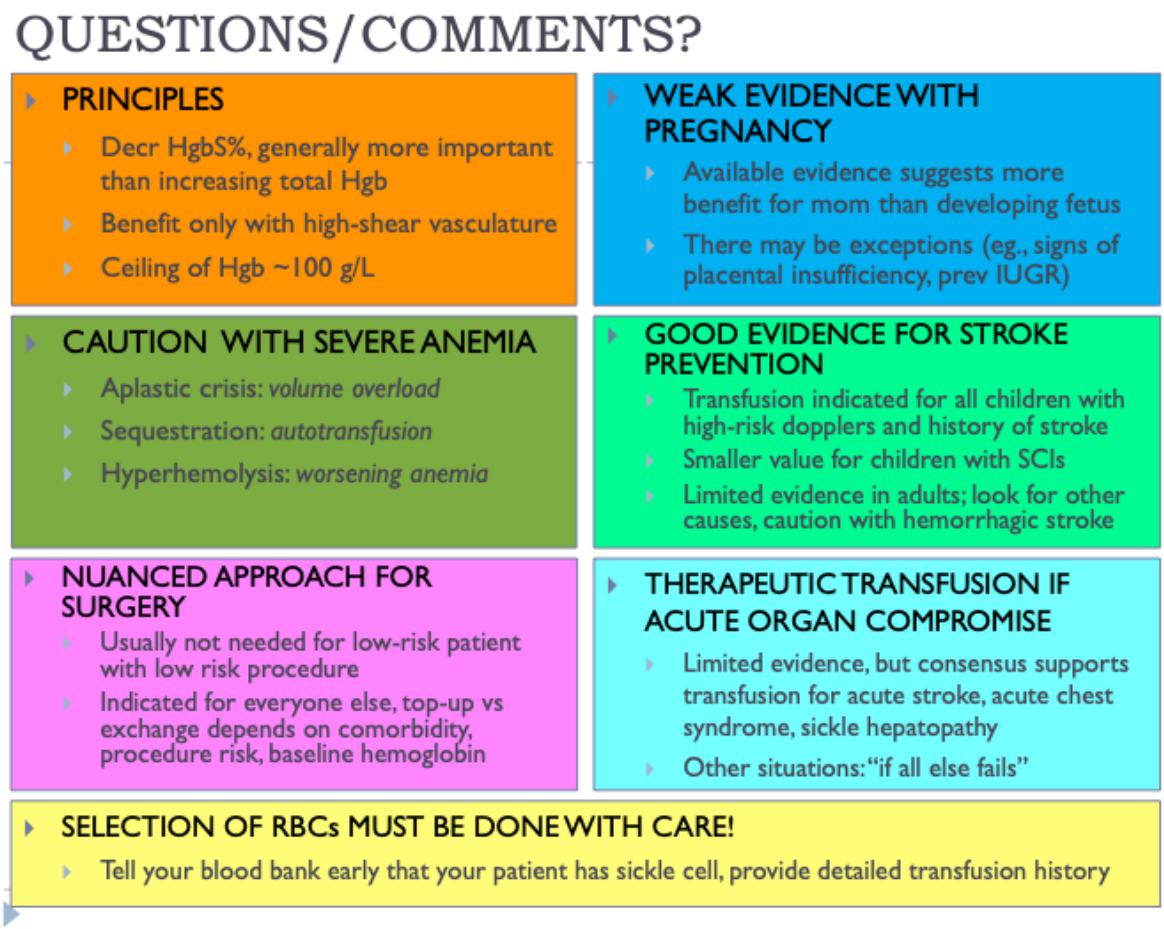
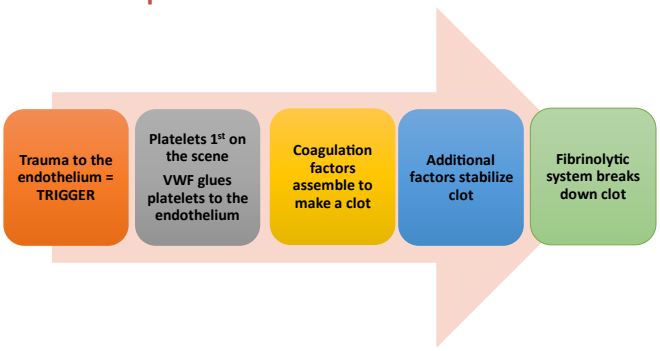
Dr. Jacob Pendergrast, Sickle Cell Disease



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

Hemostasis Simplified



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

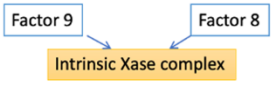
A normal PT and aPTT does not rule out a bleeding disorder.

Von Willebrand Disease

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

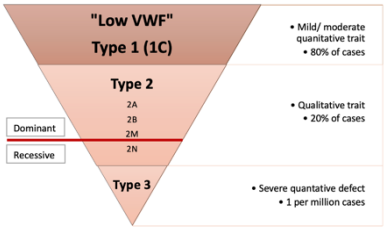
Bleeding Symptoms		Treatment
Mucocutaneous ▪ Heavy menstrual bleeding ▪ Epistaxis ▪ Bruising ▪ Excessive bleeding from minor wounds ▪ GI bleeding ▪ Oral cavity/post-dental procedure ▪ Post-operative ▪ Post-partum	**Musculoskeletal** (Type 2N, 3) ▪ Hemarthrosis ▪ Soft tissue, muscle hematomas	**Call Hematology/Transfusion Medicine** Principle of treatment: Increase or replace VWF ▪ DDAVP (Desmopressin) ▪ VWF:FVIII Concentrate (Humate P, Wilate) ▪ Adjunctive anti-fibrinolytic agent (TXA)

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 ▪ Heavy menstrual bleeding (symptomatic carriers)	**Call Hematology/Transfusion Medicine** Principle of treatment: Replace deficient factor	
	▪ Factor VIII: Xyntha, Kovaltry, Nuwiq, Adynovate, Jivi, ▪ Factor IX: Benefix, Rebinyn ▪ DDAVP (Desmopressin) – mild hemophilia (FVIII>10%)	Non-factor therapies: Emicizumab ▪ 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)