Coagulation Disorders

Coagulopathy and Hypercoagulability

	Path	Clinical/Diagnosis	Treatment
VWD	AD/AR def. of VWF → abnormal mucocutaneous bleeding	Bruising, mucosal bleeding, menorrhagia. Typically VWF Ag and Activity low, may have decreased FVIII activity and prolonged PTT	Bleed: DDAVP (if responder) Severe bleed: VWF conc. Menorrhagia: OCPs Avoid aspirin use
Hemophilia	X-linked inheritance Hemophilia A: FVIII Def Hemophilia B: FIX Def	Hemarthrosis, ICH, mucosal bleeding, epistaxis, occ. hematuria, GI bleed. Prolonged PTT, decreased FVIII or FIX activity; PT and plt wnl	Hemophilia A: FVIII concentrates (DDAVP for some mild pts) Hemophilia B: FIX
Vit K Def (Warfarin Use)	Dec. synthesis of FX, IX,VII, II, Protein C, S Epi: neonates, antibiotics, malabsorp. (panc, celiac, IBD)	Easy bruising, mucosal bleeding, melena, hematuria, ICH (newborns) Inc. PT and PTT	Vit K (PO or IM) Acute bleed: FFP or PCC
DIC	Widespread pathologic intravascular coagulation → plt/factor consump., hemolysis Causes: **STOP M**aking **T**hrombi (**S**epsis, **T**rauma, **O**B comp, **P**ancreatitis, **M**alig, **T**ransfusion	Bleeding from wound/surgical site Hemoptysis, venous/art. Thrombosis → organ ischemia. HypoTN, jaundice, ext. cyanosis. Dec. Plts, fibrinogen, haptoglobin Inc. PT/PTT, D-Dimer, LDH	Treat underlying cause Aggressive support Acute bleed: Plt transfusion + FFP +/- RBC transfusion.
Inherited Hyper-Coagulable States	Factor V Leiden	FV cannot be inactivated by Prot C	Life-long anticoagulation in the setting of homozygous inheritance and prior venous thromboembolism (VTE)
	Prothrombin 20210 mutation	Increased Prothrombin levels	
	Antithrombin deficiency	Reduced inactivation of F2 (thrombin)	
	Protein C or S Deficiency	Reduced F5/8 inactivation, purpura fulminans w/ homozygous protein C def.	

Lab Changes by Disorder

	Platelet count	BT (NO LONGER PERFORMED)	PT	PTT
ITP	↓	↑	-	-
TTP-HUS	↓	↑	-	-
Hemophilia A/B	-	↑	-	↑
VWF Deficiency	-	↑	-	-/↑
DIC	↓	-	↑	↑
Vit K def/Warfarin	-	↑	↑	↑/-
End-stage Liver Disease	↓/-	↑/-	↑	↑

Hematologic Disorders of the Newborn/Child

	Pathogenesis	Clinical	Diagnosis	Treatment
Anemia of Prematurity	(1) Impaired EPO prod (2) Shortened RBC life (3) Iatrogenic blood loss	Asymptomatic Apnea, poor wt gain, tachycardia	Hemoglobin/Hct Reticulocyte count, Smear	Dec. phlebotomy Iron supplementation Transfusions
Transient Erythroblastopenia of Childhood	Acquired red cell aplasia (6 mo - 5 yo)	Gradual pallor, fatigue, etc.	Normocytic/chromic anemia, Hb (3-8), Reticulocyte count	Self-resolving

Hematologic Disorders continued on next page →