
HEMOSTASIS

by

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How to Approach to The Diagnosis of Bleeding Disorders

Evaluation of the patient

- History : age, sex, duration
- Physical Examination : site , type
- Laboratory Evaluation

History

- Surgical challenges
- Accidents & trauma injuries
- Dental extractions
- Menstrual history
- Spontaneous
- Family history
- Sites: Joints, scalp. Skin and mucous membrane

Type of Bleeding

- ecchymoses
- petechiae
- epistaxis
- deep soft tissue bleed
- hemarthroses
- GIT bleeding

Genetic cause ?

- Duration of bleeding history
- Onset
- Family history of relevant
- Age
- Sex

Medical History

- Liver disease: cirrhosis, jaundice, gall stone, hepatitis
- Renal disease : dialysis
- Malignancies and chemotherapy
- Antibiotic therapy, aspirin
- Poor nutrition (Vitamin K or C)
- Anticoagulants: warfarin, heparin,

Physical Examination

- current hemorrhage
  - nature and extent
- intercurrent illnesses
  - liver disease
- petechiae/ecchymoses

Laboratory Assessment

- Screening tests
  - BT, CBC
  - PT
  - aPTT
  - platelet count
  - Platelets function
  - fibrinogen
  - thrombin time

Specific Laboratory Tests

- Mixing studies
  - patient and normal plasma mixed 1:1
  - incubated 2 hours at 37° C
  - perform clotting assay as usual
- Uncorrected - circulating anticoagulant
- Corrected - factor deficiency

Circulating Anticoagulant

- Lupus anticoagulant/APA syndrome
  - rarely have associated bleeding
  - tend to thrombosis
- Acquired factor inhibitors
  - Factor VIII most common

Factor deficiencies

- Hemophilia A or B
  - Factor VIII or IX assays
  - Probably mild unless bleeding patient is an infant male
  - Send to Hemophilia Treatment Center
- von Willbrand's disease
  - most common genetic bleeding disorder
  - many different types

von Will brand's Disease

- autosomal dominant except Type III
- patients range from asymptomatic to spontaneous bleeding similar to a severe hemophiliac
- characterized by mucocutaneous bleeding

Lab Testing

- aPTT
- Factor VIII activity
- von Willebrand's Factor
- Ristocetin Cofactor
- von Willebrand's Factor multimers

von Willebrand's Disease

- Type I
  - normal molecule in abnormally low quantities
  - normal distribution of multimers
- Type II
  - abnormal molecule
  - abnormal distribution of multimers with decrease in the largest molecular weight forms
- Type III
  - Severe ,complete absence

Other Hereditary Defects

- Other Factor deficiencies
- Platelet defects
  - very rare
  - platelet aggregation studies
  - electron microscopy
  - bleeding time

Others ?

- Vitamin K deficiency
  - drug-induced/malabsorption
  - rarely nutritional in an outpatient
- Liver Disease
  - long PT +/- aPTT
  - poor clearance of coagulation products
- DIC

Liver Disease

- Decreased synthesis of factors
- Synthesis of abnormal factors
- Increased fibrinolysis
- Thrombocytopenia

Disseminated Intravascular Coagulation

- deficient factors
- platelets

Bone Marrow Diseases

- Acute leukemias
- Myelodysplasia
- Myeloproliferative disorders
  - Polycythemia RV
  - dysfunctional platelets