Bleeding Disorder

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Coagulation system

VASCULAR PHASE

PLATELET PHASE

PLASMA PHASE

Coagulation system

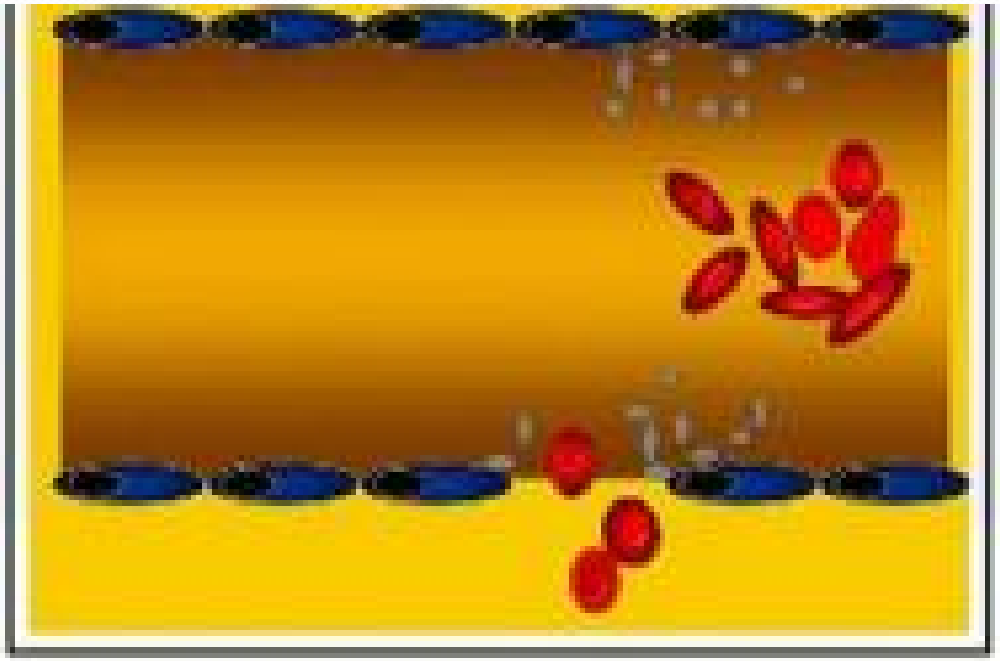
Blood vessel injury →Vasoconstriction

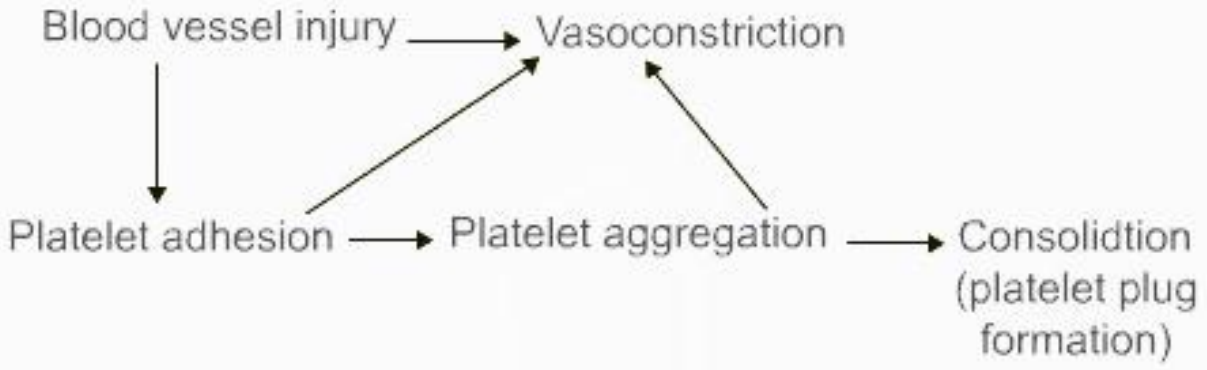
VASCULAR PHASE

PLATELET PHASE

PLASMA PHASE

Simplistic View of Vascular Injury

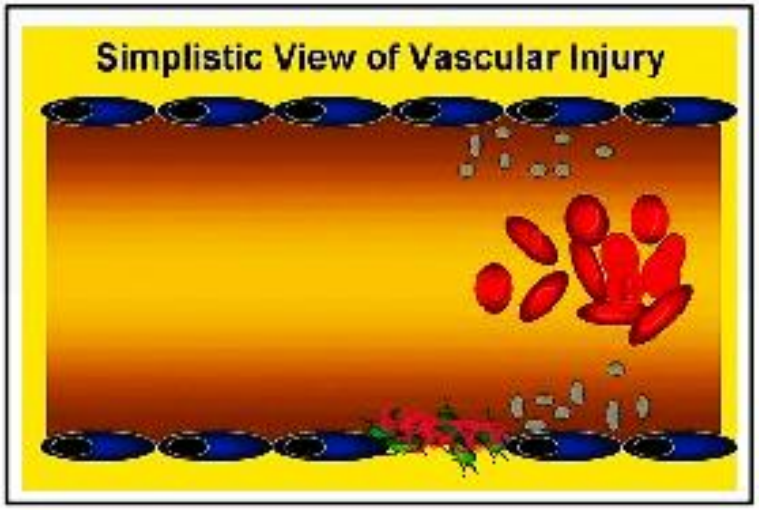


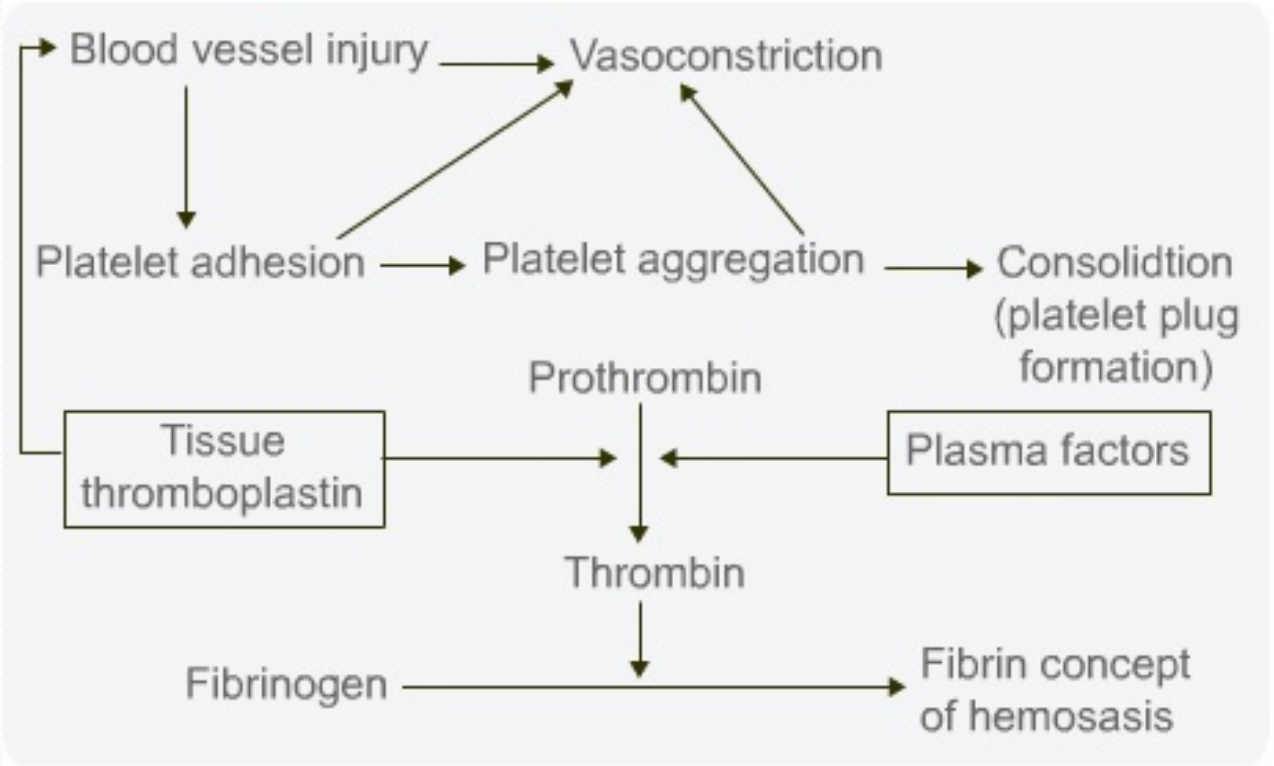


VASCULAR PHASE

PLATELET PHASE

PLASMA PHASE





VASCULAR PHASE

PLATELET PHASE

PLASMA PHASE

Practical Approach to a Child With BLEEDING HISTORY:

- AGE OF ONSET
- SEX, FAMILY HISTORY
- LOCATION &TYPE OF BLEEDING
- DURATION OF BLEEDING
- MEDICATIONS
- ASSOCIAED SYMPOMS
- REVIEW OF SYSTEMS

Detailed history (Symptoms)

Epistaxis, gingival bleeding , easy bruising, menorrhagia, hematuria, GI bleeding, hemarthrosis , prolonged bleeding after lacerations

Response to hemostatic challenge

Circum cision, surgery, phlebotomy,

immunization/intramuscular injection, suture placement/removal, dental procedures

Medications

Antiplatelet drugs (nonsteroidal anti-inflammatory drugs)

Anti-coagulants (Warfarin, heparin, low-molecular-weight heparin)

Family history

Symptoms, response to hemostatic challenge (siblings, parents,aunts, uncles, grandparents), transfusions after surgeries.

Clinical Characteristic	Primary Hemostatic Defect	Clotting Factor Deficiency
Site of bleeding	Skin, mucous membran	Soft tissues, muscles, joints
Bleeding after minor cuts	Yes	Not usually
Petechiae	Present	Absent
Ecchymosis	Small, superficial	Larger, deeper, palpable
Hemarthrosis	Rare	Common
Bleeding after trauma/surgery	Immediate	Delayed

Complete blood count (CBC)

Quantitative assessment of platelets

Etiology

1:Consumption Syndromes
2: Decreased Production
3:Sequestration

Primary Platelet Consumption Syndromes

Immune thrombocytopenias

Non immune thrombocytopenias

Immune

- ITP
- Autoimmune diseases(SLE, Evans)
- Neoplasia
- Infection(HIV)
- Drug-induced (including HIT)

Non Immune

- Infection(Bacteremia ,fungemia,Viral ,Protozoa)
- Thrombotic microangiopathic disorders(TTP, HUS)
- Drug-induced via direct platelet effects (protamine)
- Type 2b VWD or platelet-type VWD

Decreased Platelet Production

- Hereditary disorders
- Acquired disorders

Hereditary disorders (Aplastic anemia Congenital)

Congenital amegakaryocytic thromboytopenia(CAMT)

Anemia Fanconia

Decreased Platelet Production

- Acquired aplastic anemia
- - Drug(chemotherapy , valproic acid)
- -Radiation

-Infection

- -Infiltration by abnormal cells (Leukemia, solid tumors, osteopetrosis, storage diseases)

qualiative assessment of platelets

- Hereditary disorders
- Acquired disorders

Bleeding time

A normal BT is 3 to 9 minutes.
The BT is an approximate measure of the relationship between platelet number and function, as originally demonstrated.

Prolonged Bleeding time

- Impaired platelet function
- Platelet counts reduced below 80,000
- Impaired vascular integrity

Prolonged Bleeding time

The BT can also be prolonged with congenital and acquired platelet function defects ,certain drugs, uremia, VWD, vasculitis (e.g., Henoch-Schonlein purpura),and connective tissue disorders such as Ehlers-Danlos syndrome.

Platelet Function Analyzer (PFA 100)

Often prolonged with impaired platelet function

Factor Disorder

Extrinsic system
Intrinsic system
Common system
FXIII
Fibrinolysis system

Prothrombin time (PT)

(Assesses extrinsic system)

FVII

PTT

(Assesses intrinsic system)

FXII(HMK,PK), FXI, FIX and FVIII assays

Common confirmatory coagulation tests

FX, FV, FII and fibrinogen

Thrombin Time

The TT measures the thrombin-induced conversion of fibrinogen to fibrin
Useful test to diagnose when both the PT and PTT are prolonged.
Prolonged when fibrinogen is reduced or abnormal

- Presence of inhibitors (fibrin degradation products, D-dimers)

FXIII deficiency & increased fibrinolysis

In a patient with a significant bleeding history, if all screening tests and von Willebrandpanel are normal consider FXIII, PAI activity, alpha 2 antiplasmin

FXIII deficiency

Urea Clot Lysis Assay: Useful screen for FXIII deficiency outside of the newborn period.

FXIII Assay

Euglobulin Clot Lysis Time

Shortened in conditions characterized by increased fibrinolysis (antiplasmin deficiency, plasminogen activator inhibitor 1 deficiency.

Reptilase time

Reptilase, a snake venom protease, clots fibrinogen in the presence of heparin and thus can be used to identify heparin as the cause of a prolonged TT. Thus, in the presence of heparin theTT is prolonged, whereas the reptilase time is normal.

Mixing studies

Mixing studies (performed to evaluate a prolonged PT or PTT) Continued prolongation indicates presence of a coagulation inhibitor.

Vascular Disorder

Hereditary disorders(Ehlers-Danlos syndrome)
Acquired disorders (Henoch-Schonlein purpura)

Evaluation VWD

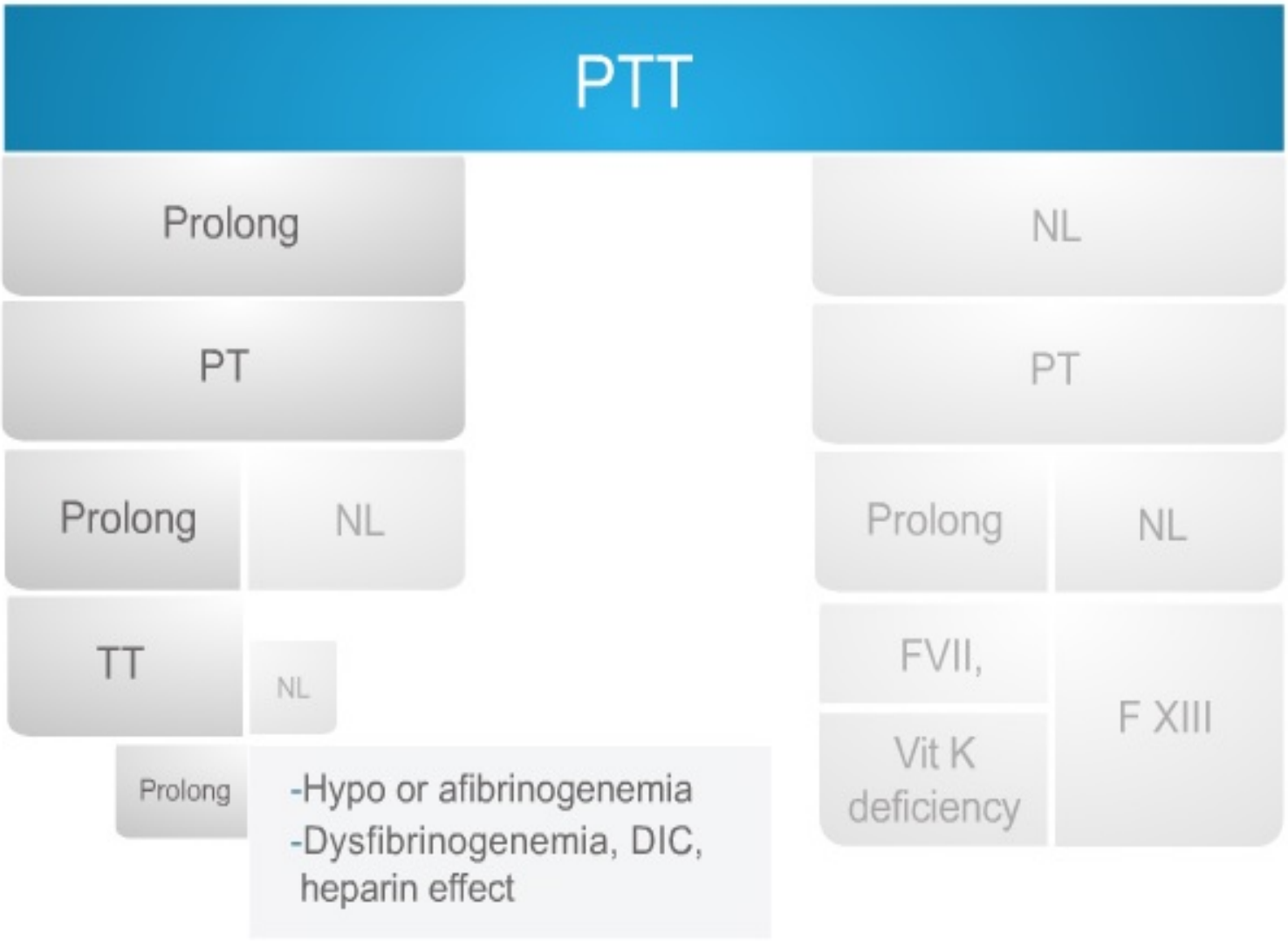
Recommended tests for VWD include :
APTT, factor VIII assay, ristocetin cofactor assay as a measure of VWF activity, VWF antigen assay, ABO blood group typing.

Underlying disorder

It is important to note whether the patient has underlying medical disorder that may affect hemostasis, such as hepatic or renal disease, malabsorption syndrome.

Laboratory approach to 2° H. defect

Laboratory approach to 2° H. defect


PTT
Prolong
PT
Prolong
NL
TT
NL
Prolong
-Hypo or afibrinogenemia
-Dysfibrinogenemia, DIC, heparin effect
NL
PT
Prolong
NL
FVII,
Vit K deficiency
F XIII

Thanks For Attention