



# Haemostasis and Bleeding Disorders

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# Objectives

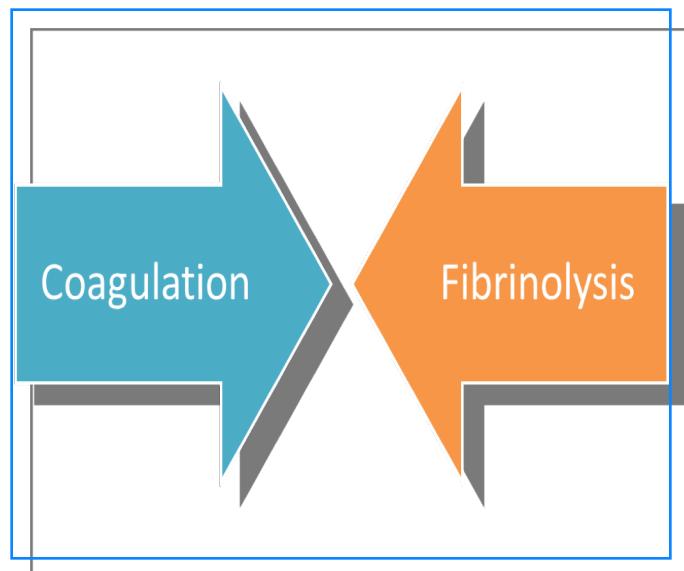
- What is haemostasis?
- Components of haemostasis
- What are bleeding disorders?
- How they could be presented clinically?

# What is Haemostasis?

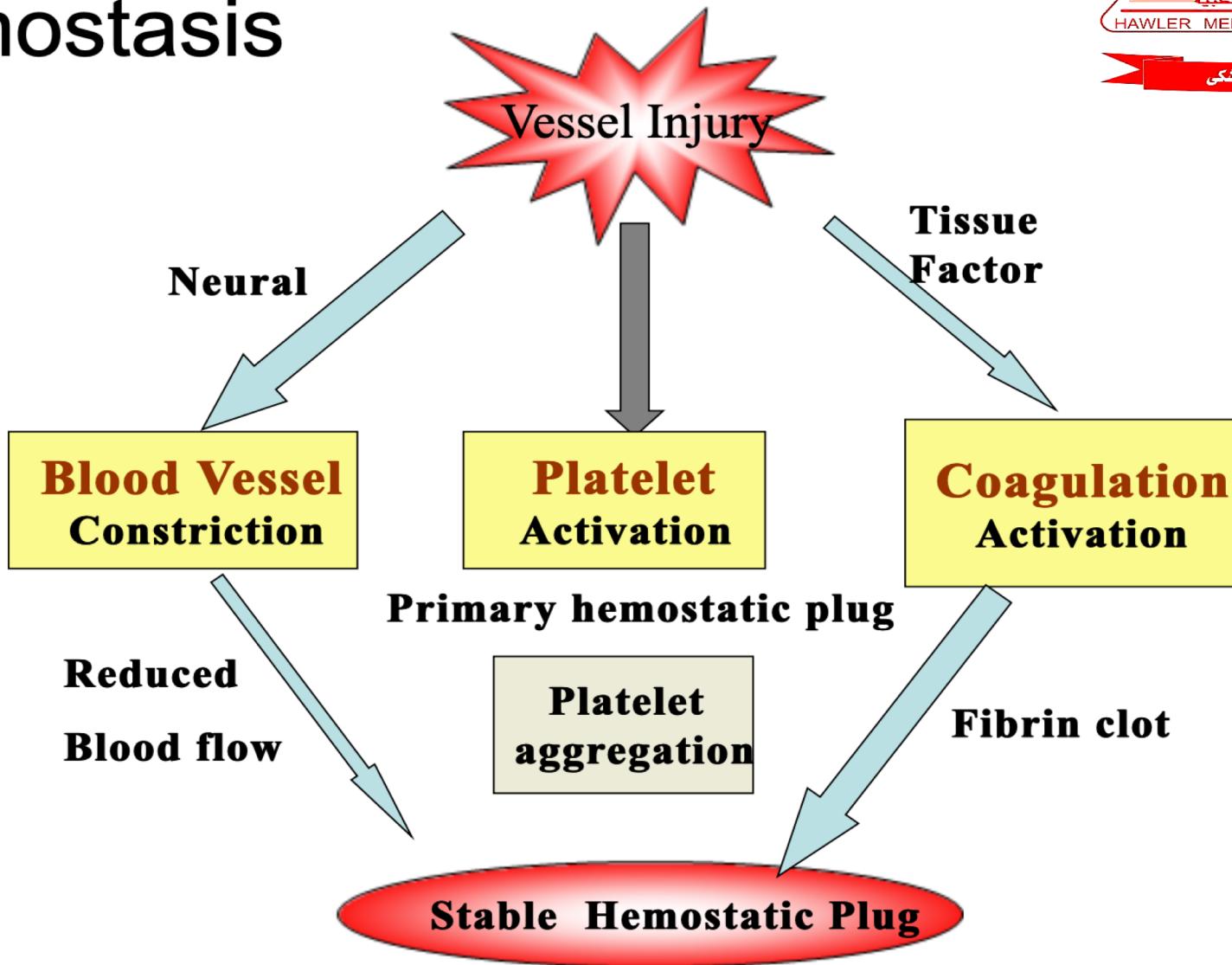
- It is the normal physiological response that prevents significant blood loss after vascular injury. It serves to maintain the integrity of the circulatory system.
- However the process can become imbalanced, leading to significant morbidity and mortality.

## Components of Haemostasis System:

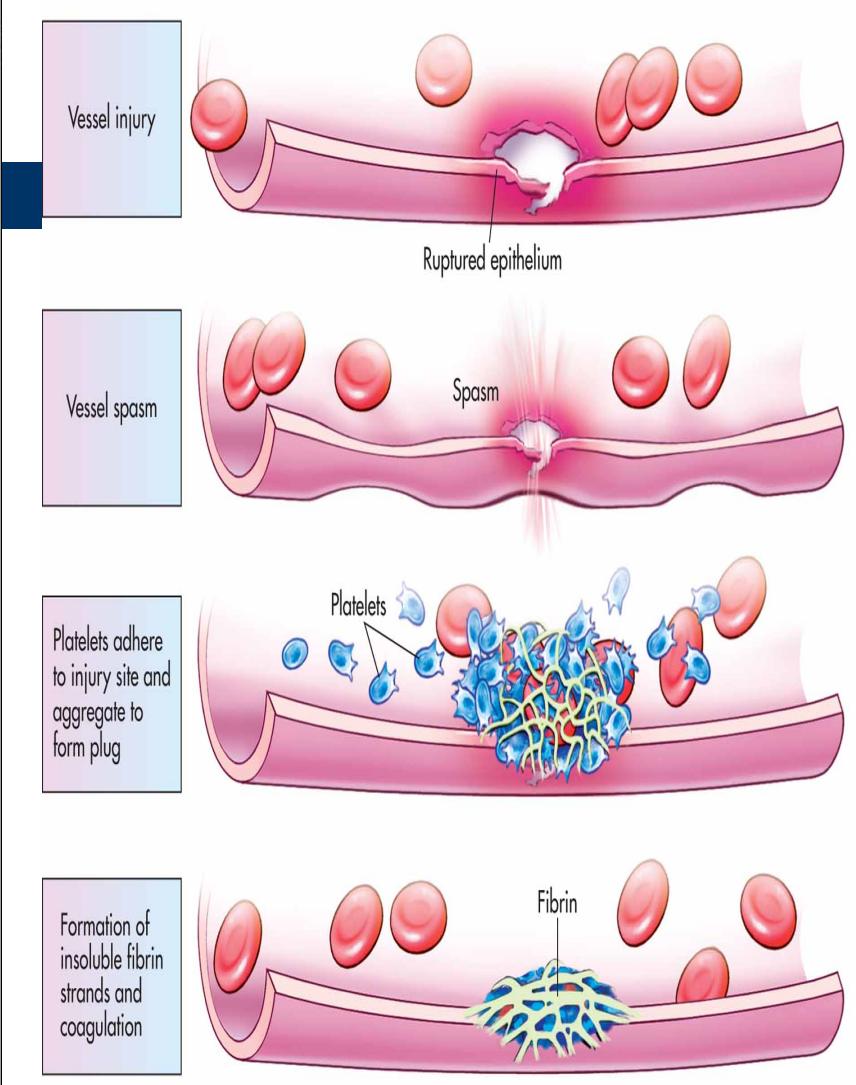
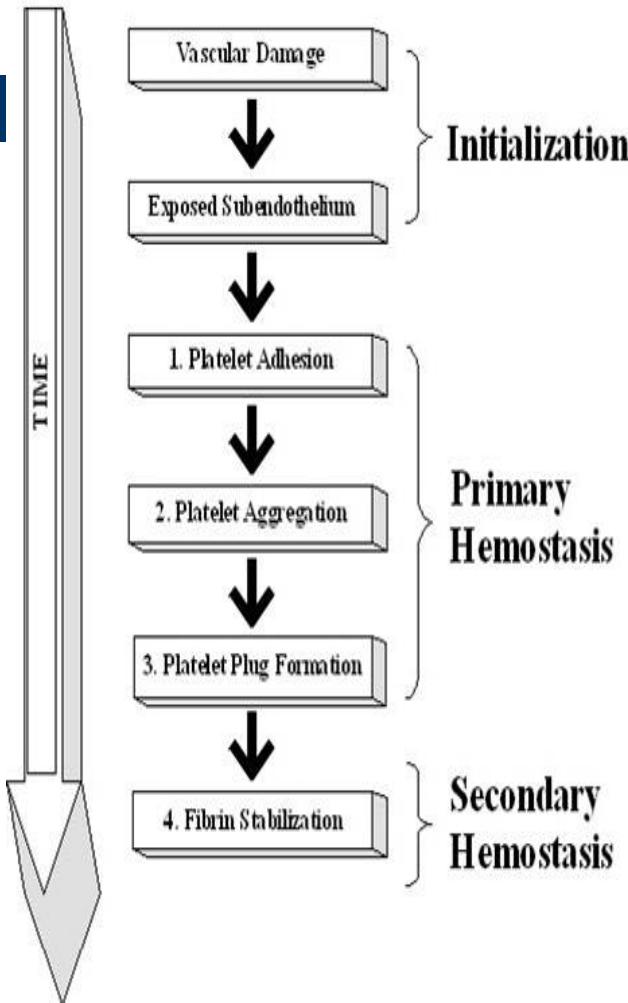
- Blood vessel
- Platelets
- Clotting factors
- Coagulation inhibitors:  
(protein C, protein S,  
antithrombin)
- Fibrinolytic system:  
(plasminogen, plasmin)



# Hemostasis

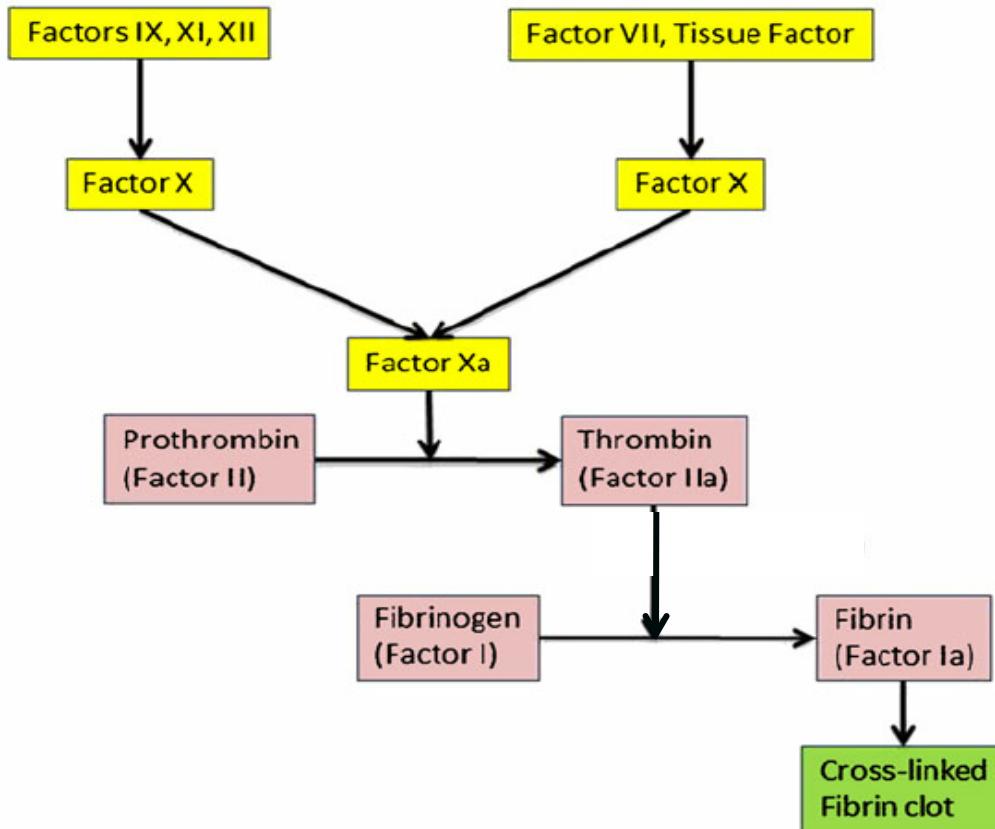


# Overview of Hemostatic Mechanisms



# Coagulation pathway

Intrinsic (contact activation) Pathway      Extrinsic (tissue factor) Pathway



- What are the Causes of prolonged aPTT ?

# Causes of prolonged aPTT

- Congenital deficiencies of intrinsic system clotting factors such as factors VIII, IX, XI, and XII, including hemophilia A and hemophilia B (Christmas disease).
- Von Willebrand disease
- Hypofibrinogenemia
- Liver disease
- Vitamin K defecincy
- DIC
- Heparin therapy,

- What are the Causes of prolonged PT ?

# Causes of prolonged PT:

- Warfarin use
- Vitamin K deficiency from malnutrition, biliary obstruction, malabsorption syndromes, or use of antibiotics
- Liver disease
- Deficiency or presence of an inhibitor to factors VII, X, II/prothrombin, V, or fibrinogen
- Disseminated intravascular coagulopathy (DIC)
- Fibrinogen abnormality (eg, hypofibrinogenemia, afibrinogenemia, dysfibrinogenemia)
- Massive blood transfusion due to dilution of plasma clotting proteins

# What are the bleeding disorders?

- They are a group of disorders that share the inability to form a proper blood clot.
- They are characterized by prolonged bleeding after surgery, trauma, menstruation and sometimes it may be spontaneous.
- The defect either from platelets, clotting factors or from blood vessels; they may be inherited or acquired.

# Simplified WHO grading of bleeding

Grade	Signs
0	No bleeding
1	Petechia, ecchymosis, occult blood in body secretions, and mild vaginal spotting
2	Gross bleeding not requiring RBC transfusions (eg, epistaxis, hematuria, and hematemesis)
3	Bleeding requiring RBC transfusions
4	Life-threatening bleeding

Note: Data from Slichter.<sup>45</sup>

Abbreviations: RBC, red blood cell; WHO, World Health Organization.

# Platelet Disorders

## Thrombocytopenia:

- Congenital: e.g. Wiscott-Aldrich syndrome
- Impaired bone marrow production (aplastic anemia, bone marrow infiltration like leukemia).
- Increased platelet destruction: Immune thrombocytopenia
- Excess sequestration in spleen: hypersplenism

## Disorders of Platelet Function:

- Inherited: Glanzmanns thrombasthenia
- Acquired: aspirin, uremia.

# Disorders of Clotting Factors

- *Inherited:* Hemophilia, Von Willebrand disease and congenital fibrinogen deficiency.
- *Acquired:* liver disease, vitamin K deficiency, disseminated intravascular coagulopathy and anticoagulant therapy.

# Disorders of blood vessels

- Hereditary Haemorrhagic Telangiectasia (HHT)
- Connective tissue disorders: like Marfan's syndrome
- Senile purpura
- Henoch-Schönlein purpura
- Vitamin C deficiency (Scurvy)
- Steroid

# Symptoms:

- Common bleeding symptoms
  - Nose bleeds
  - Skin bruising
  - Gingival bleeding
  - Bleeding from small wounds
  - Menorrhagia
  - Postpartum bleeding

# Symptoms.....

- Postoperative and invasive procedures
  - Dental extractions
- Other bleeding symptoms
  - Gastrointestinal bleeding
  - Urinary bleeding
  - Hematomas
  - Hemarthroses
  - Hemoptysis
  - Central nervous system bleeding

# History taking:

- **Who:** who is the patient, age, sex, race and family history?
- **When:** when did the bleeding occur (onset of bleeding)? Is it related to drug ingestion or any underlying disorder? did it develop after surgery or trauma?
- **Where:** sites of bleeding, skin, muscle etc.
- **What:** description of the type of bleeding.

# Physical Examination

- Identifying petechiae, purpura, ecchymoses and hematomas.
- Petechiae: is a small (1–2 mm) red or purple spot on the skin.
- Purpura: red or purple discolored spots on the skin that do not blanch on applying pressure, they measure (3–10 mm).
- Ecchymoses: is a subcutaneous spot of bleeding with diameter larger than 1 cm.

# Physical Examination.....

- Focus on findings that may suggest the underling causes of bleeding such as evidence of liver disease (jaundice, splenomegaly), telangiectasia (eg, hereditary hemorrhagic telangiectasia).
- Signs of anemia
- Vital signs (BP, PR, RR, Temp.)

## Case 1

- A 40 year old female with no comorbidities noticed the sudden appearance of multiple bruises on her extremities and mild epistaxis. She had no other symptoms and denied taking any medications.
- CBC identified a platelet count of  $30 \times 10^9/L$ , an extensive work-up revealing no other significant abnormalities.
- A bone marrow aspiration and biopsy was unremarkable, apart from numerous megakaryocytes.

# Immune thrombocytopenia (ITP)

- It is a bleeding disorder caused by thrombocytopenia not associated with a systemic disease.
- Typically, it is chronic in adults, but it is usually acute and self-limited in children.
- Spleen size is normal in the absence of another underlying condition.
- Diagnosis requires that other disorders be excluded through selective tests.

## Case 2

- A 20 year old male presents with bruising and severe pain in the right arm after a heavy work yesterday. His history is notable for prior excess bleeding during tooth extraction and recurrent bleeding in knee joint.

CBP	Normal
Bleeding time	3 min.
PT	11 sec.
PTT	60 sec.

- What further points in the history should be focused on?
- What is the diagnosis?

# Haemophilia

- X-linked recessive, It is a male disease, extremely rarely affect female.
- Up to 33% of patients have no family history, (i.e. spontaneous mutation).
- The genetic mutation will cause absence or low level factor VIII (haemophilia A) or factor IX (haemophilia B).
- Incidence ~ 30-100/1.000.000
- May be mild, moderate or severe.
- The main problem is intra-articular bleeding.

## Case 3

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- A 20 year old female presented with fever and many skin bruises over the last 5 days.
- CBP showed Hb 7g/dl, WBC  $2 \times 10^9/L$  and platelets  $20 \times 10^9/L$ . Bone marrow aspiration was very hypocellular.

## Case 4

- A 28 year old woman presents with fatigue for more than 2 months. She has lifelong recurrent epistaxis. Her father also had repeated epistaxis. On examination she was pale and there were multiple blue-red lesions involving the lips and nostrils.

Hb	7 g/dl
MCV	64 fl
MCH	24 pg
WBC	$6 \times 10^9/L$
Platelets	$410 \times 10^9/L$
PT	Normal
PTT	Normal

# Hereditary Hemorrhagic Telangiectasia

- An autosomal dominant disorder
- Telangiectasia (skin, mucous membrane and internal organs)
- Pulmonary and cerebral arterio-venous malformation in minority
- Recurrent epistaxis, gastrointestinal bleeding, anemia

# Acute Leukemia

- They are malignant diseases arising from haemopoietic stem cells or early progenitors
- Characterized by the appearance of increased number of immature cells in the bone marrow and blood
- There is bone marrow infiltration (anemia, neutropenia and thrombocytopenia), and organ infiltration (spleen, liver and brain).
- Coagulation may be disturbed (always check coagulation screen).
- The main 2 types are acute myeloid leukemia (AML) and acute lymphoblastic leukemia (ALL).

# Von Willebrand disease (VWD)

- Von Willebrand factor performs two main function: mediate platelet-vessel wall adhesion (& aggregation) and it acts as a carrier for factor VIII
- VWD is the most common inherited bleeding disorder
- Affecting up to 1% of the population.
- Most cases are autosomal dominant
- It affects male and female equally.
- Type I and III are quantitative while type II is a qualitative defect

# Vitamin K

- Vitamin K is needed for the synthesis of factors II, VII, IX and X.
- Vitamin K is vital to the carboxylation of glutamic acid residues which is needed for the activation of these factors.
- The most common circumstance in which vitamin K deficiency leads to bleeding are: hemorrhagic disease of the newborn, nutritional deficits, malabsorption or alteration in intestinal flora.
- Treatment must be directed at the underlying disorder and vitamin K supplementation.



# Qs



*Thank You*