Approach to bleeding disorders

Dr Nahid Raeissi
Ped. Hematologist & Oncologist
Isfahan University of Medical Sciences

Normal hemostasis

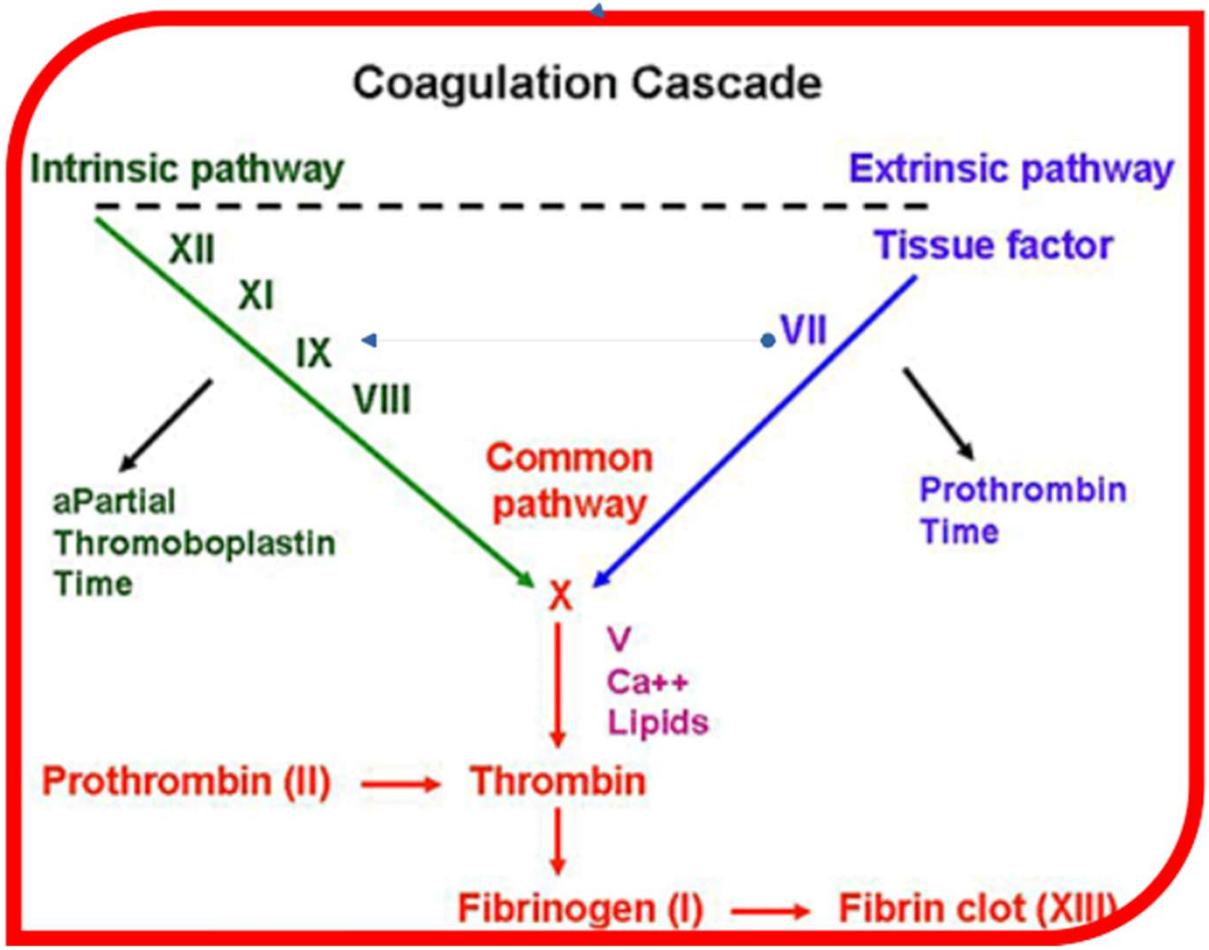
In health, hemostasis is the biologic process that limits hemorrhage after blood vessel injury.
If a vessel wall is damaged, a number of mechanisms are promptly activated to limit bleeding by a complex series of interrelated reactions involving endothelial cells, platelets, plasma coagulation factors, anticoagulant proteins and fibrinolytic proteins.

Primary hemostasis

- Primary hemostasis occurs after damage to the vessel wall, and involves vasoconstriction and adhesion of platelets in a monolayer on exposed subendothelial fibrils. Subsequently, further platelets aggregate to form a platelet plug, which .stems the flow of blood

Secondary hemostasis

- ***Secondary hemostasis*** ***involves activation of the coagulation system, leading to the generation of fibrin strands, which are laid down between platelets and .reinforce the platelet plug***


Coagulation Cascade
Intrinsic pathway
Extrinsic pathway
XII
XI
IX
VIII
Tissue factor
VII
aPartial
Thromoboplastin
Time
Common
pathway
Prothrombin
Time
X
V
Ca++
Lipids
Prothrombin (II)
Thrombin
Fibrinogen (I)
Fibrin clot (XIII)

Assessment of bleeding symptoms

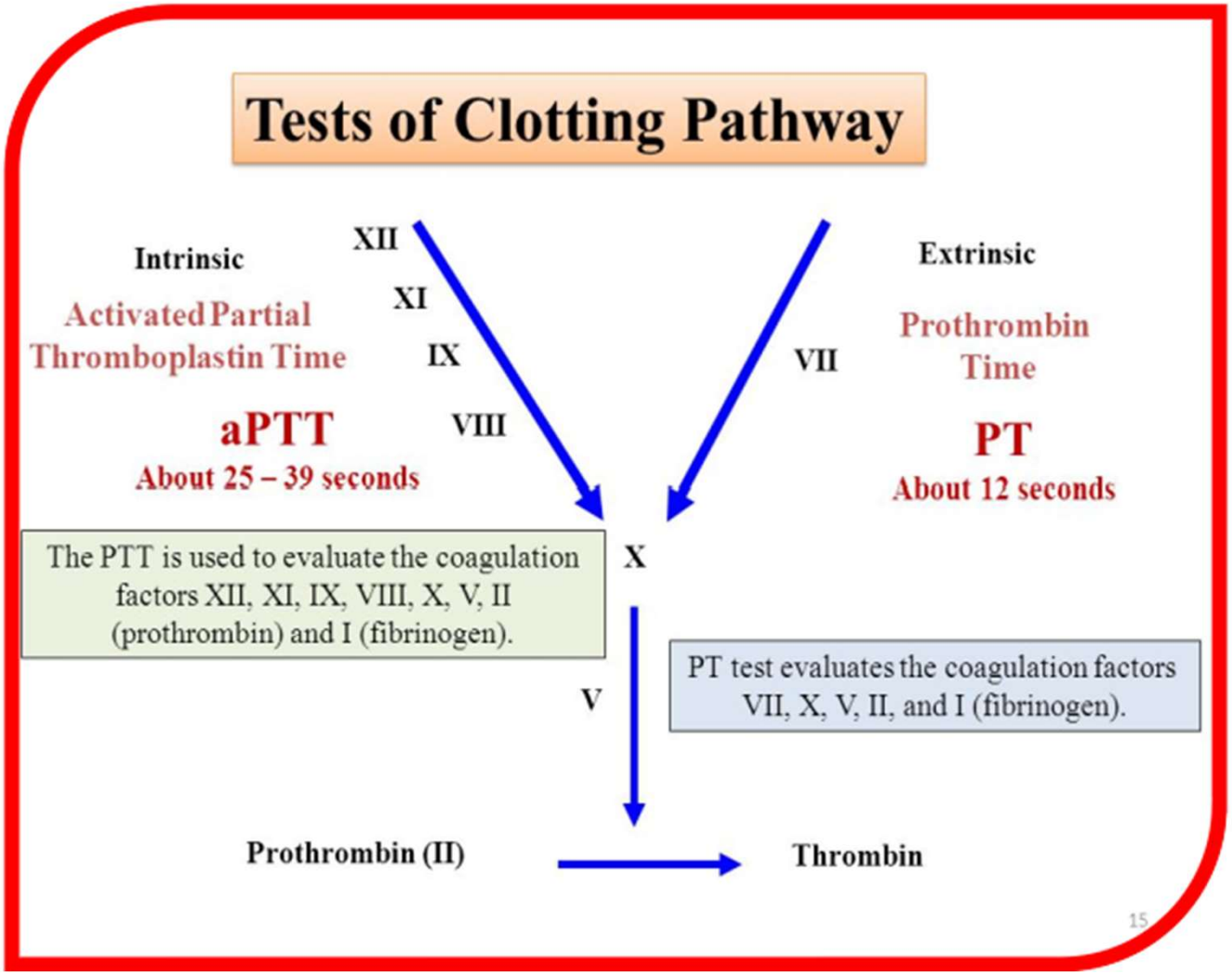
The assessment of an individual patient is a two-stage process:

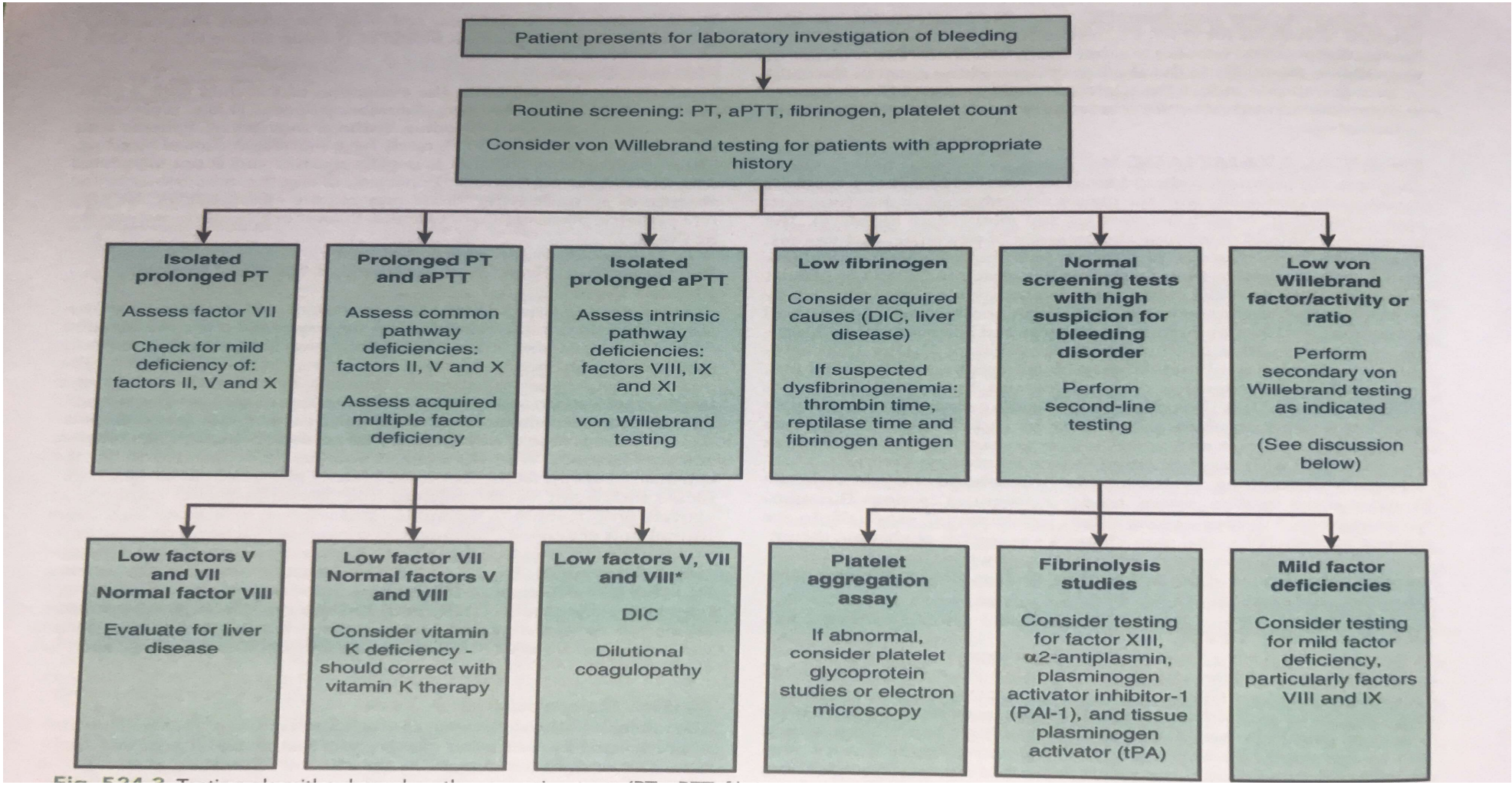
1. History (Patient & family history, Drugs, Systemic illness) and focused clinical examination; and
2. Laboratory investigation

Kind of haemorrhage	Primary haemostasis	Secondary haemostasis
Petechiae- Purpura	++	--
Hemarthrosis	--	++
Ecchymosis - Hematoma	+	+
Beginning of haemorrhage after trauma	Immediate	Late
Mucosal bleeding	Spontaneous	With trauma
Local pressure	Effective	Non effective

Kind of haemorrhage	Primary haemostasis	Secondary haemostasis
Petechiae- Purpura	++	--
Hemarthrosis	--	++
Ecchymosis - Hematoma	+	+
Beginning of haemorrhage after trauma	Immediate	Late
Mucosal bleeding	Spontaneous	With trauma
Local pressure	Effective	Non effective


Petechiae
Purpura


Tests of Clotting Pathway
Intrinsic
Activated Partial Thromboplastin Time
aPTT
About 25 – 39 seconds
XII
XI
IX
VIII
VII
Extrinsic
Prothrombin Time
PT
About 12 seconds
The PTT is used to evaluate the coagulation factors XII, XI, IX, VIII, X, V, II (prothrombin) and I (fibrinogen).
X
V
PT test evaluates the coagulation factors VII, X, V, II, and I (fibrinogen).
Prothrombin (II)
Thrombin
15


Patient presents for laboratory investigation of bleeding
Routine screening: PT, aPTT, fibrinogen, platelet count
Consider von Willebrand testing for patients with appropriate history
Isolated prolonged PT
Assess factor VII
Check for mild deficiency of: factors II, V and X
Prolonged PT and aPTT
Assess common pathway deficiencies: factors II, V and X
Assess acquired multiple factor deficiency
Isolated prolonged aPTT
Assess intrinsic pathway deficiencies: factors VIII, IX and XI
von Willebrand testing
Low fibrinogen
Consider acquired causes (DIC, liver disease)
If suspected dysfibrinogenemia: thrombin time, reptilase time and fibrinogen antigen
Normal screening tests with high suspicion for bleeding disorder
Perform second-line testing
Low von Willebrand factor/activity or ratio
Perform secondary von Willebrand testing as indicated
(See discussion below)
Low factors V and VII Normal factor VIII
Evaluate for liver disease
Low factor VII Normal factors V and VIII
Consider vitamin K deficiency - should correct with vitamin K therapy
Low factors V, VII and VIII*
DIC
Dilutional coagulopathy
Platelet aggregation assay
If abnormal, consider platelet glycoprotein studies or electron microscopy
Fibrinolysis studies
Consider testing for factor XIII, α2-antiplasmin, plasminogen activator inhibitor-1 (PAI-1), and tissue plasminogen activator (tPA)
Mild factor deficiencies
Consider testing for mild factor deficiency, particularly factors VIII and IX

Click to add Title

- **Thank you for your attention**