Introduction to Coagulation and its Disorders

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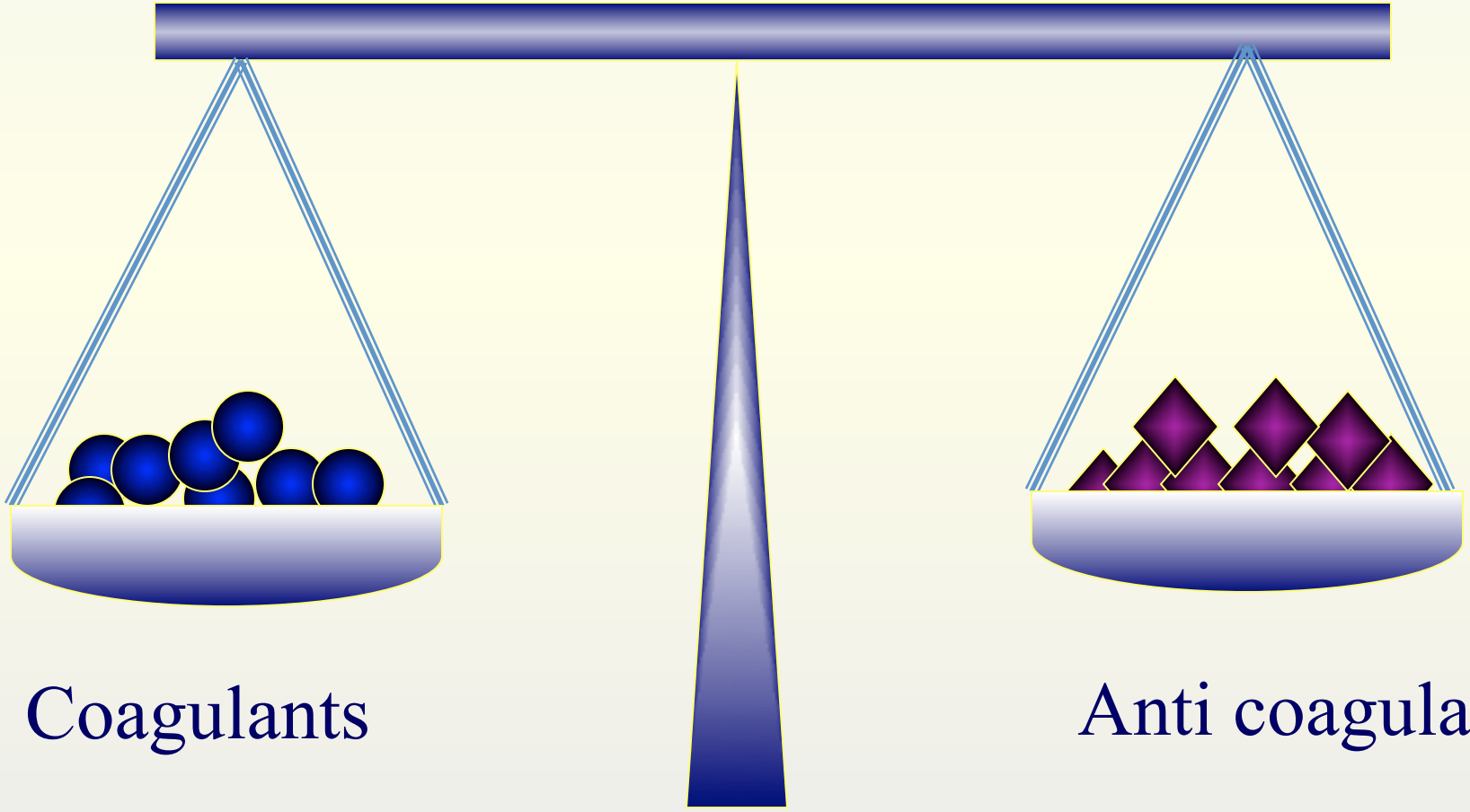
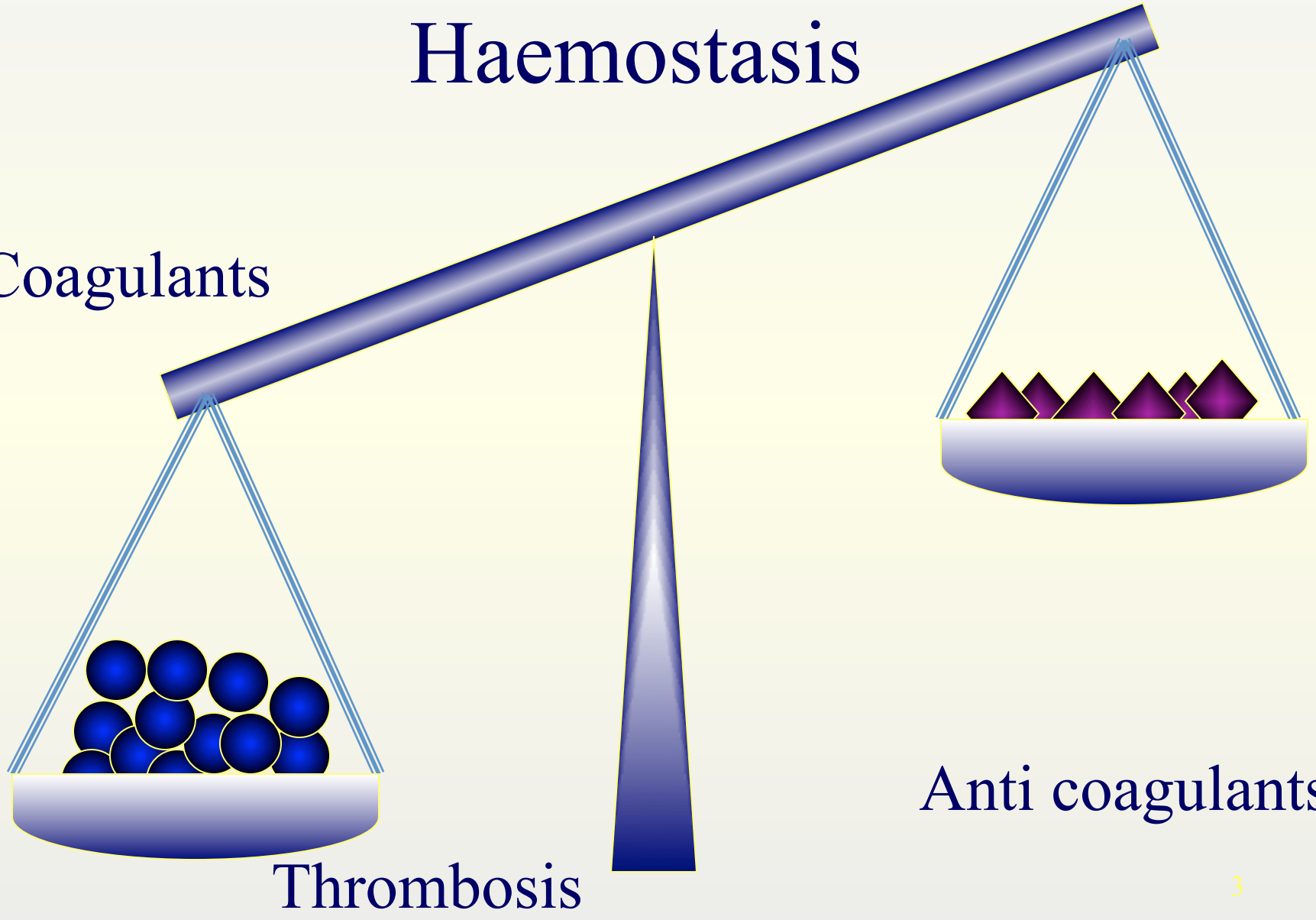
Haemostasis



Coagulants

Anti coagulants

Haemostasis



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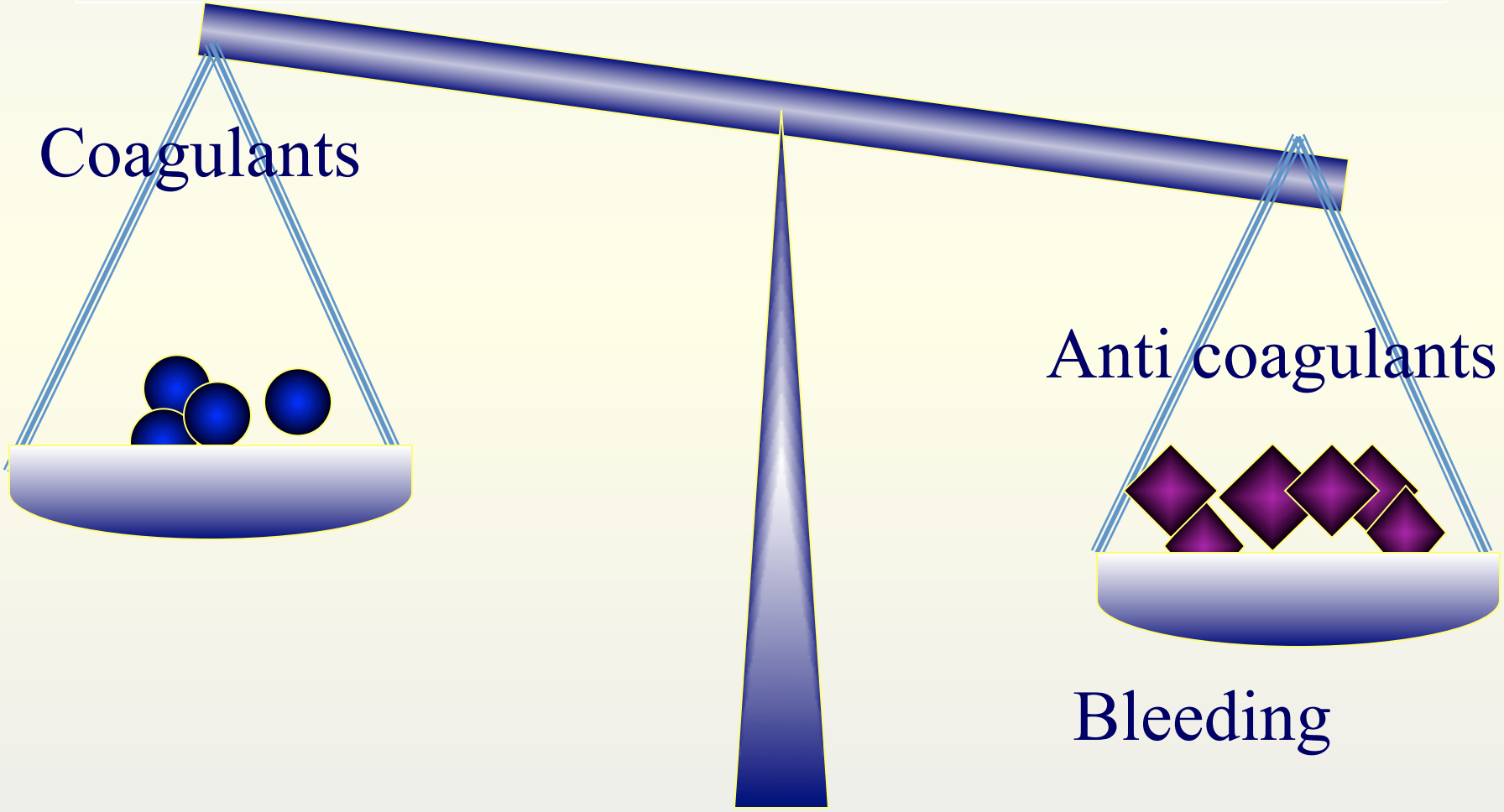

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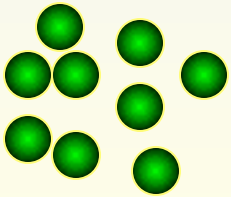


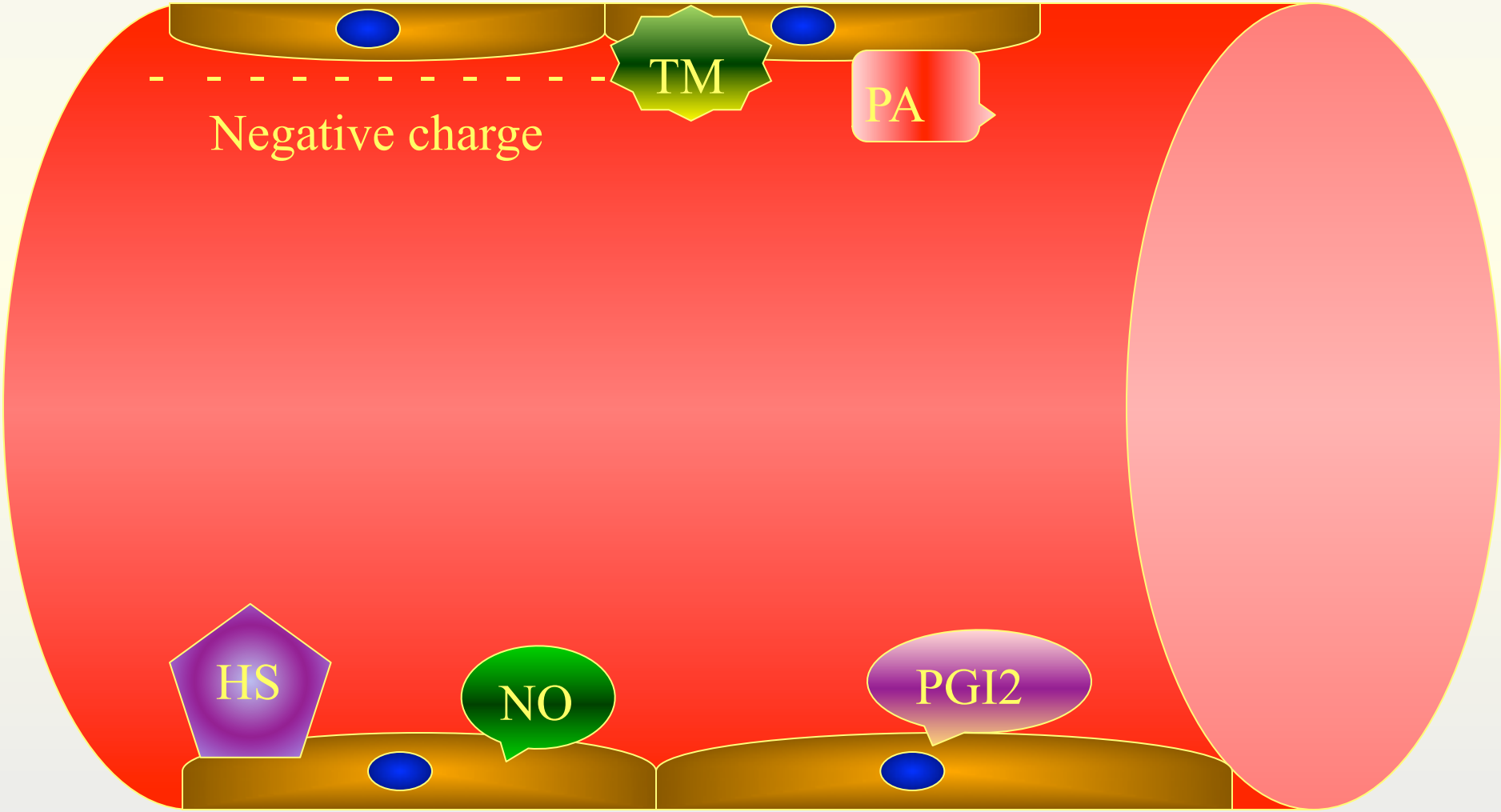
Platelets - Thrombocytopaenia
Functional defect

Vessels - Vascular defect

Coagulation system — Deficiency / Defect

Fibrinolytic system — Deficiency / Defect

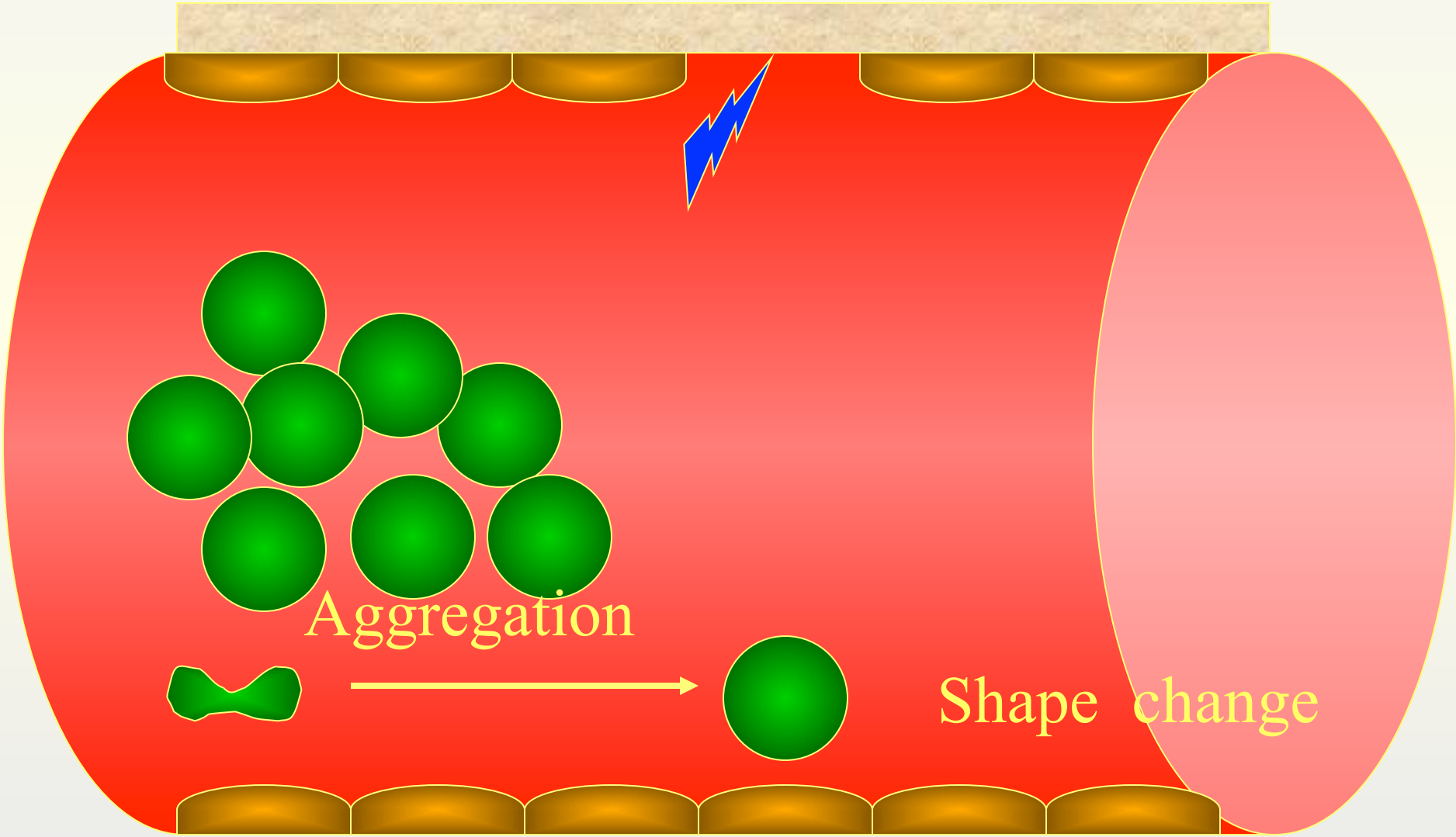
Endothelial cells



Haemostasis

Vasoconstriction

Endothelial Injury



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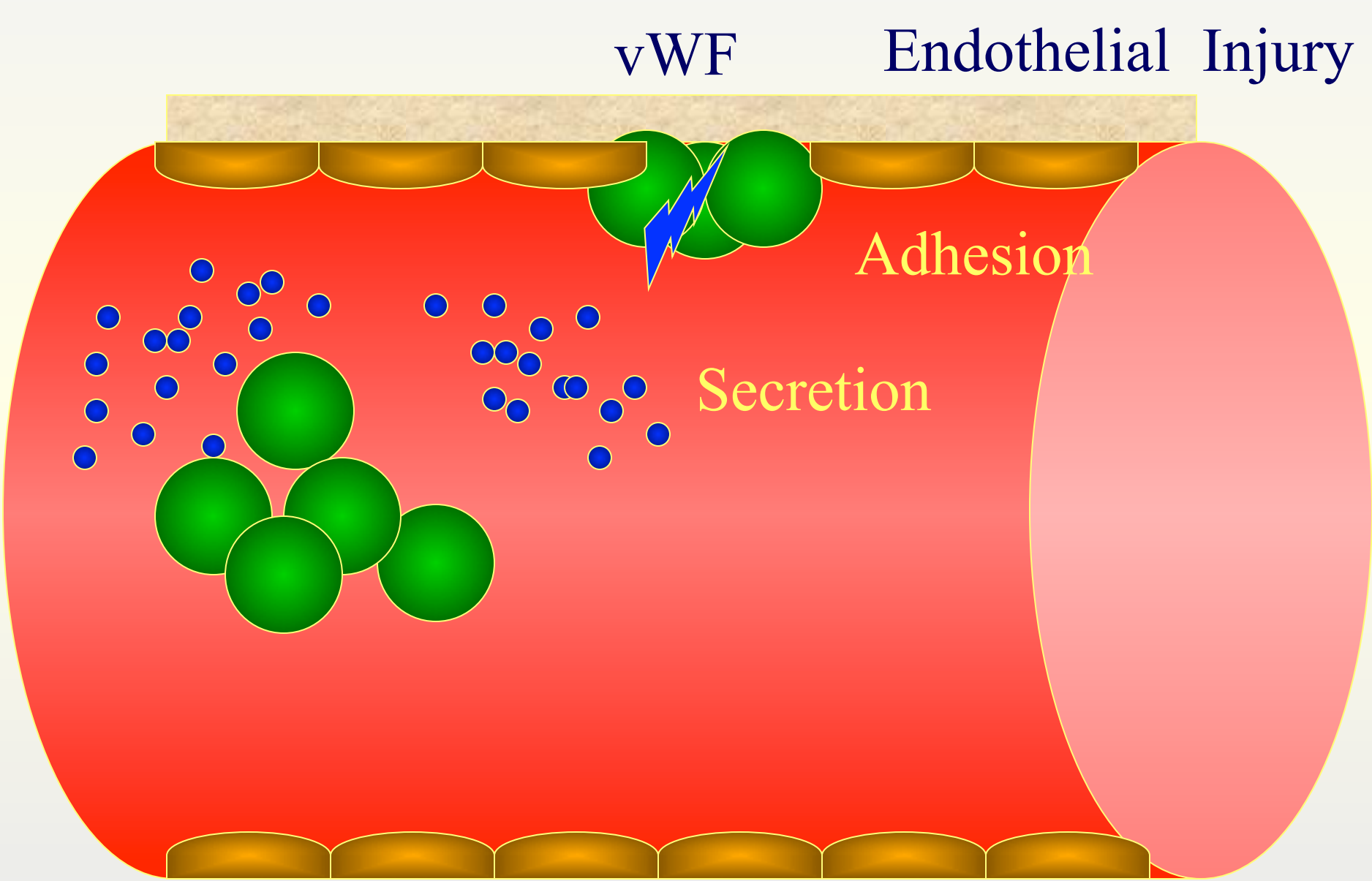
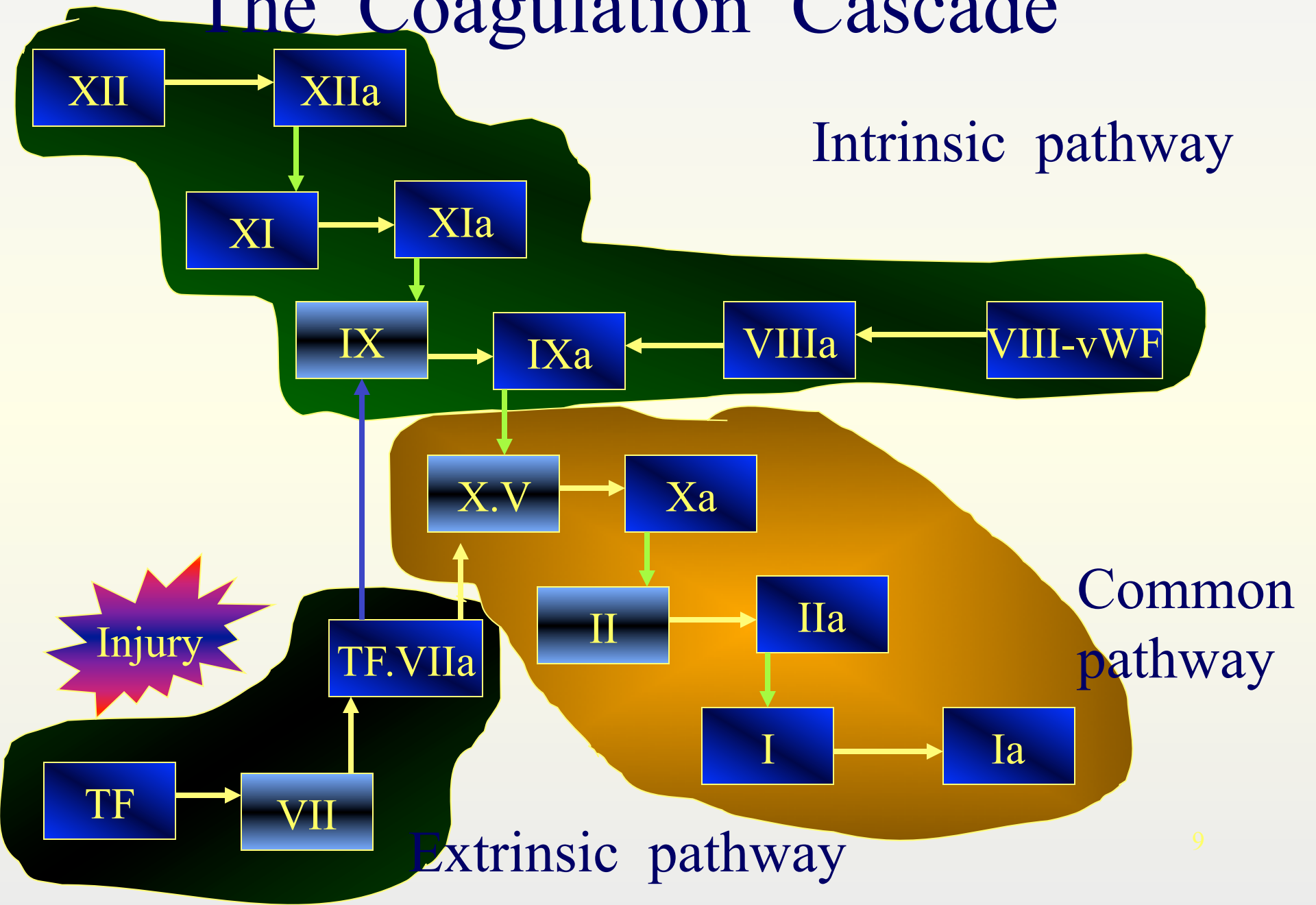


The Coagulation Cascade



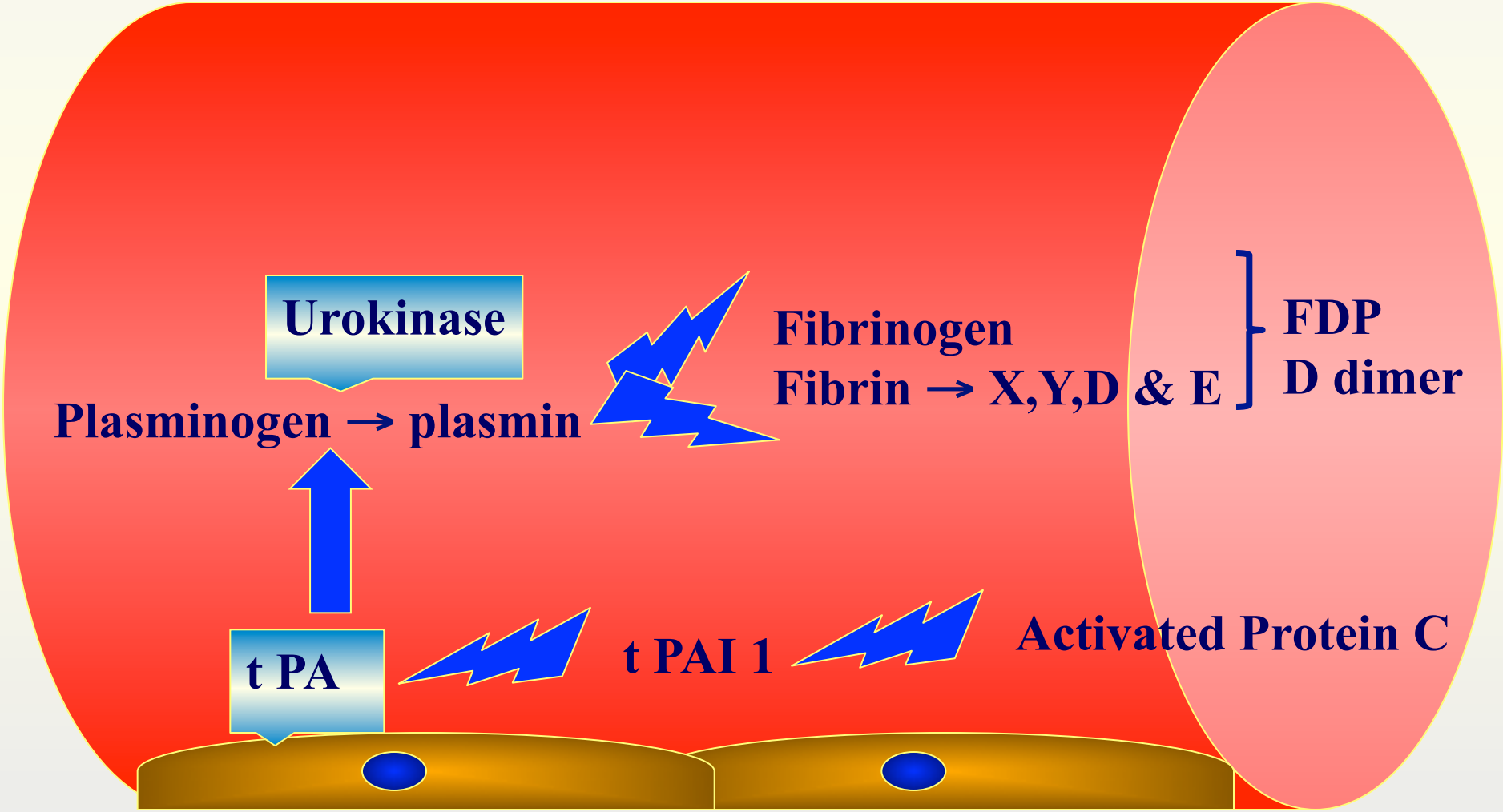
Physiological limitation of coagulation - Natural anticoagulants

- Antithrombin
- Activated protein C - inactivates factor V and factor VIII
- Protein S - enhances binding of activated protein C to phospholipid surface.
- Bound to C4b binding protein b

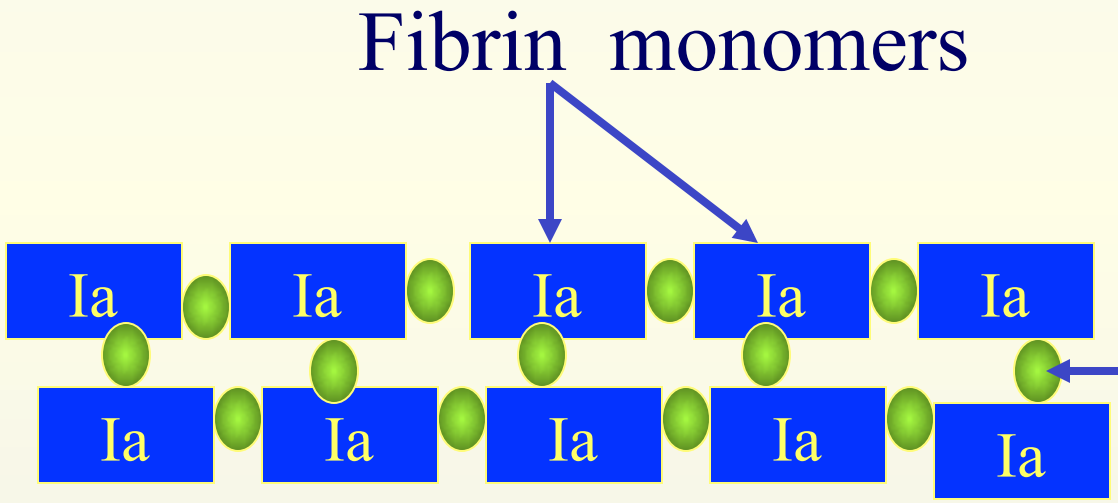
Other inhibitors

- α2 macroglobulin
- α1-antitrypsin
- α2-antiplasmin.

Fibrinolysis



Coagulation



Factor XIII binds And stabilizes the fibrin clot

Investigation of bleeding disorders

- Is there a generalized haemostatic defect?
- Bleeding from multiple sites
- Spontaneous bleeding
- Excessive bleeding after injury.

Disseminated intra vascular coagulation

Investigation of bleeding disorders

- Is the defect inherited or acquired?
- Family history
- Severe inherited defects become apparent in infancy
- Mild inherited defects present later in life
- - Excessive bleeding after surgery
- - Childbirth
- - Dental extractions or trauma.

Investigation of a bleeding disorder

- Some defects are revealed by routine coagulation screens before surgical procedures.
- Is the bleeding of a
- Vascular
- platelet defect or
- Coagulation defect?

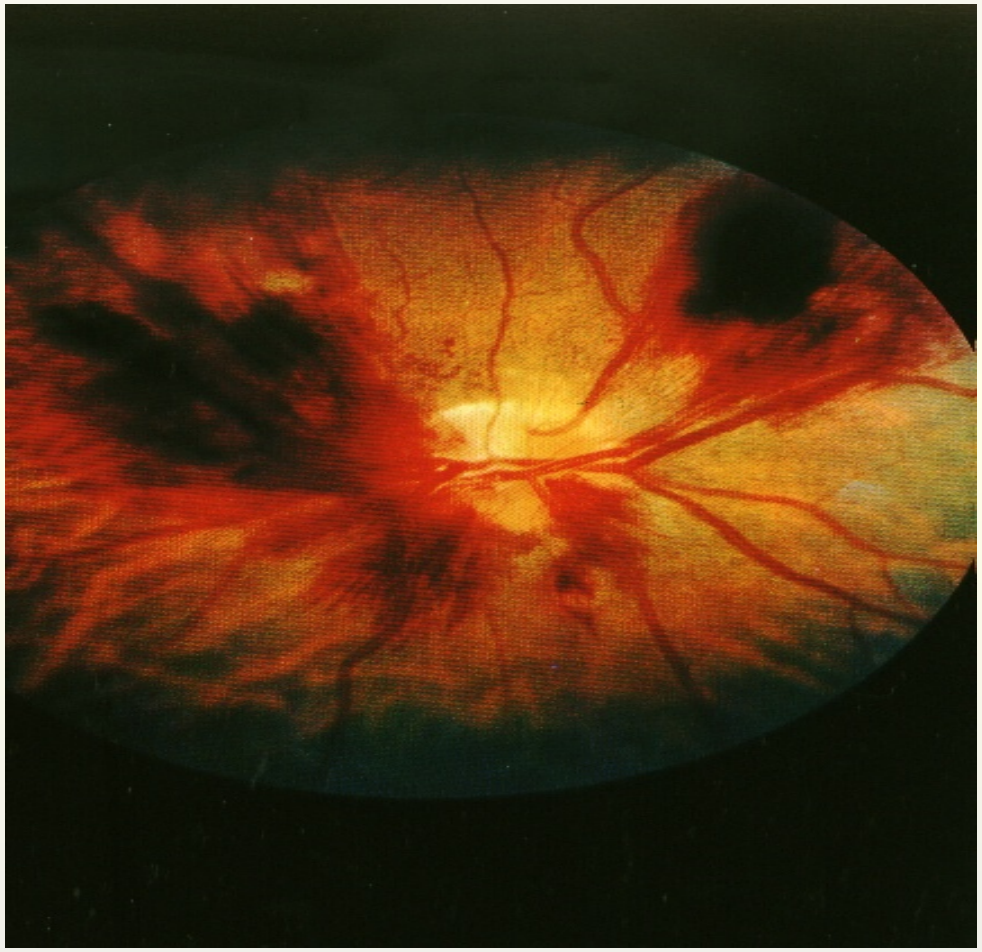
Vascular / Platelet bleeding

- Easy bruising
- Spontaneous bleeding from small vessels
- Purpura & petechiae
- Ecchymoses
- Bleeding mucous membranes - mouth.

Vascular disorders

- Easy bruising
- Bleeding into the skin
- Bleeding from mucous membranes
- Bleeding time normal

Platelet type Haemorrhages



Purpura due to infections

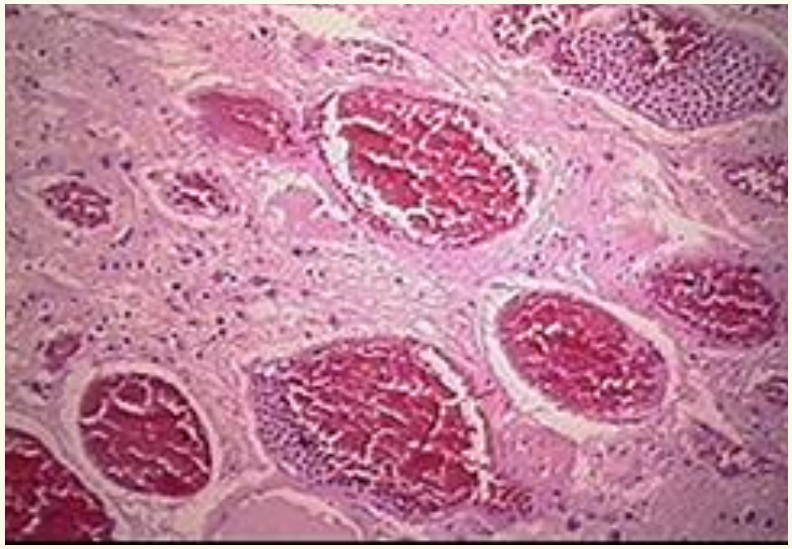
- Meningococcal septicaemia

Gum bleeding in Dengue

Vessels

- Haemangioma
- Telangiectasis
- Vitamin C deficiency

Hereditary haemorrhagic telangiectasia



Hereditary haemorrhagic telangiectasia

- Autosomal dominant
- Mutations in blood vessel development
- Dilatation of capillaries and small arterioles
- Characteristic small red spots
- Blanch on pressure
- Recurrent epistaxis and chronic GI bleeds
- Vascular malformations also occur in pulmonary, hepatic cerebral and spine

Coagulation Factor disorders

- Bleeding after injury
- Haemarthroses
- Muscle haematomas
- A short delay between the event and haemorrhage or haematoma formation

Haemarthrosis & muscle Haematomas

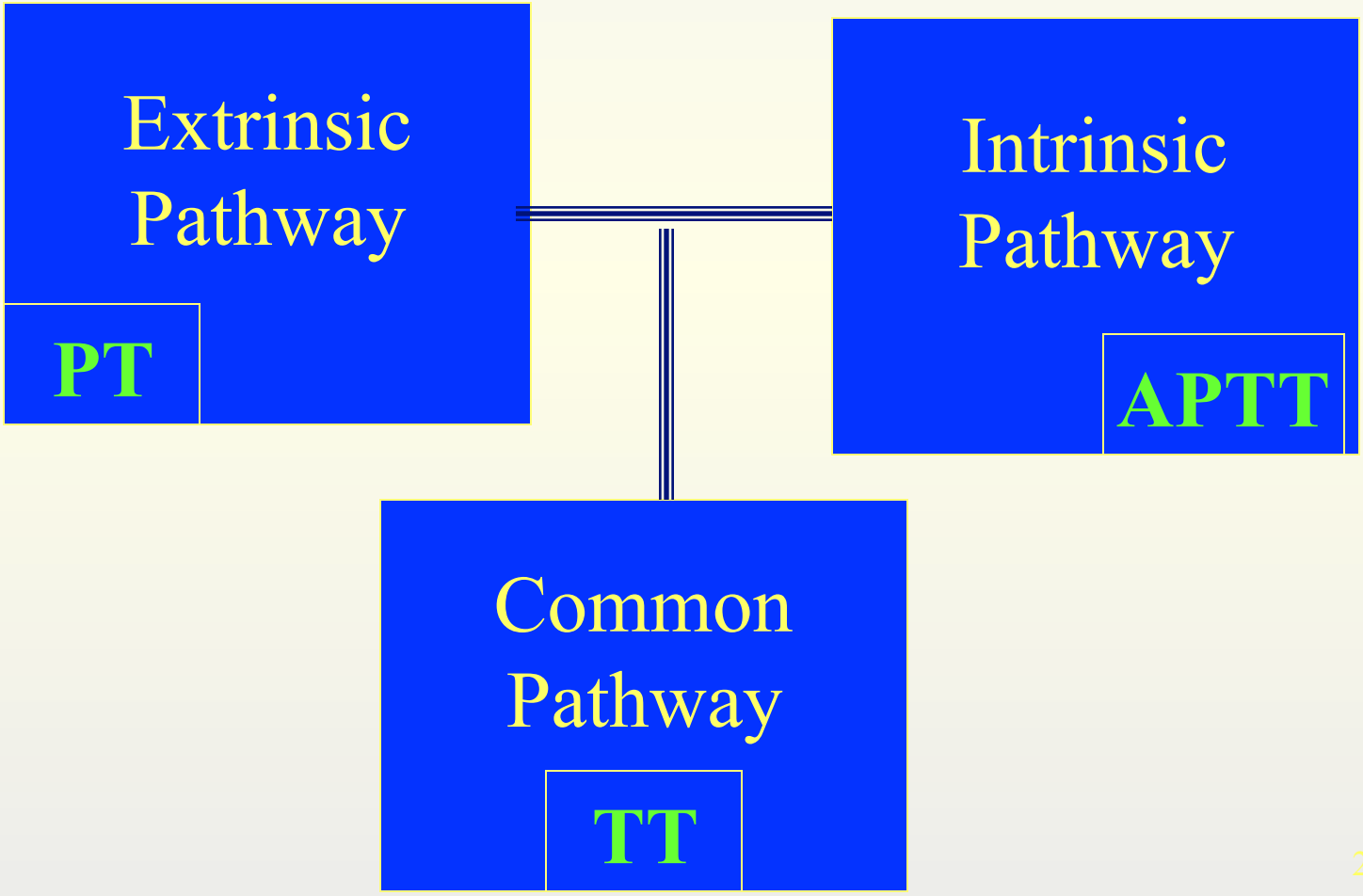
Lab Investigations

- Blood count
- Blood film –
- Number and morphology of platelets

Coagulation tests

- PT
- APTT
- TT
- Correction tests to differentiate prolonged times
- Coagulation factor deficiencies
- Inhibitors of coagulation
- Factor assays

Coagulation tests



Special tests of coagulation

- Fibrinogen and FDPs
- Platelet function tests - platelet aggregation
- Platelet granule contents
- Bleeding time - should not be performed at low platelet counts

Easy bruising syndrome

- Common benign disorder occurring in healthy women
- Bruises on arms, legs and trunk
- Senile purpura and purpura due to steroids due to atrophy of vascular supporting tissue

Self inflicted injuries

- Unexplained bleeding or bruising
- Abuse
- Self-inflicted