Approach to child with bleeding and coagulation disorder

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Learning objectives

- To recognize causes of bleeding in children
- To recognize importance of history and examination
- To differentiate between platelets and coagulation disorders

Introduction

- Bleeding is a diagnostic challenge in children
- Excessive bleeding suggest underlying bleeding disorder
- Minor bleeding is common in children
- Bleeding disorder can be inherited or acquired
- Coagulation, vascular disorder and platelet deficiency are main bleeding disorder
- Evaluation include history, exam and lab tests

Types of Bleeding Disorders

- Coagulation Disorders- congenital/ acquired
  - Hemophilia A (factor VIII deficiency)
  - Hemophilia B (factor IX deficiency)
  - von Willebrand Disease (vWD)
  - Other
- Platelet disorders
  - Thrombocytopenia
  - Platelet functional defect
- Vascular
  - HSP
  - Connective tissue disorder
  - Scurvy

History

- Detailed history
- Types of bleeding
- Emphasis on the child's age, sex, clinical presentation, family history
- General medical history: Malignancy, Sepsis, Drug use, Liver disease etc

Physical examination

- Petechiae: Small pinhead hemorrhagic spot <3 mm
- Purpura (latin mean purple): Red or purpuric discoloration of skin that do not blanch on pressure, size 3-10 mm
- Ecchymosis: size >1 cm
- Hematoma: Collection of blood under skin or muscle of more than 1 cm
- Hemarthrosis: Collection of blood in joints

Physical examination

- Mucocutaneous bleeding suggests a disorder of primary hemostasis, i.e. VWD or platelet dysfunction/deficiency, or a vascular disorder.
- In males, deep hematomas, hemarthroses, or evidence of chronic joint abnormalities suggests hemophilia
- Acquired bleeding disorders may present in the context of coexisting illness
- Anemia
- Thrombocytopenia
- Loose joints and lax skin

Physical examination

- Lymphadenopathy and/or organomegaly suggest an infiltrative process such as malignancy or a storage disease.
- Signs of liver failure suggest acquired coagulation factor deficiencies
- Additional congenital anomalies may suggest the presence of a syndromic bleeding disorder

Table 1. Clinical abnormalities associated with inherited bleeding disorders

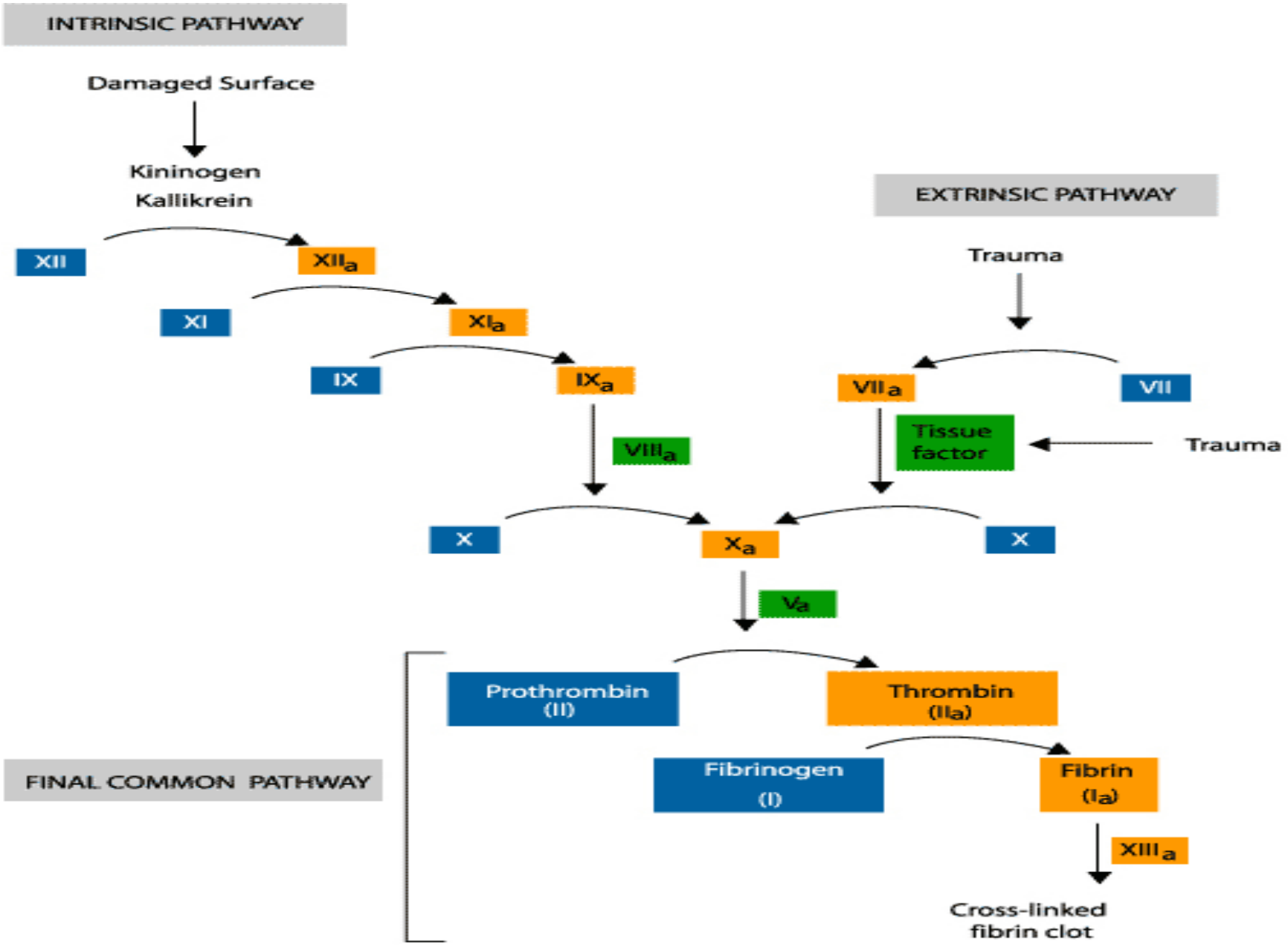
Coagulation defects	
FXIII deficiency	poor wound healing, severe scar formation
Platelet function defects	
Hermansky-Pudlak syndrome	oculocutaneous albinism
Chediak-Higashi syndrome	oculocutaneous albinism, infections, neutrophil peroxidase-positive inclusions
ARC syndrome	arthrogryposis, renal dysfunction, cholestasis
MYH9-related disorders	cataracts, sensorineural hearing defect, nephritis
Leukocyte adhesion deficiency type III	recurrent severe infections, delayed separation of the umbilical cord, neutrophilia
Thrombocytopenia	
Wiskott-Aldrich syndrome	eczema, immunodeficiency
Thrombocytopenia with absent radii, amegakaryocytic thrombocytopenia with radioulnar synostosis	skeletal defects
DiGeorge/velocardiofacial syndrome	cleft palate, cardiac defects, facial anomalies, learning disabilities
Paris-Trousseau/Jacobsen syndrome	cardiac defects, craniofacial anomalies, mental retardation
X-linked thrombocytopenia and dyserythropoiesis with or without anemia/X-linked thrombocytopenia-thalassemia	microcytosis of red blood cells, unbalanced hemoglobin chain synthesis resembling β-thalassemia minor

Clinical features of bleeding disorder

Clinical sign/ symptoms	Platelet/ vascular abnormality	Coagulation factor deficiency
Ecchymosis	Small and superficial	Large and deep
Petechiae	Frequent	Never
Mucosal haemorrhage	Frequent	Uncommon
Muscle/ joint or internal hemorrhage	Uncommon	Frequent
Bleeding after trauma/ surgery	Immediate, stops with pressure	Delayed(1-2 d later) does not stop with pressure
Example	vWD, ITP	Hemophilia A and B

Laboratory investigations

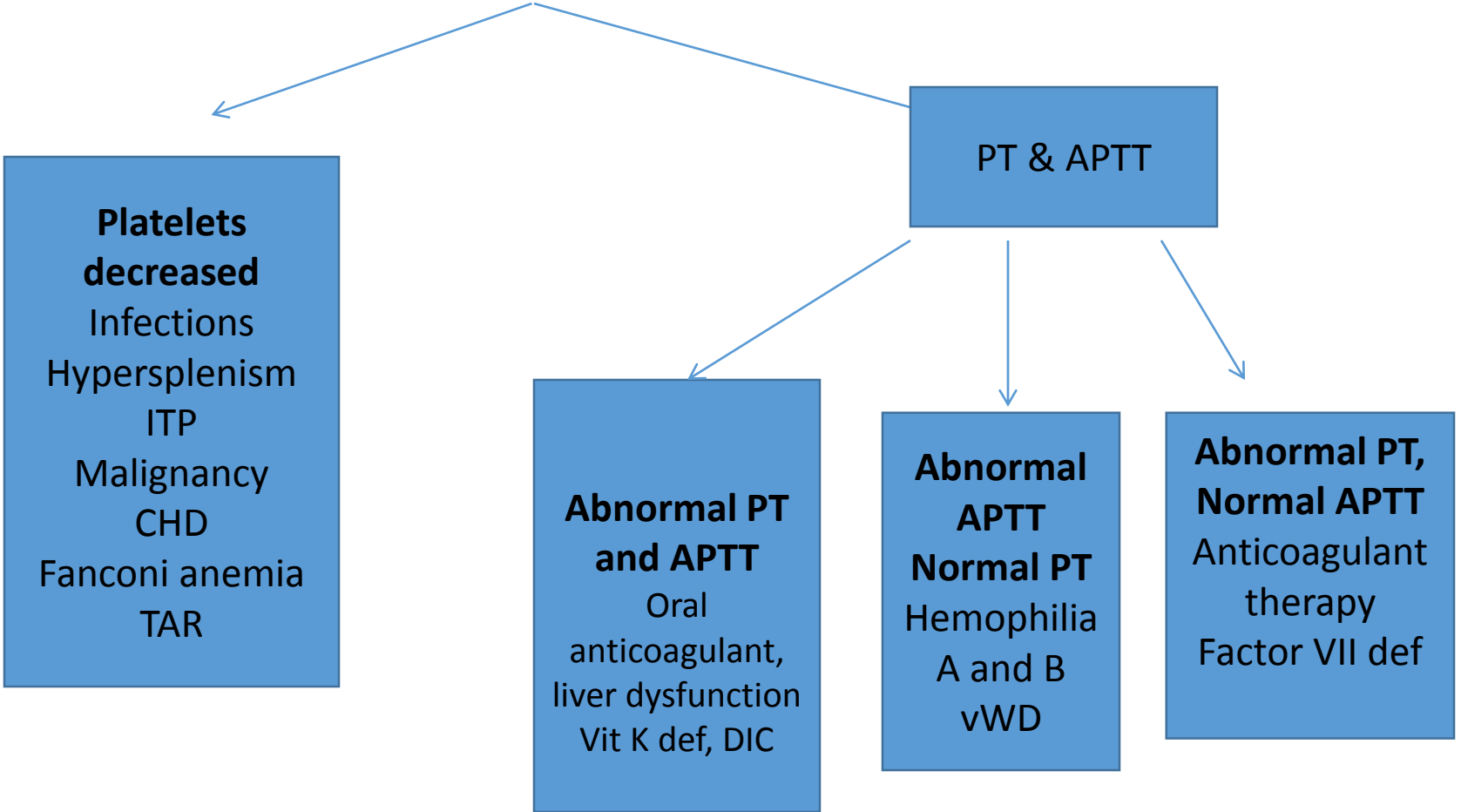
- CBC
- Peripheral smear
- Bleeding time – platelet no/ functions
- PT- (12±2 sec, 2, 5, 7, 10)
- aPTT- (25-40 sec- 5, 8, 9, 10, 11, 12)
- Platelet function testing

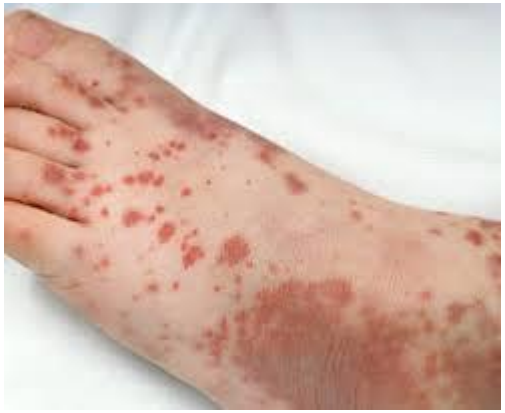

INTRINSIC PATHWAY
Damaged Surface
Kininogen
Kallikrein
XII
XIIa
XI
XIa
IX
IXa
VIIIa
EXTRINSIC PATHWAY
Trauma
VIIa
VII
Tissue factor
Trauma
X
Xa
X
Va
FINAL COMMON PATHWAY
Prothrombin (II)
Thrombin (IIa)
Fibrinogen (I)
Fibrin (Ia)
XIIIa
Cross-linked fibrin clot
AnaesthesiaUK

GC	BT	PT	APTT	P/C	Diagnosis
Well	N	N	↑	N	Hemophilia A and B
Well	N	↑	N	N	Factor 7 deficiency
Well	N	↑	↑	N	Vitamin K deficiency
Sick	↑	↑	↑	↓	DIC, Liver disease
Well	↑	N	N	↓	ITP
Sick	↑	N	N	↓	Aplastic anemia, leukemia
Well	↑	N	N	N	Qualitative platelet defect

Lab workup

History and physical examination



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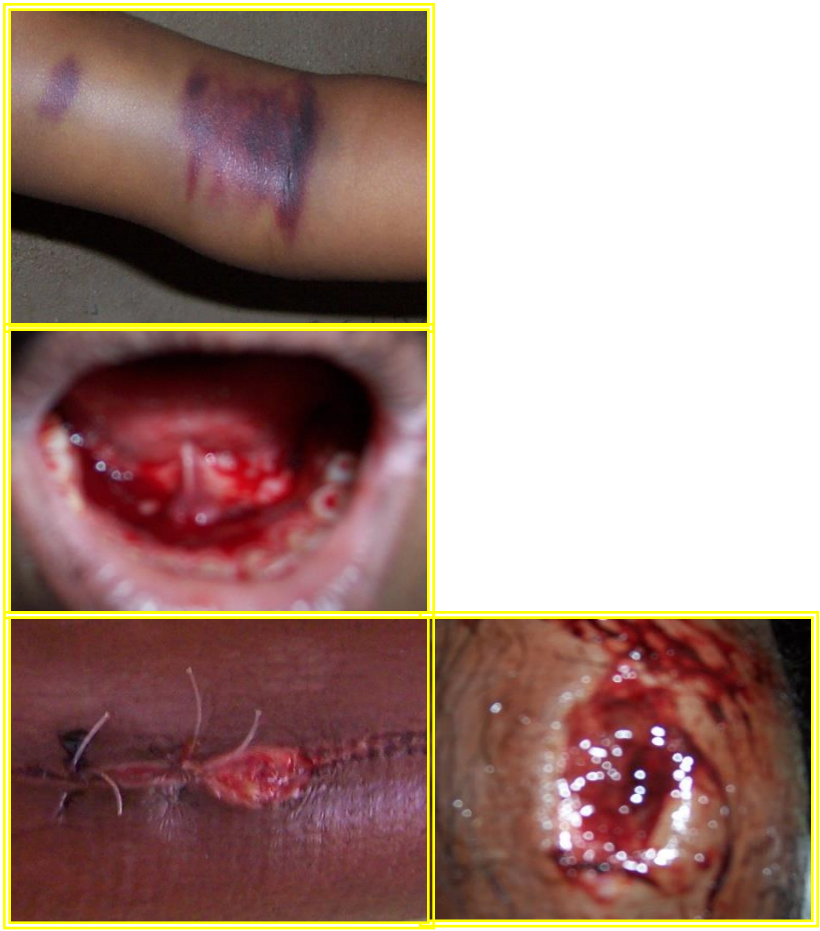
Clinical manifestations of hemophilia

Hemophilia can affect any organ in the body

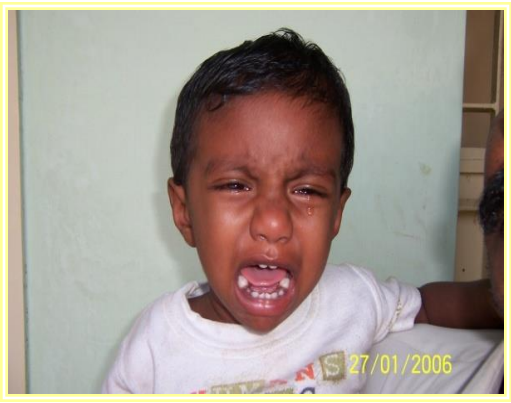
Early symptoms

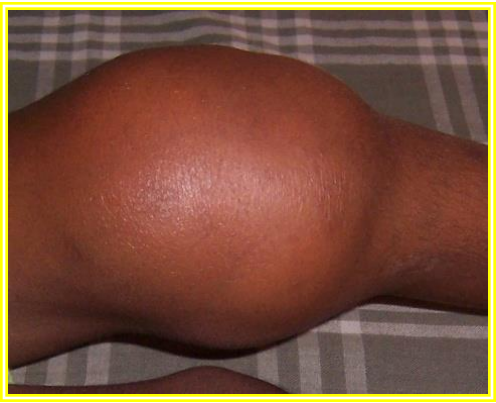
From Childhood

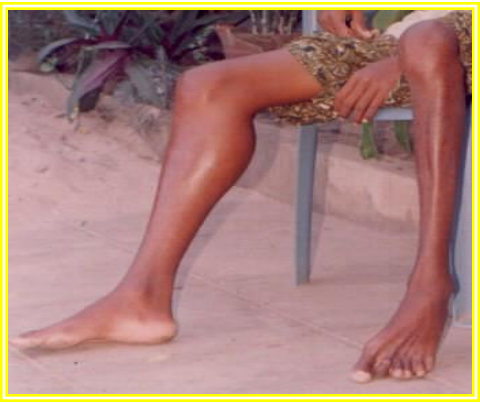
- Blue patches and bruises on the skin.
- Gum bleeds
- Frenulum bleeds.
- Cuts and Wounds which Bleed a long time.



Early symptoms

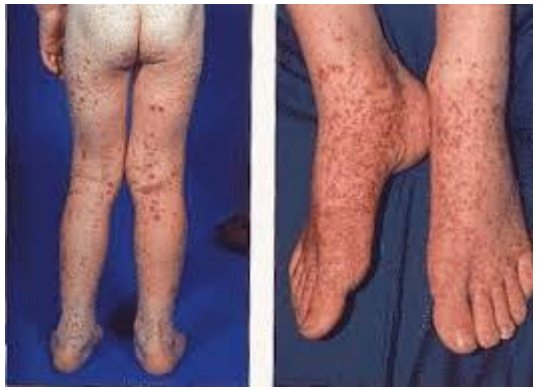






- Unbearable pain in Muscles and joints
- Swelling.
- With out Injury

HSP: Typical rash



Quiz

Q1. All of following are true except

a) Intrinsic pathway of coagulation can be determined by PT
b) PT is increased in hemophilia
c) Platelets are normal in HSP
d) None

Thank You