

HEMOPHILIA:

COAGULATION DISORDERS CLASSIFICATION:

- | | |
|----------------------------|------------------------|
| → A HEREDITARY COAGULATION | B ACQUIRED COAGULATION |
| → Hemophilia A (VIII) AR | → Vitamin K deficiency |
| → Hemophilia B (IX) AR | → Liver disease |
| → VWD | → Other |
| → other | |

NOTE:

Def of factor VIII - von W Factor complex:

↓
has two components (Plasma VIII)
VWF factor (This protein factor VIII)

Function of VWF

- Subendothelial injury contain VWF (get exposed due to Any trauma or injury). This facilitate Adhesion of platelets to B.V

Initial step is hemostatic plug

Inadequate VWF is Responsible for bleeding

↓
Thus def of VWF lead to 2nd def of factor VIII

HEMOPHILIA - A:

- Most common hereditary diseases
- X Linked Recessive disease. — Reduced VIII Activity

↓
Life threatening bleeding

{ due to inadequate coagulation & inappropriate clot Removal }

Mode of Inheritance:

X^H X^H
X^h X^h

- Genes are located on long arm of X chromosome.
- Males with single X chromosome (X^H) suffer from Hemophilia.
- Heterozygous female are carriers.

Clinical Features

- easy bruising & haemorrhage into joints & soft tissue.
- Haematuria, Resp + intestinal obstruction → Prolonged bleeding.
- Petechiae are Absent.
↳ pin point haemorrhages

Lab Findings

- BT: Normal
- CT - Prolonged
- APTT (Prolonged Activated Partial Thromboplastin time) → Prolonged
- Factor VIII assay → ↓ sect
- FDP

Platelet count - Normal

Complication

- Deforming Arthritis & contractures.
- Anemia.

tlf → Recombinant VIII factor.

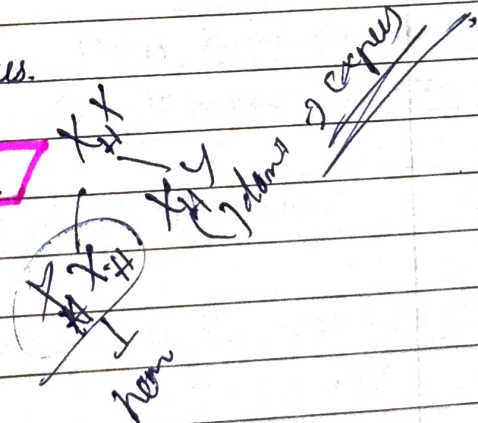
X^HX^H

X^HY

X^HX^H

X^HX^h

X^hY



CLL

Tumors composed of monomorphic small B lymphocytes in P-S & B-M

Lb: $\rightarrow$ 1161, WBLM, chronic lymphocytic leukaemia

WBL $\rightarrow$ lymphocytes - soccer ball chromatin
 BM involvement - nodular, diffuse, interstitial
 lymphocytic infiltration

MYELOPROLIFERATIVE NEOPLASMS:

chronic myeloproliferative disorders

chronic neutrophilic leukaemia

Polycythemia vera

chronic eosinophilic leukaemia

Essential thrombocythosis

chronic myeloid leukaemia

