

Hemophilia

Blood home $\rightarrow$ loves to Blood.

Hemophilia A & B

$\Rightarrow$ It is X linked Recessive disorder, affects only men and females are carriers.

- Two Types

i) Hemophilia A $\rightarrow$ VIII Factor Deficiency $\sim 1/10,000$,

ii) Hemophilia B $\rightarrow$ IX Factor Deficiency - Rarer Common (Christmas disease)

Causes: (For acquired)

- Liver Failure
- Vitamin K Deficiency.
- Autoimmune against clotting factor.
- Disseminated Intravascular Coagulation.

Clinical Features $\rightarrow$ Nearly identical for A & B.

- Easy Bruising (Ecchymosis)
- Hematomas (In muscles, calf & prostates);
- Prolonged bleeding after cut or surgery.
- oozing after tooth extraction. Epistaxis.
- GIT, GUT. Bleeding.
- Haemarthrosis (Bleeding into joints), Swollen painful joints,
- Bleeding into Brain $\rightarrow$ Stroke, Rised Intracranial pressure.

Investigations

Blood $\rightarrow$ Normal Platelets, Prothrombin time $\rightarrow$ Normal

$\rightarrow$ (Test for Extrinsic & Common pathway)
 $\rightarrow$ (II, X, V, II, I)

Partial Thromboplastin Time $\rightarrow$ Increased

- Test for intrinsic & common pathway (XII, XI, IX, VIII, X, V, II, I)

$\rightarrow$ $\downarrow$ VIII $\rightarrow$ A, $\downarrow$ IX $\rightarrow$ B.

$\rightarrow$ Gene therapy.