Hereditary bleeding disorders caused by deficiencies of either clotting factor VIII or IX

**General**
- Deficiency of factor VIII
  - X-linked recessive
  - Occurs 1/10,000 male births
  - Flip tip inversion of factor VIII gene
  - 30% have no family history
  - High rate of new mutations
  - Often in early life or following surgery/trauma
  - Presentation depending on severity
  - Leads to crippling arthropathy
  - Can cause nerve palsy and compartment syndrome
  - No petechiae or ecchymoses
  - Haemarthrosis
  - Haematoma

**Clinical Features**
- Increased APTT
- Decreased factor VIII assay
- Normal PT
- Normal platelet count and bleeding time
- Avoid NSAIDs/IM injections
- Stimulates release of factor VIII from endothelial cells
- Not for severe Haemophilia A
- Give Desmopresin
- Recombinant factor VIII if severe bleeding

**Diagnosis**
- Haemophilia A
- Christmas Disease
  - Factor IX deficiency
  - X-linked recessive
  - Clinically identical to Haemophilia A
  - To differentiate check individual factor levels

**Management**
- Haemophilia A
- Christmas Disease
- Factor IX deficiency
  - Clinically identical to Haemophilia A
  - To differentiate check individual factor levels