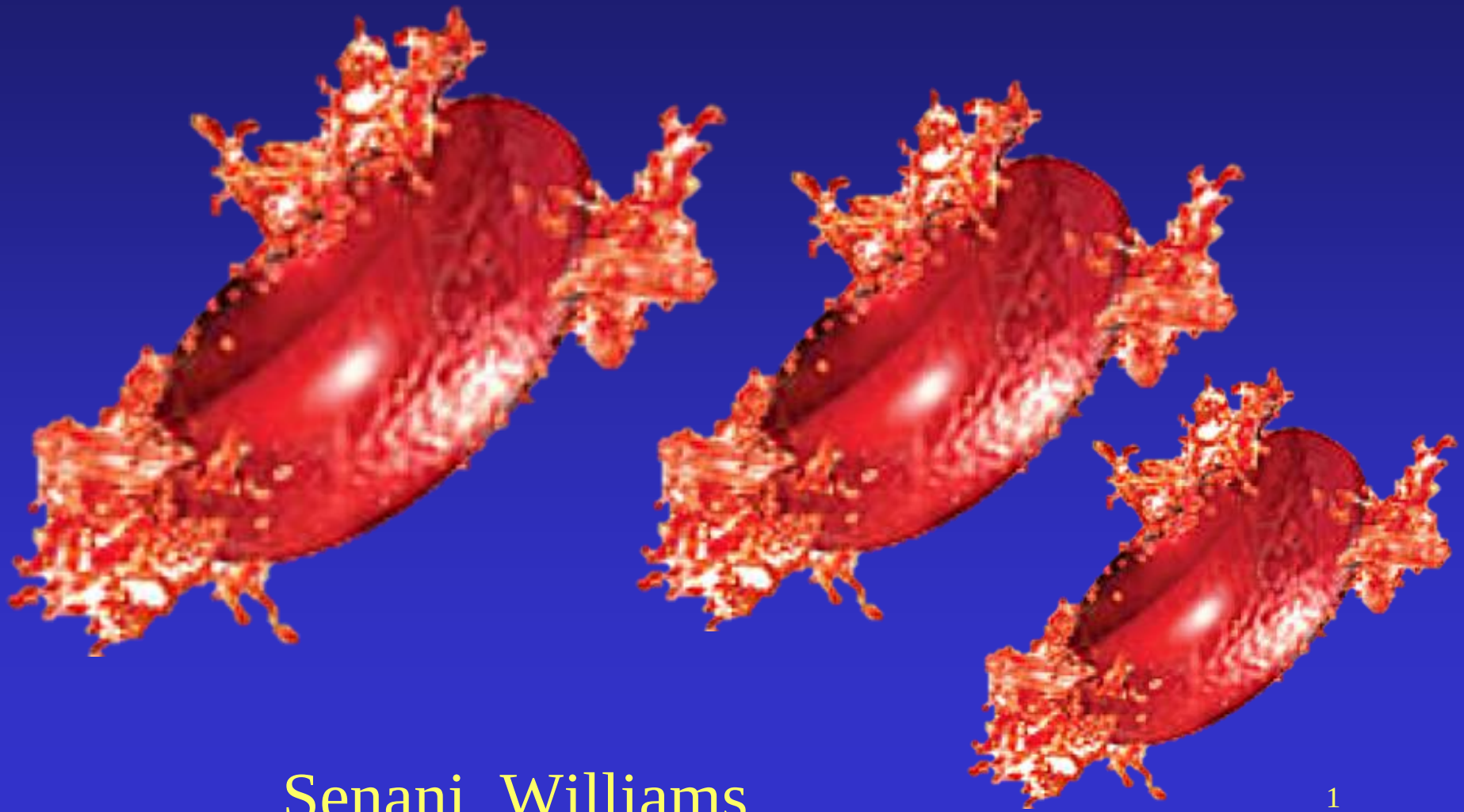
General Considerations of Haemolytic Anaemia



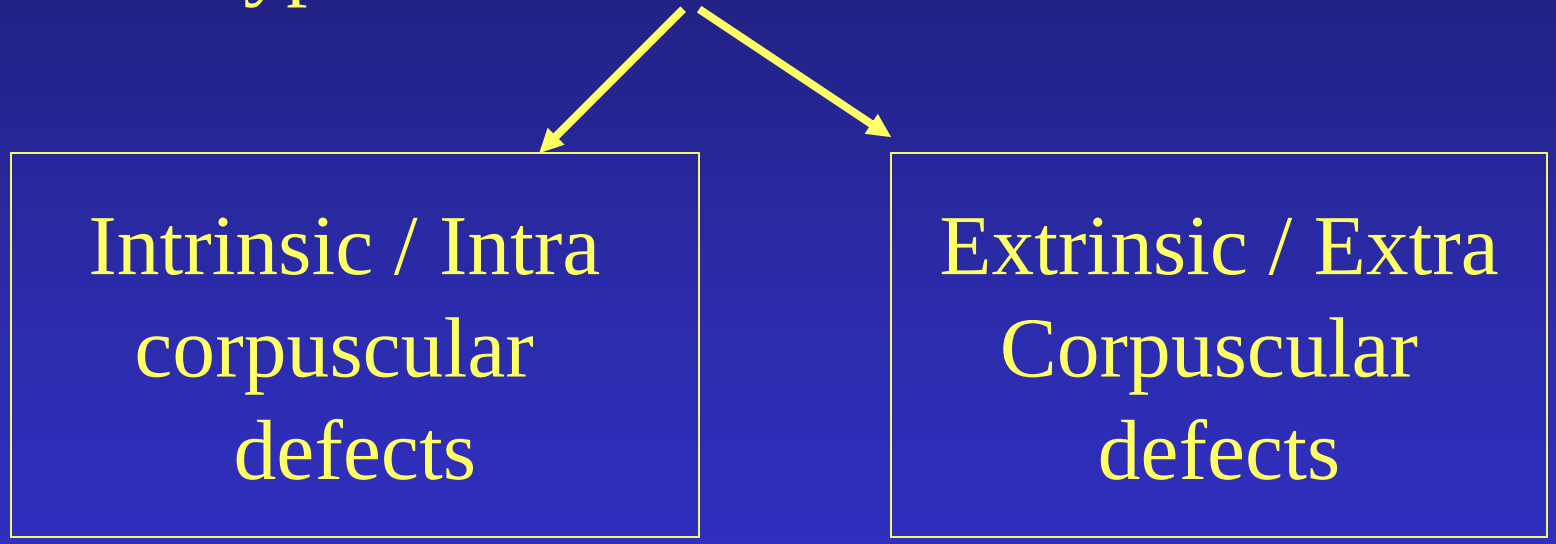
Senani Williams

Haemolytic Anaemia

- Premature destruction of RBC's - < 120 days
- Erythroid hyperplasia in BM – up to 8 times the normal
- ↓ Hb occurs with decompensation

Haemolytic Anaemia

- 2 types of defects

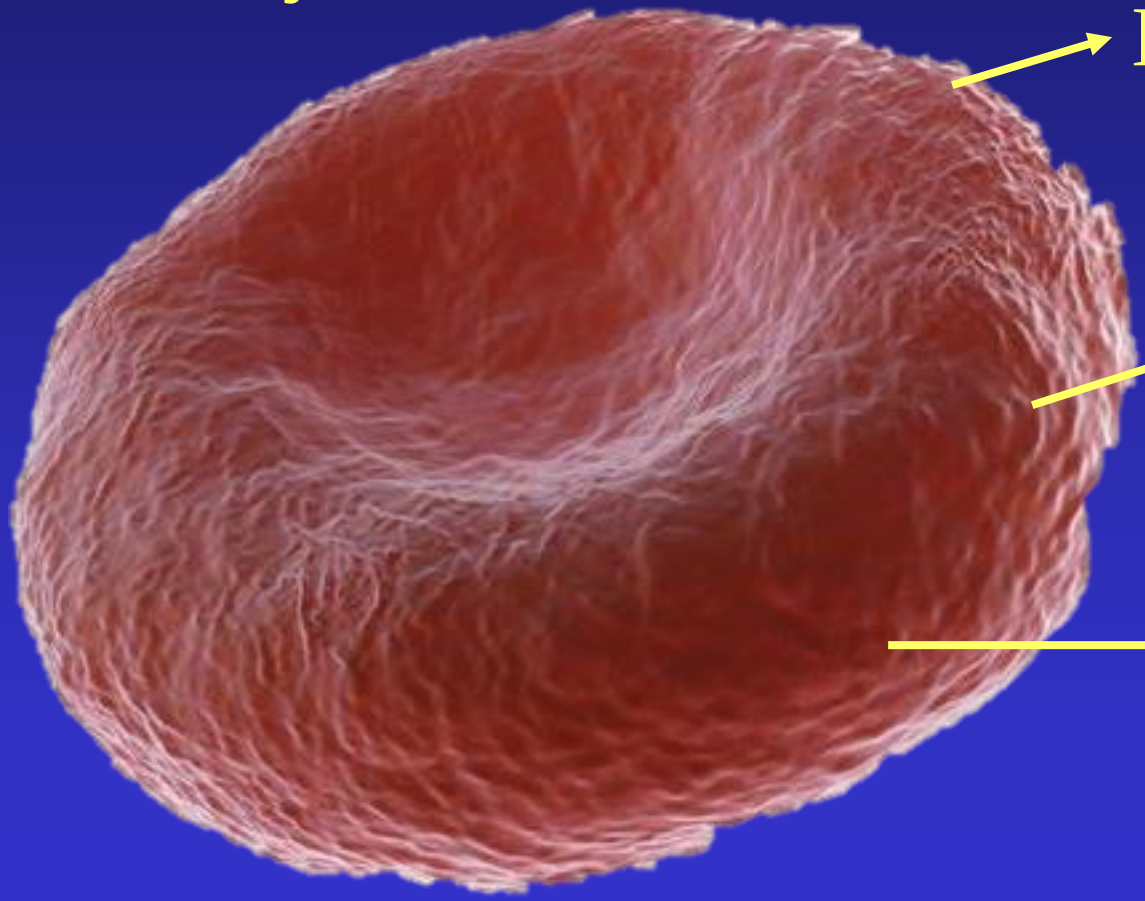


Usually Inherited

Usually Acquired

Intrinsic / Intra corpuscular Defects

Usually Inherited



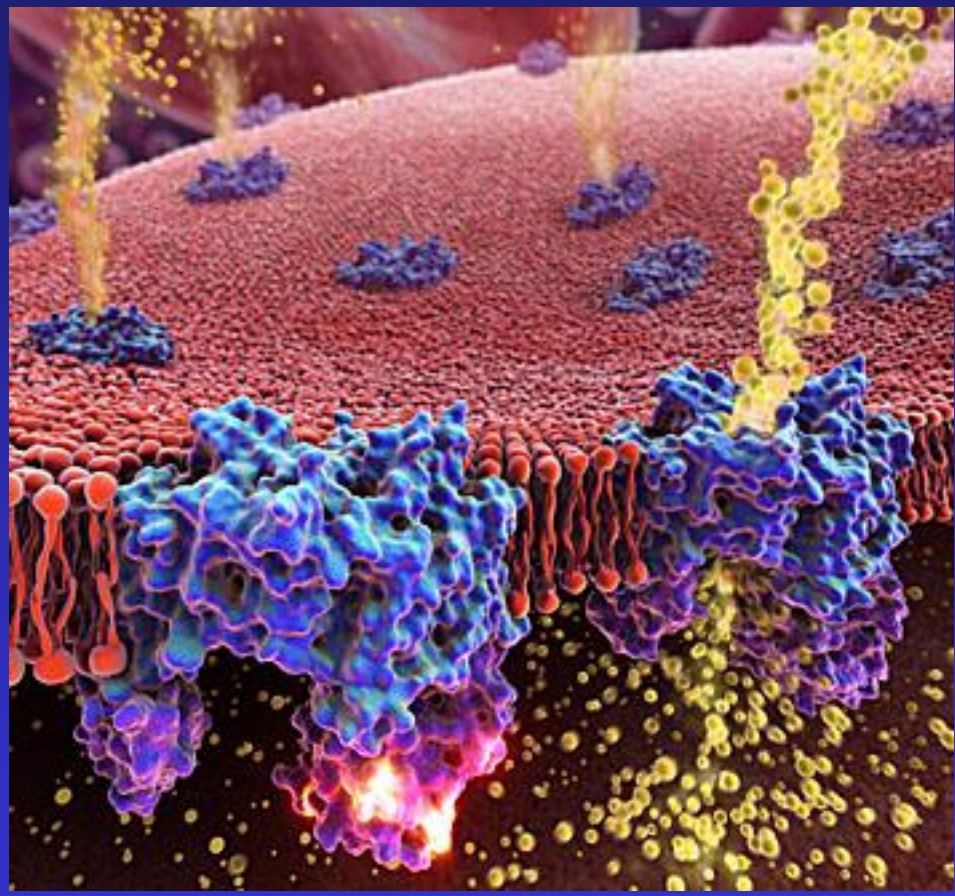
Membrane defects

Enzyme defects

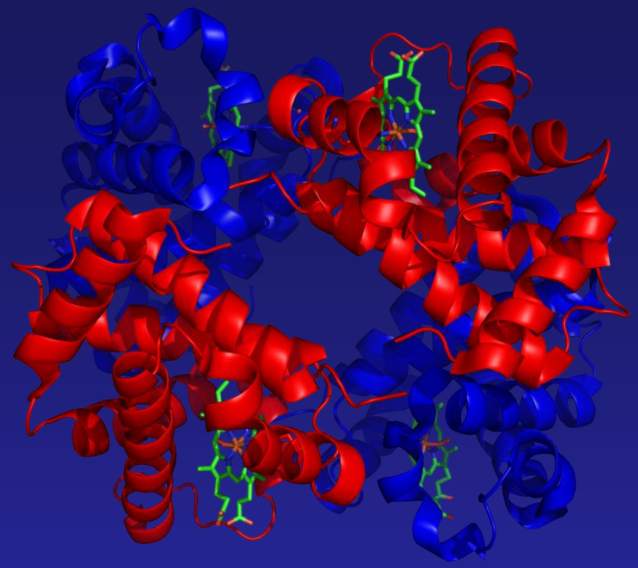
Hb

Membrane Defects

- All inherited except
- Paroxysmal Nocturnal Haemoglobinuria



Haemoglobin defects



- 2 types of defects

Structurally abnormal haemoglobin

Haemoglobinopathy – HbS

Abnormal amounts of structurally normal globin synthesized Thalassaemia

Enzyme Defects

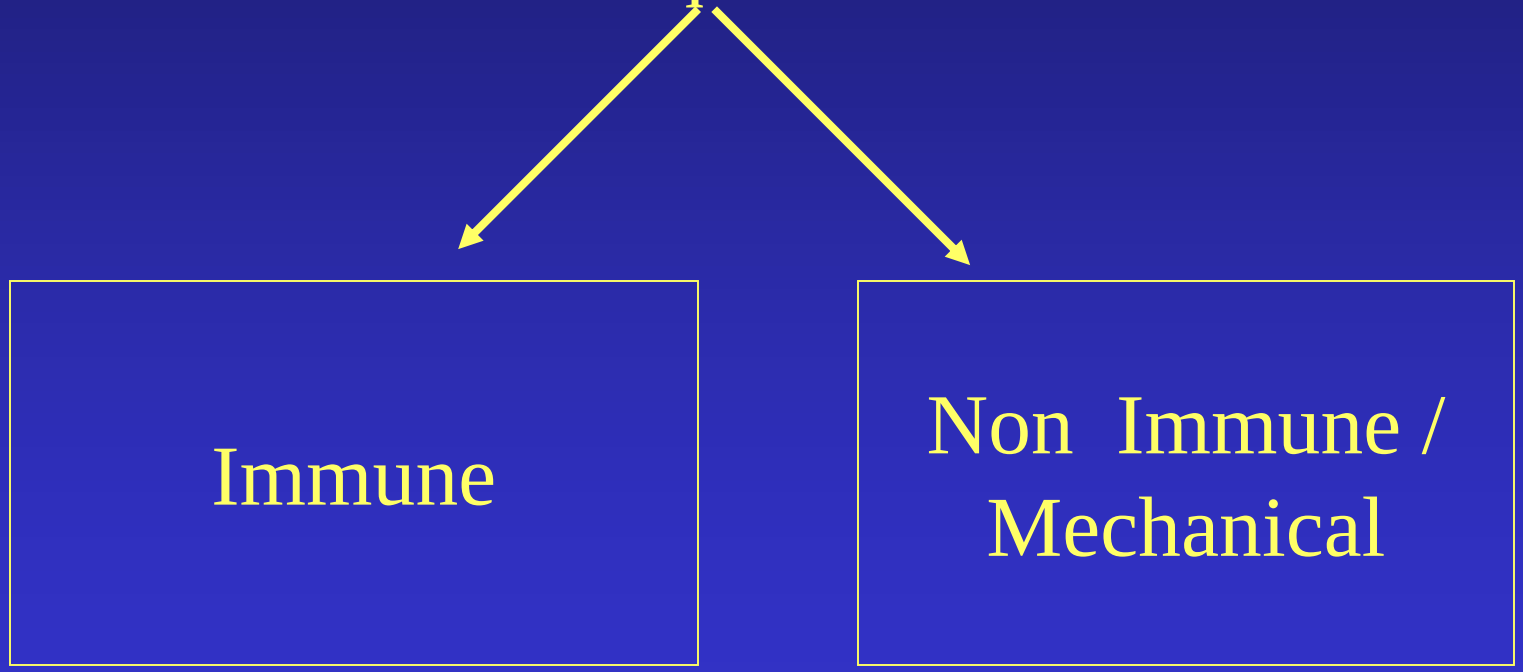
- G6PD deficiency

- Pyruvate Kinase deficiency

Extrinsic / Extra corpuscular Defects

Extrinsic / Extra corpuscular Defects

- Most often acquired



Extrinsic Defects / Non immune causes of Haemolytic Anaemia

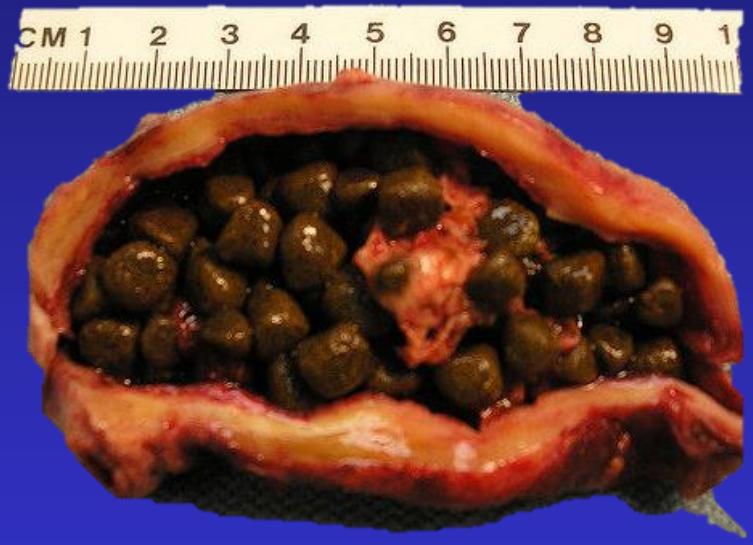
- Burns
- Infections
- Drugs
- Chemicals
- Rheumatoid arthritis
- CRF
- Liver disease
- Pb
- Malignancy

Evidence of Haemolysis

- Evidence of ↑Hb breakdown
- Evidence of bone marrow erythroid hyperplasia
- Evidence of ↓RBC lifespan

Evidence of ↑ Hb breakdown

- ↑ S. unconj. Bilirubin
- +/- jaundice



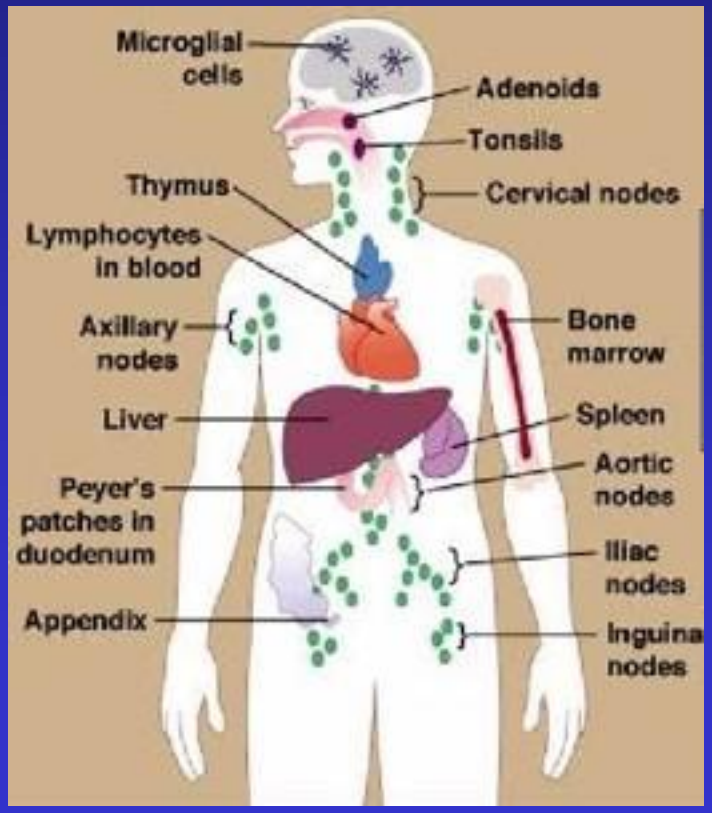
- pigment gall stones - black, amorphous
- ↑ urobilinogen
- ↑ LDH (LDH $_2$ – fraction)

Evidence of Reduced red cell life span

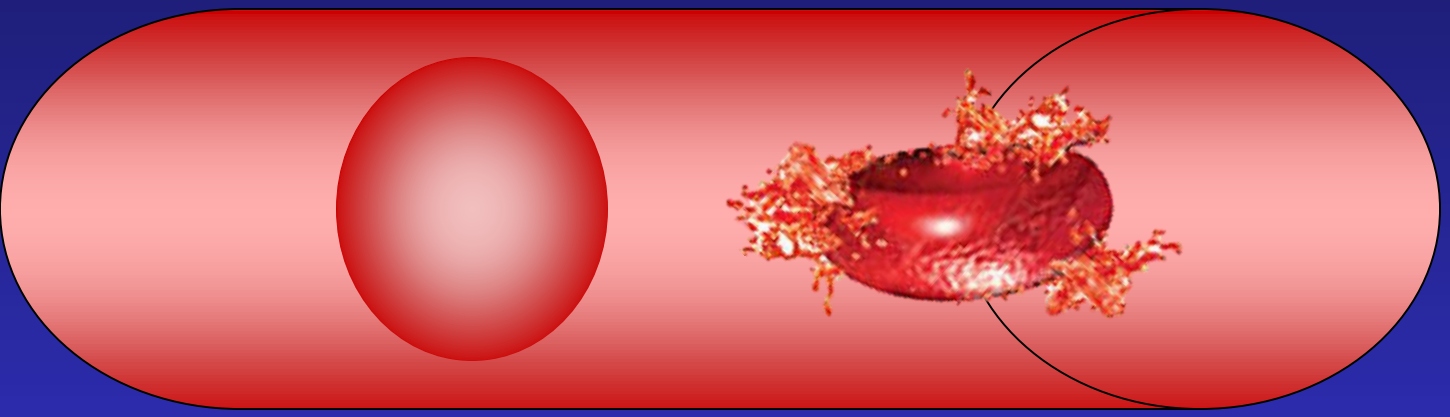
- ^{51}Cr labelled RBCs

Where is the destruction occurring ?

Intravascular



Intra vascular haemolysis

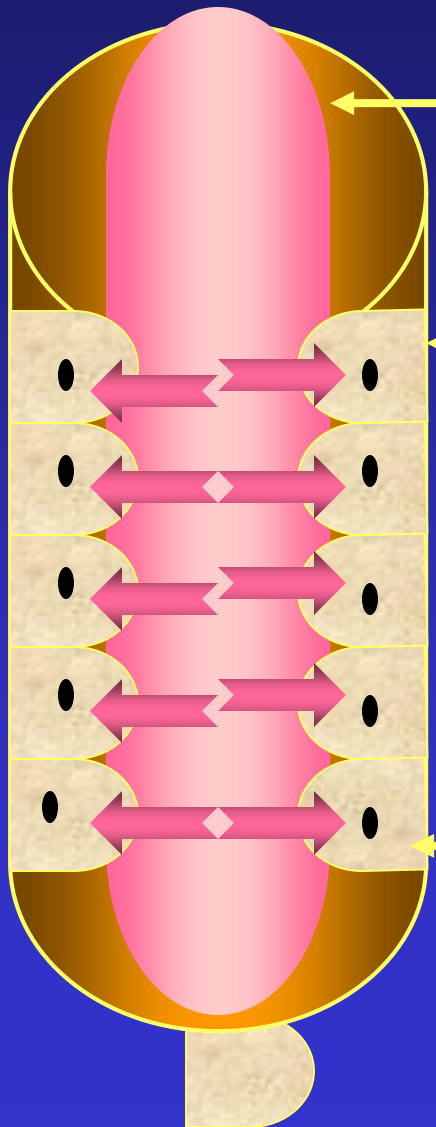


Haemoglobin breakdown products in plasma and urine

Usually an acute release of free Hb

Intra vascular haemolysis

Haemosiderinuria



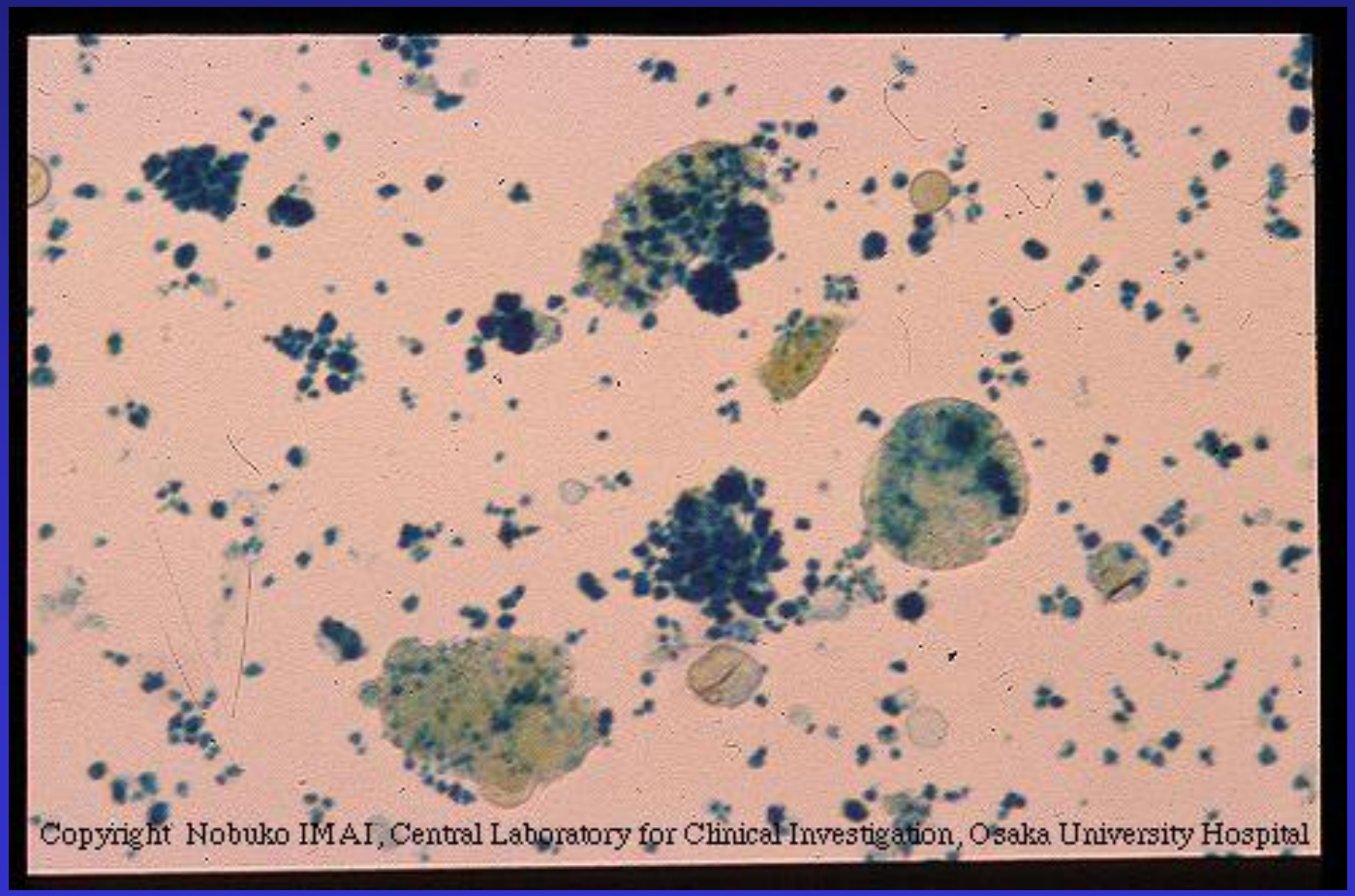
Filtered plasma containing Hb

Renal tubular epithelial cells absorbing Hb

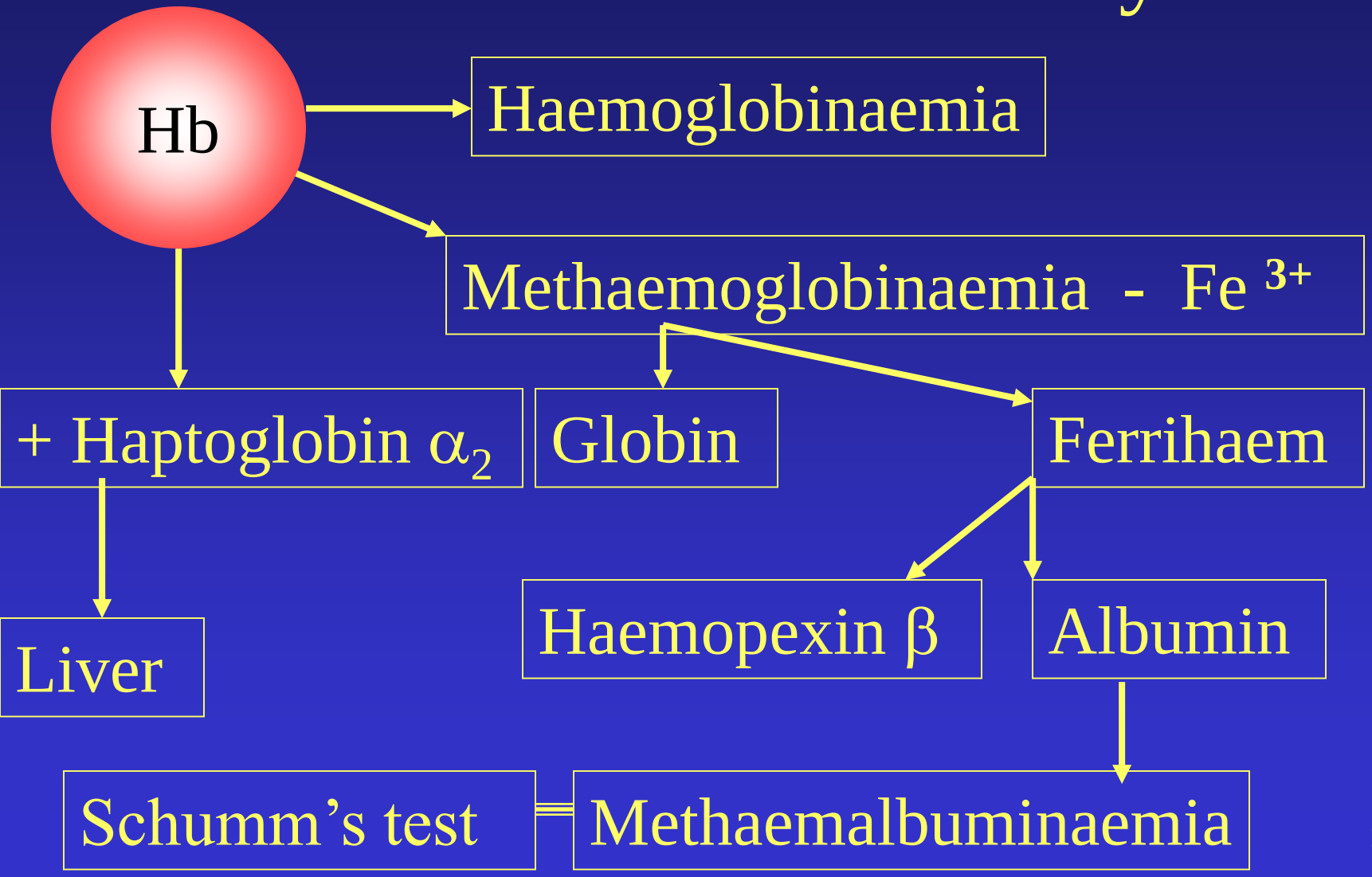
Stored as Haemosiderin

Renal tubule

Haemosiderinuria



Intra vascular Haemolysis



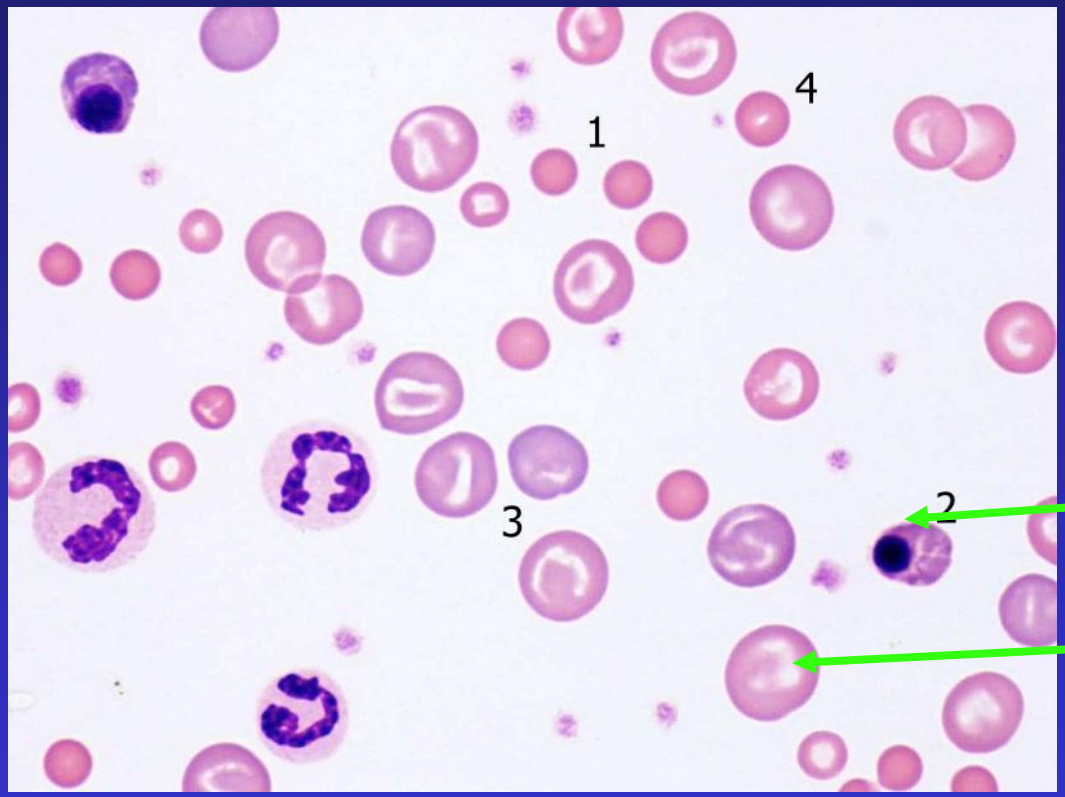
Causes of intravascular haemolysis

Homework

Extra Vascular Haemolysis

- Unconjugated serum bilirubin ↑
- **NO** free haemoglobin
- Exaggeration of normal removal of senescent red cells

Blood Picture

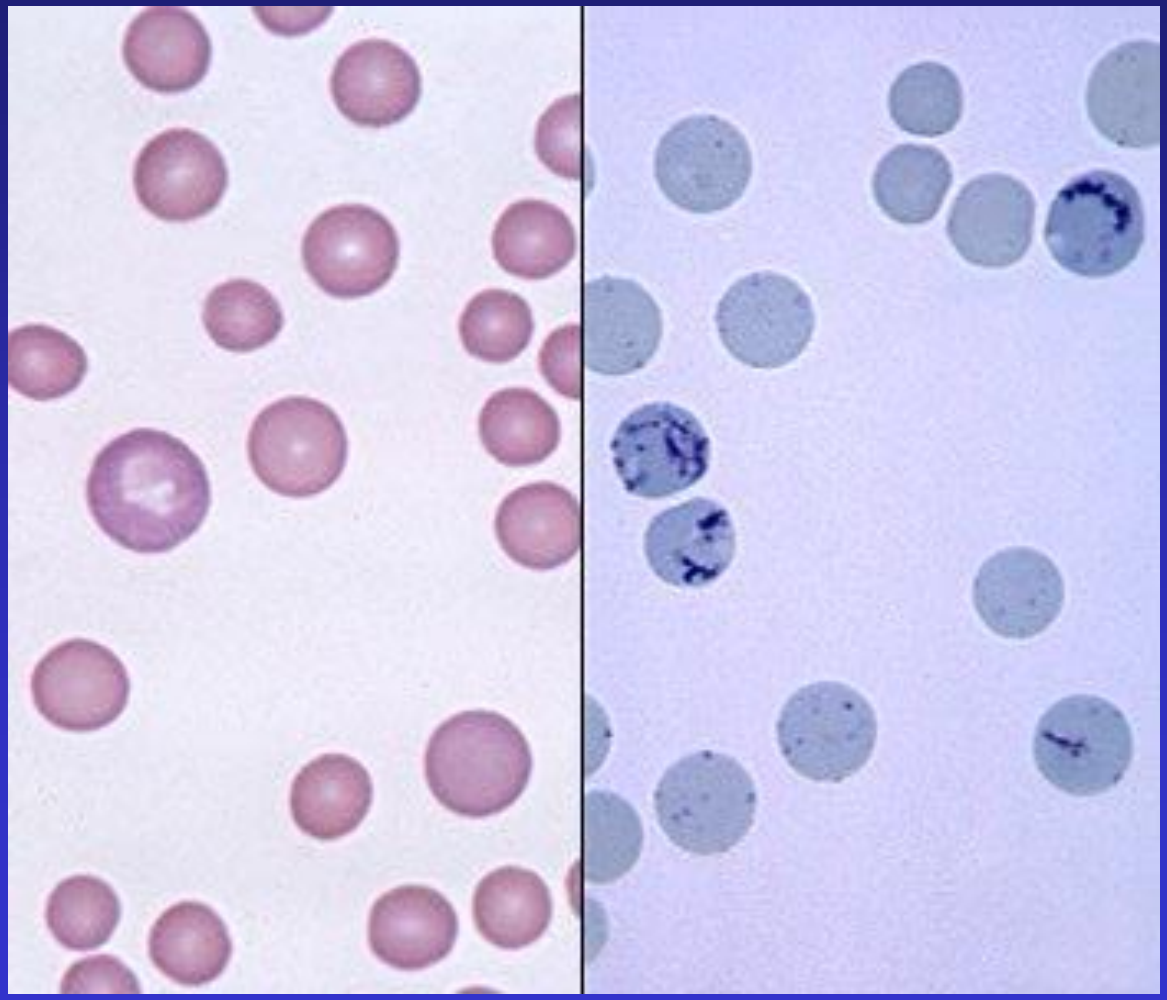


Nucleated red cells

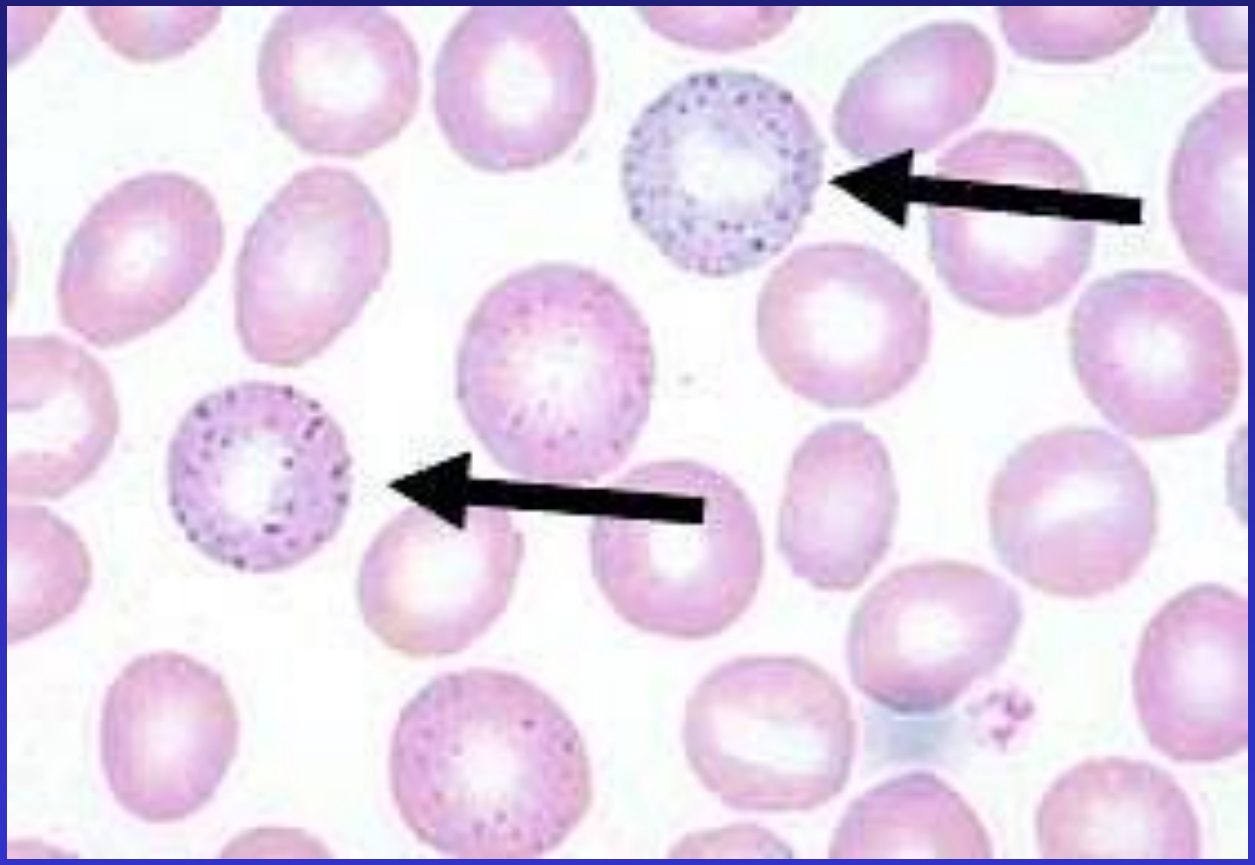
Polychromatic macrocytes

“shift reticulocytes”

Polychromatic cells : Reticulocytes

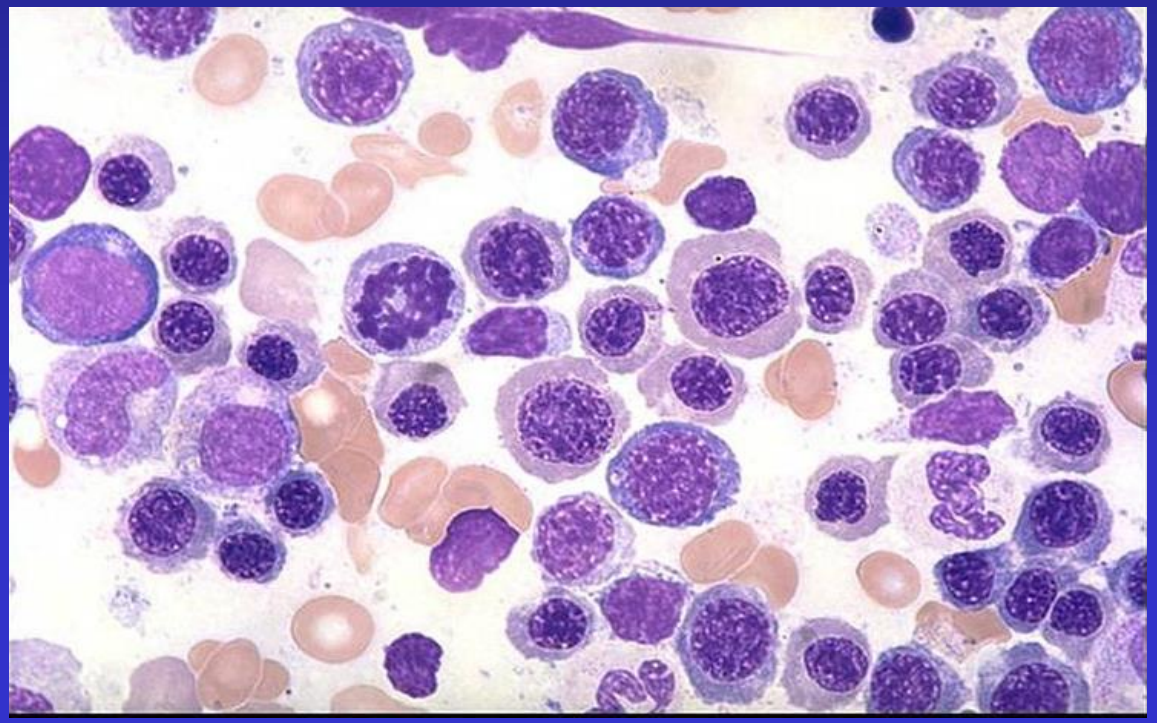


Basophilic stippling



Bone Marrow

- Hypercellular
- Reduced fat spaces
- Reversed M : E ratio
- Erythroid hyperplasia
- - normoblastic



Skeletal (Radiological) Abnormalities

- Skull - broadening of diploic space hair on end appearance in children

Skeletal (Radiological) Abnormalities

- Irregular bony sclerosis in sickle cell anaemia due to vascular thrombosis & infarction