Anemia classification, blood loss anemia and hereditary spherocytosis

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DEFINITION

Anemia (*An = without, emia =blood*) is a decrease in the RBC count, hemoglobin and/or Hematocrit values resulting in a lower ability for the blood to carry oxygen to body tissues .

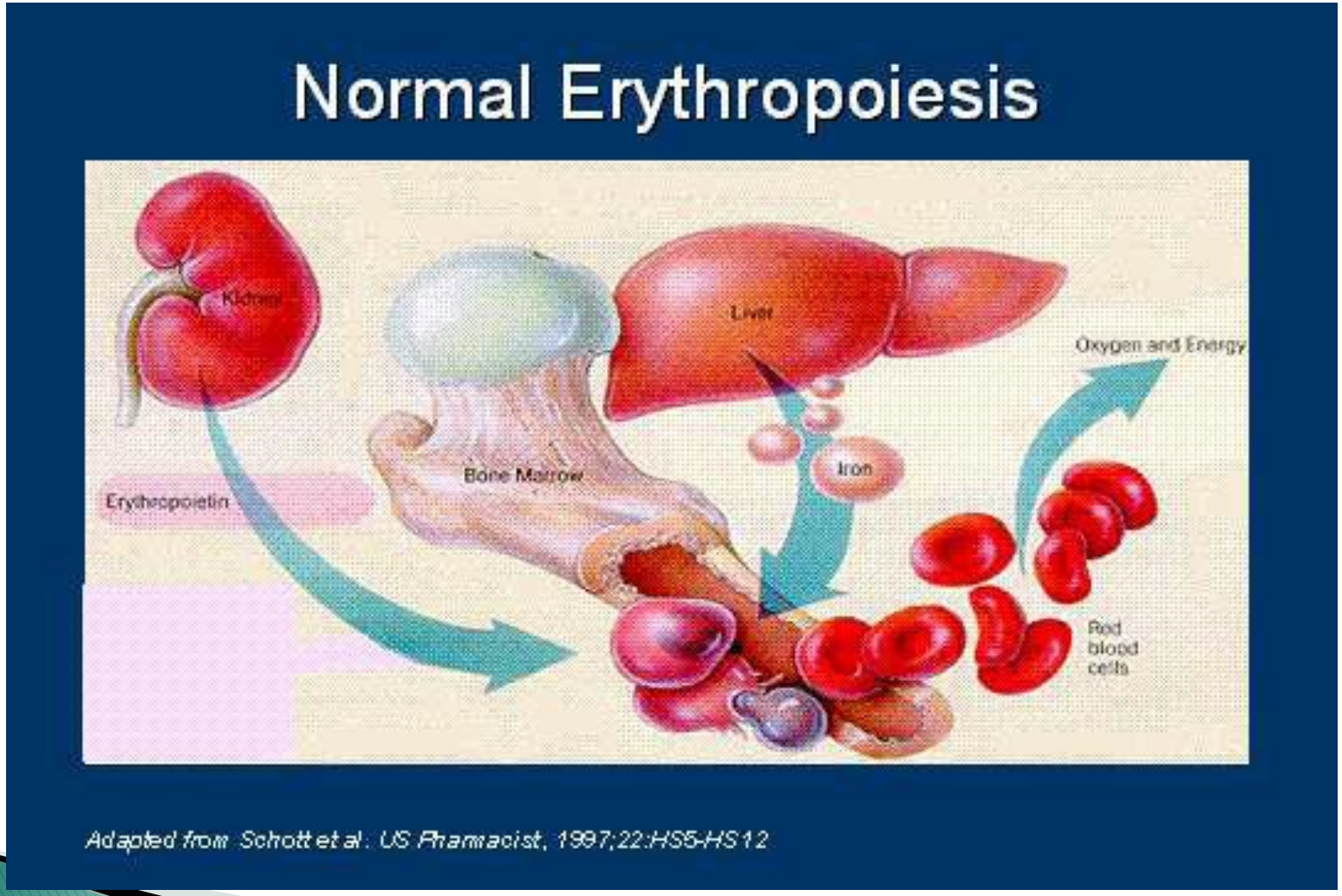
DEFINITION OF ANAEMIA

- “ ANEMIA is present when the haemoglobin level in the blood is below the lower extreme of the normal range for the age and sex of the individual.”

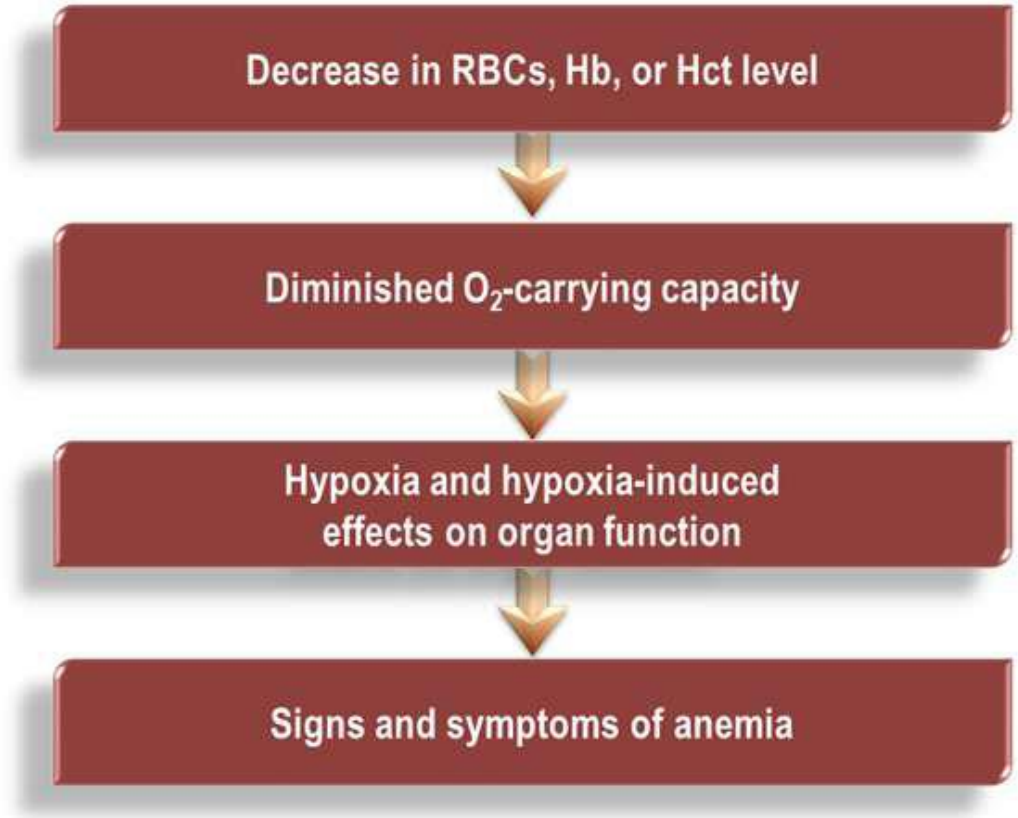
Laboratory Definition of Anemia

- **Hb; (G/dl)**
  Women: <12.0
  Men: < 13.5
- **Hct:**
  Women: < 36
  Men: <41

PATHOPHYSIOLOGY



PATHOPHYSIOLOGY



CLASSIFICATION OF ANAEMIA

Classification

A. Underlying mechanism (ETIOLOGICAL)
   1) blood loss
   2)increased destruction
   3)decreased production

B. Morphology of erythrocytes
   1) **size** (micro-, macro-, normocytic)
   2) **shape** (spherocytosis, stomato-,...)
   3) **color** (degree of hemoglobinization normo- hypo-, hyperchromic)

ETIOLOGICAL CLASSIFICATION

A. BLOOD LOSS ANAEMIA

- 1. OVERT BLOOD LOSS
- SURGERY
- ACCIDENT
  - EPISTAXIS
  - RECTAL BLEEDING
  - MENORRHAGIA
  - RECURRENT BLEEDING FROM ANY OTHER SITE
- 2. OCCULT BLOOD LOSS
- GI BLEEDING
- GENTI-URINARY BLEEDING

B.EXCESSIVE DESTRUCTION OF RBC *(HEMOLYTIC ANEMIA)*

1. INTRACORPUSCULAR DEFECT

I) Membrane defects;

Hereditary spherocytosis

Hereditary ovalocytosis, etc.

II) Enzyme defects :

G-6PD deficiency,

PK def., etc.

III) Hemoglobin defects :

Thalassemia, sickle cell anemia Hemoglobinopathies

B.Excessive Destruction of RBC CONT….

- 2.EXTRACORPUSCULAR DEFECT

1) Mechanical :

March hemolytic anemia

MAHA (Microangiopathic HA)

2) Chemical/Physical

3) Infection : *Clostridium tetani*

4)Antibodies : HTR, SLE

5) Hypersplenism

C. IMPAIRED RBC PRODUCTION

- 1. Abnormal bone marrow
  - Aplastic anemia
  - Myelophthisis : Myelofibrosis, Leukemia,
  - Cancer metastasis
- 2. Essential factors deficiency
  - Deficiency anemia : Fe, Vit. B12, Folic acid, etc
  - Anemia in renal disease : Erythropoietin
- 3. Stimulation factor deficiency
  - Anemia in chronic disease
  - Anemia in hypopituitarism
  - Anemia in hypothyroidism

MORPHOLOGICAL CLASSIFICATION OF ANEMIA

1.Microcytic Hypochromic Anemia

- MCV < 80
  MCHC < 31
  1. iron deficiency anemia :
  Chronic blood loss,
  Inadequate diet, Malabsorption, Increased demand, etc.
  2. Abnormal globin synthesis :
  Thalassemia with or
  other Hemoglobinopathies
  3. Abnormal porphyrin and heme synthesis :
  Pyridoxine responsive anemia, etc.
- 4. Other abnormal Fe metabolism :

2.Normocytic Normochromic Anemia

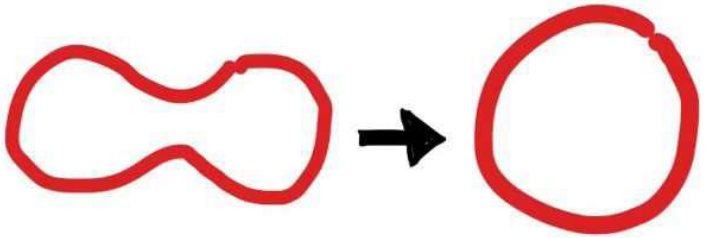
MCV 82 – 92
MCHC > 30

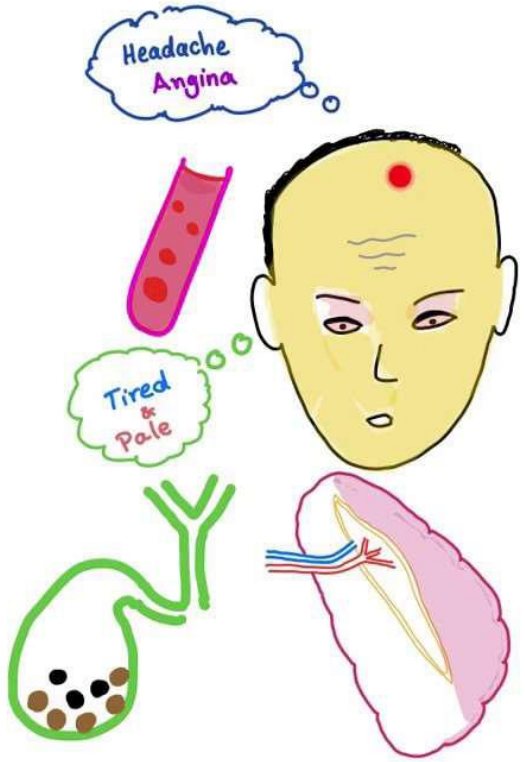
- 1. Blood loss
- 2. Increased plasma volume : Pregnancy, Overhydration
- 3. Hemolytic anemia : depend on each cause
- 4. Hypoplastic marrow : Aplastic anemia, RBC aplasia
- 5. Infiltrate BM : Leukemia, Multiple myeloma, Myelofibrosis, etc.
- 6. Abnormal endocrine : Hypothyroidism, Adrenal insufficiency, etc.
- 7. Kidney disease / Liver disease / Cirrhosis

3.Macrocytic Anemia

- MCV > 94
  MCHC > 31
- (Megaloblastic dyserythropoiesis)
  1.Vit. B12 deficiency : Pernicious anemia
  2. Folic acid deficiency :
  CAUSES ; Nutritional megaloblastic anemia, Sprue,
  Other malabsorption
  Inborn errors of metabolism : Orotic aciduria,
  Abnormal DNA synthesis : Chemotherapy, Anticonvulsant, Oral contraceptives

Hereditary Spherocytosis





HEREDITARY SPHEROCYTOSIS(HS)

Definition

HS is an inherited disorder caused by intrinsic defects in the red cell membrane skeleton that render red cells spheroid, less deformable, and vulnerable to splenic sequestration and destruction

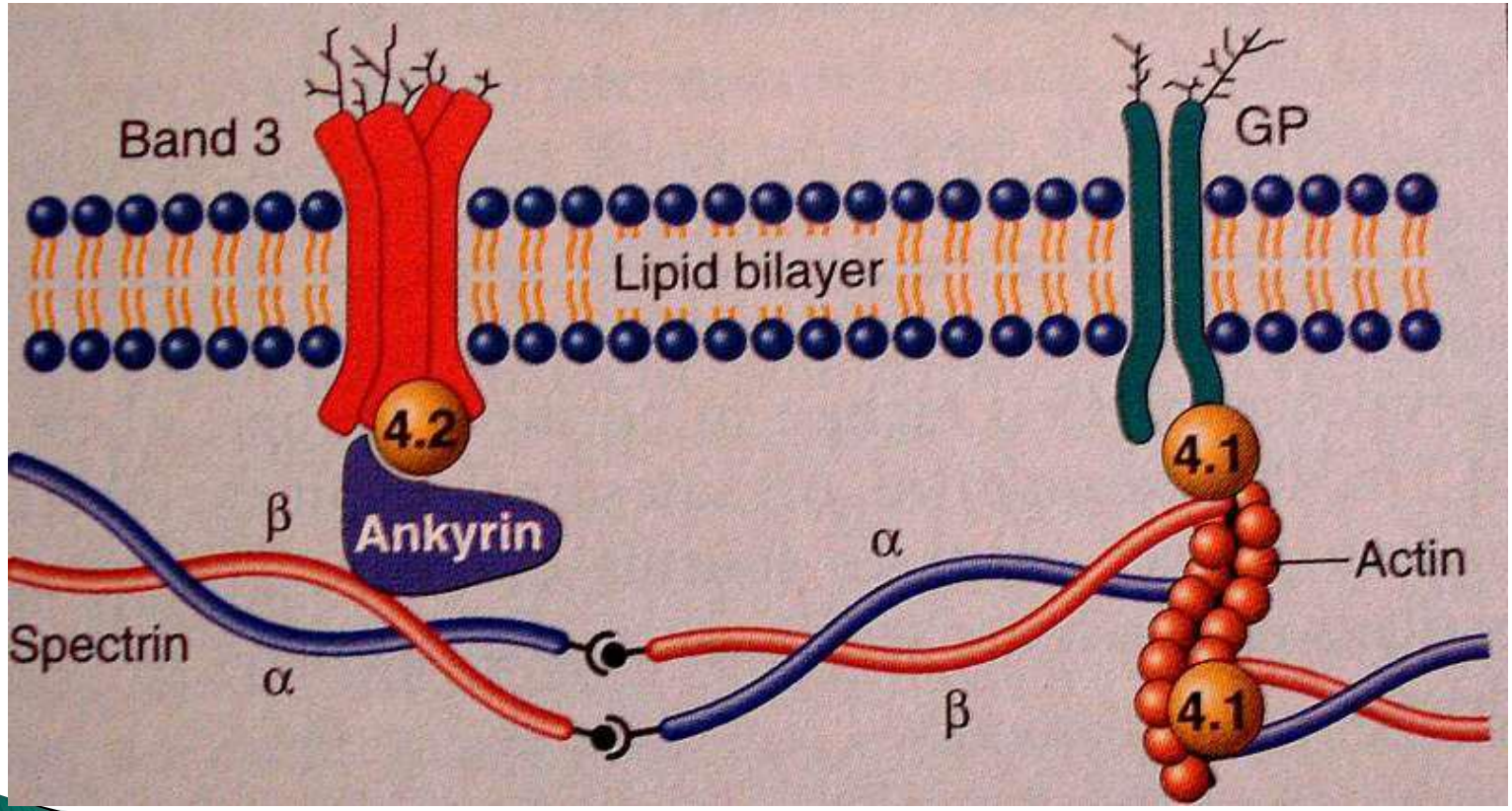
Red cell membrane

- The red cell membrane consists of:
  - Proteins 50%
  - Lipids 40%
  - Carbohydrates: 10%

Functions of red cell membrane

- Erythrocyte membrane that is normal in structure and function is essential to survival of red cell
- Maintains stability and normal discoid shape of cell
- Preserve cell deformability
- Retain selective permeability

Normal structure of red cell membrane



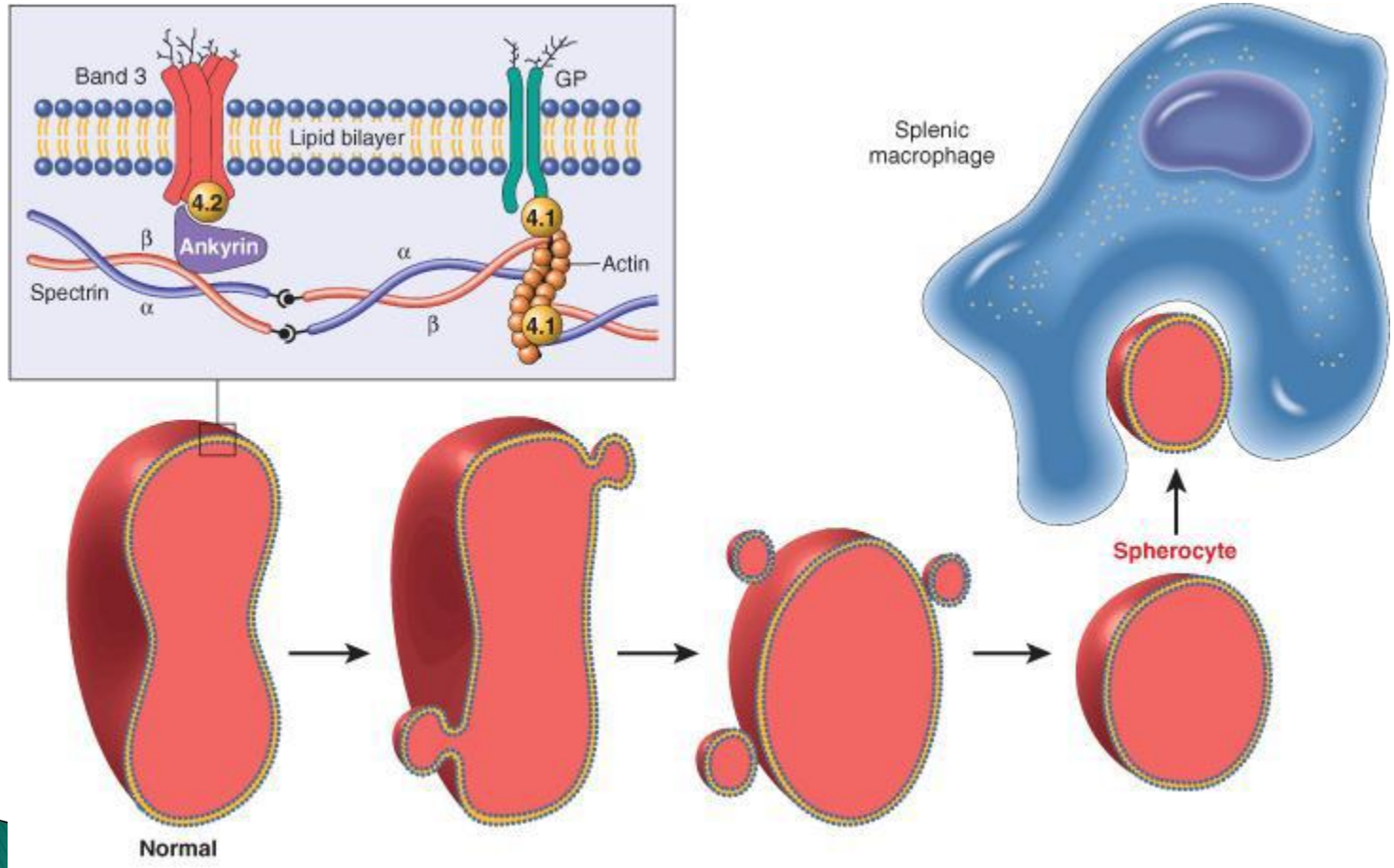
Pathophysiology of HS

- Defect in the proteins which anchor the lipid bi layer with the underlying cytoskeleton.
    1. Defect in the Spectrin
    2. Defect in the Ankyrin
- These defects results in unstable membrane but with normal volume.

Pathophysiology of hs

- They lose membrane further when in the circulation
- **Assume Spheroidal shape and**
- **smaller size (Microspherocytes)**
- **Reduced cellular flexibility**
- **Destroyed in the spleen**

spherocytosis



Clinical features

- May be clinically apparent at any age from infancy to old age
- Equal sex incidence
- Family history may be present
- 1. Anemia – mild to moderate degree
- 2. Splenomagaly
- 3. Jaundice
- 4. Pigment gallstones

Routine chemistry testing

- Bilirubin: Increased
  [Indirect bilirubin]
- Urine urobilinogen: increased
- Methaemalbumin: Increased
- LDH: Increased
- Haptoglobin: Decreased
- Haemopexin: Decreased

Diagnosis

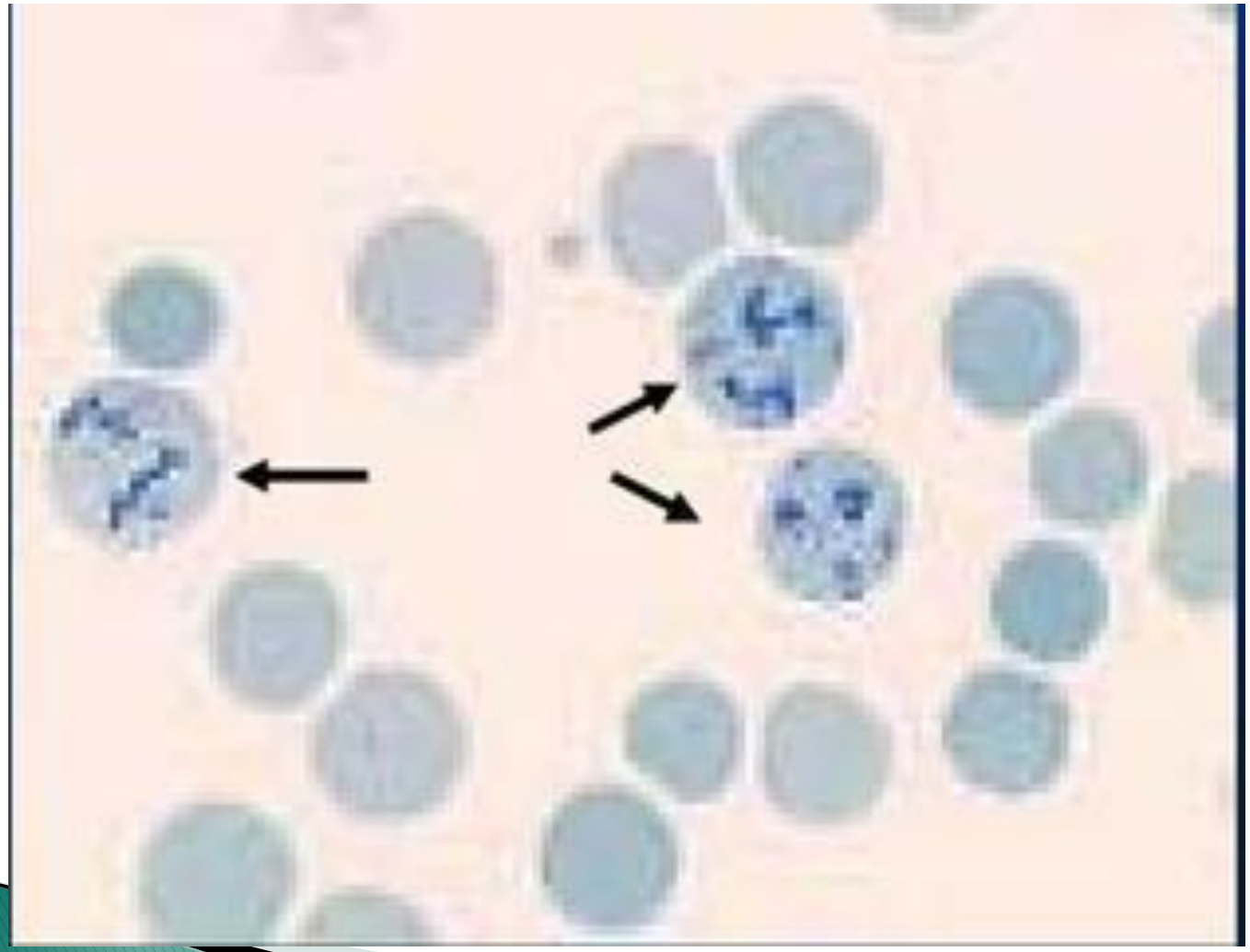
- BLOOD PICTURE
- Hb = 7-14g /dl

  [May fall below 7 in crises or May be normal in compensated hemolysis
- MCV =N or slightly reduced
- MCH= Normal
- MCHC= Increased
- Plt = normal or decreased

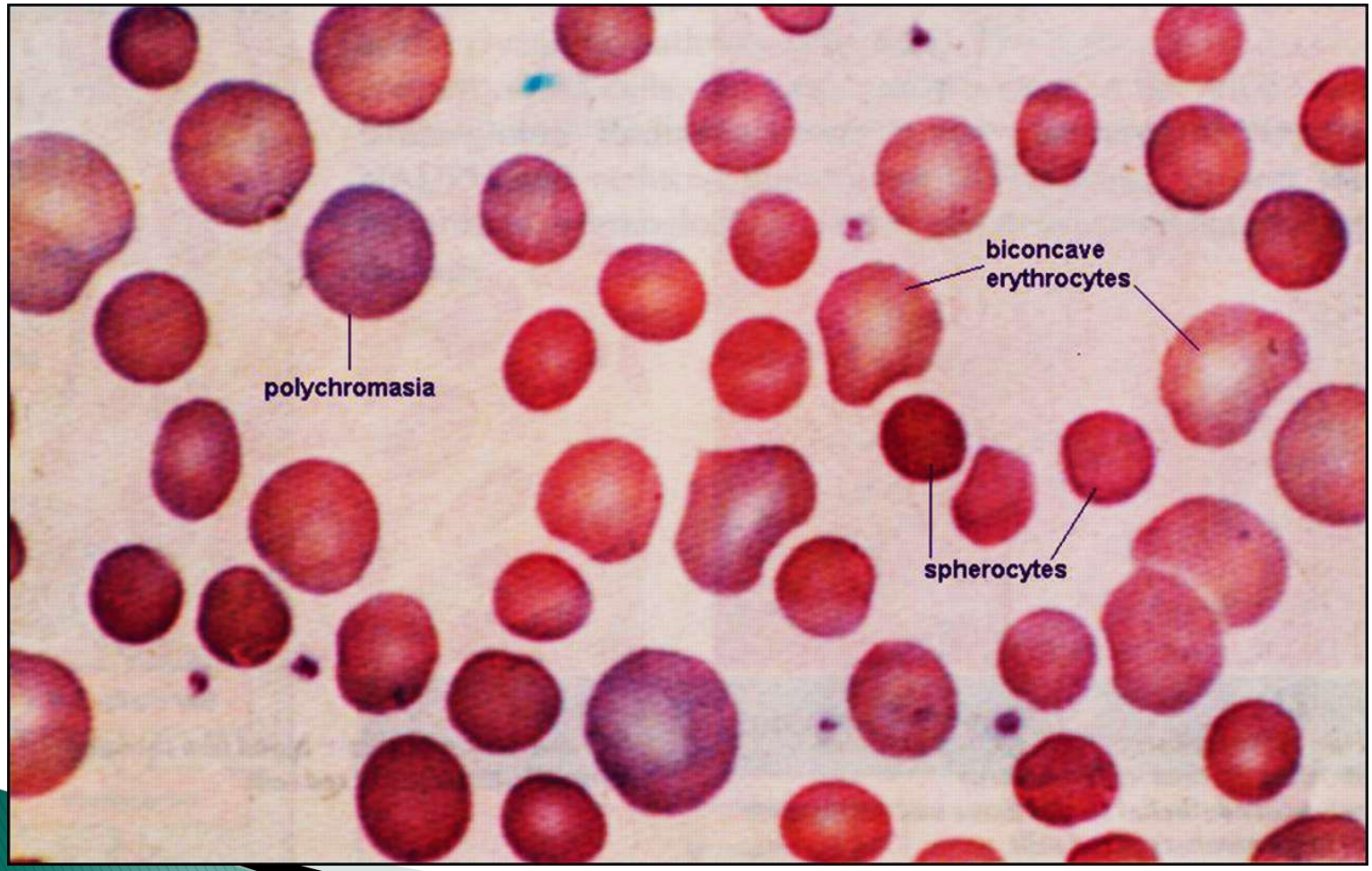
Peripheral smear

- ·ANISOCYTOSIS
- ·Spherocyte ++
- ·POIKILOCYTOSIS
- ·Polychromatic cells
- ·Target cells
- ·Nucleated RBC can be present severe cases
- ·Plt decreased if splenomegaly
- ·Retic Usually 5-20%
- ·Sometimes 50-70%

Reticulocytosis in hs



Blood smear morphology in hs



Diagnosis of HS

- OSMOTIC FRAGILITY
- Increased in Hereditary Spherocytosis

- **Red cell membrane detection (definitive dx)**
- Red cell membrane can be detected by SDS –PAGE electrophoresis

Treatment

1. Blood transfusion
2. Splenectomy

Thank you