SICKLE CELL ANAEMIA

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Course title: Advanced Hematology

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Epidemiology

- Hemoglobin S is the most common hemoglobinopathy worldwide.
- It is found most frequently in equatorial Africa and in other parts of the world.
- These are areas where falciparum malaria is endemic, suggesting that hemoglobin S arose as a protective mechanism against malaria.

Pathophysiology

- Sickle cell anemia results from a structural defect in β globin chain of the Hb.
- The abnormality in Hb S is substitution of valine for glutamic acid at the sixth amino acid position (β6 Glu→Val).
- Deoxygenated hemoglobin S tends to

polymerize into long rigid structures, which distort the cell into the characteristic sickle shape.

- Anything that causes deoxygenation of hemoglobin causes sickling, including hypoxia, acidosis, and increased temperature.
- The sickle cells are rigid and obstruct small blood vessels, resulting in tissue infarction.

Clinical Types

- 1- **Heterozygous Hemoglobin S (Sickle Trait)**
- **2- Homozygous Hemoglobin S (Sickle Cell Anemia)**

Heterozygous Hemoglobin S (Sickle Trait):

- People who are heterozygous for hemoglobin S (hemoglobin AS) are generally:
- asymptomatic,
- have a normal Hb level and complete blood count (CBC), and …
- have a normal life span.

Homozygous Hemoglobin S (Sickle Cell Anemia)

- The severity of illness in sickle cell anemia (hemoglobin SS) is variable.
- Many children become symptomatic in infancy after 3 to 4 months of age (before that time they are protected by the high levels of Hb F).

- Other people have very mild disease and may not be diagnosed until adulthood.
- This variability may be attributed to the levels of hemoglobin F and other factors.

Other Sickle Cell Diseases

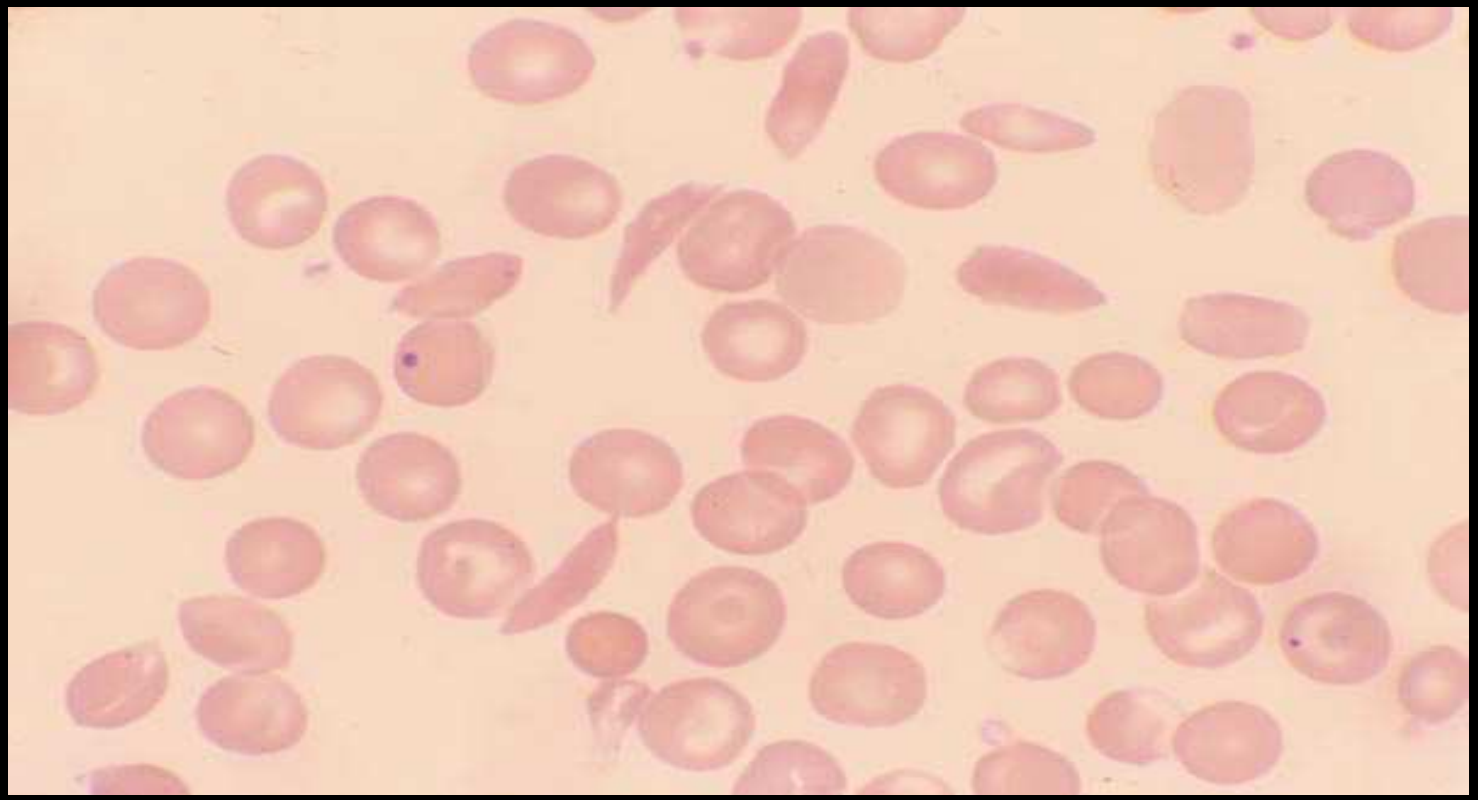
- Hemoglobin S\C,
- hemoglobin S\D
- hemoglobin S\β-thalassemia and …
- Others.
- Patients are often symptomatic and may suffer complications as homozygous sickle cell anaemia.

Diagnosis of Sickle Cell Anaemia

- **1- Sickle trait (HbAS):**
- **have a normal CBC and blood smear.**
- **Hb level is typically 5 to 8 g/dL.**
- **MCV is normal.**

2- Sickle Cell Anemia (HbSS)

- **The blood smear shows:**
- **1- target cells**
- **2- sickle cells**
- **3- Cells with various other shapes may also be seen (poikylocytosis).**
- **4- Howell-Jolly bodies (present after splenic infarction).**
- **5- Nucleated red blood cells**



Sickling and solubility tests

- These depend on the decreased solubility of deoxygenated hemoglobin S.
- Positive in patients with both sickle trait and sickle cell anemia.

Hemoglobin electrophoresis

- This test is used to separate the hemoglobin molecules based on their size and charge.
- It can also be used to distinguish between sickle trait and sickle cell anemia.

THANK YOU