



- **Definition:**

- It is an inherited autosomal recessive disease in which there is a substitution in the 6th amino acid of β -globin chain (valine instead of glutamic acid) resulting in the formation of the abnormal Hb-S.

- **Pathophysiology:**

- Factors which result in hypoxia (such as dehydration or exercise) lead to sickling of RBCs with the abnormal Hb-S → this in turn forms aggregates which will block microcirculation and cause ischemia/infarction of different organs (this is known as sickle crisis).

- **What are the different types of sickle crisis?**

- **Vaso-occlusive crisis:** characterized by pain which results from necrosis.
- **Hemolytic crisis:** characterized by \downarrow Hb.
- **Aplastic crisis:** resembling bone marrow suppression and associated with parvovirus infection.
- **Sequestration:** in the spleen.
- **Acute chest syndrome:** caused mainly by Streptococcus pneumonia.

- **What are the complications associated with SCD?**

- Bony abnormalities (e.g. osteonecrosis of femoral and humeral heads).
- Renal medullary damage.
- Autosplenectomy.
- Hepatomegaly.
- Ventricular hypertrophy.
- Pulmonary infarctions.
- Cerebrovascular accidents.
- Leg ulcers.
- Propensity to infection and sepsis with gram-positive as well as gram-negative organisms

- **Pregnancy and sickle cell disease:**

- **Diagnostic test:** hemoglobin electrophoresis will differentiate between SA trait (<40% hemoglobin S) or SS disease (>40% hemoglobin S).
- **Effects of pregnancy on SCD:**
 - ✓ \uparrow risk of UTIs and hence crisis.
 - ✓ Nausea and vomiting in pregnancy can lead to dehydration which predisposes to crisis.
 - ✓ Stress condition associated with pregnancy → \uparrow crisis.
- **Effects of SCD on pregnancy:**
 - ✓ Spontaneous abortions, IUGR, fetal death or pre-term delivery.
 - ✓ Increased risk of DVT and pulmonary embolism post-partum (most common cause of maternal death in SCD).
- **Management:**
 - ✓ Avoid hypoxia and dehydration.
 - ✓ Take folate supplements.
 - ✓ Monitor fetal growth and well-being.
 - ✓ Blood transfusion: when Hb < 7 with presence of symptoms.
 - ✓ Delivery: managed the same way as for women with cardiac disease. Epidural analgesia is ideally suited for labor and delivery. Compatible blood should always be available.