Sickle Cell Anaemia and Surgery

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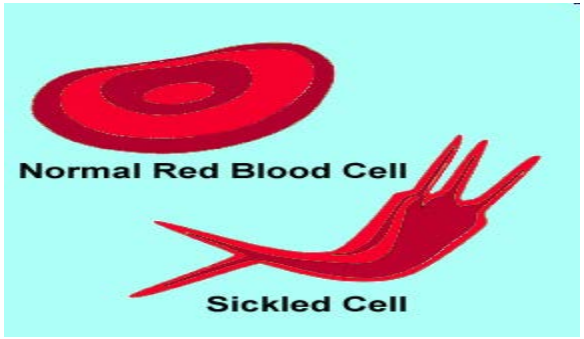
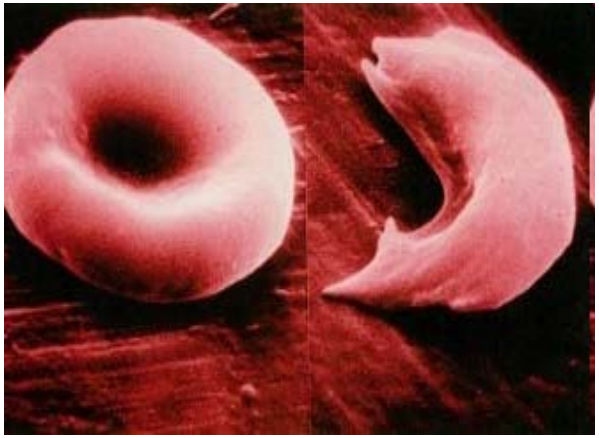
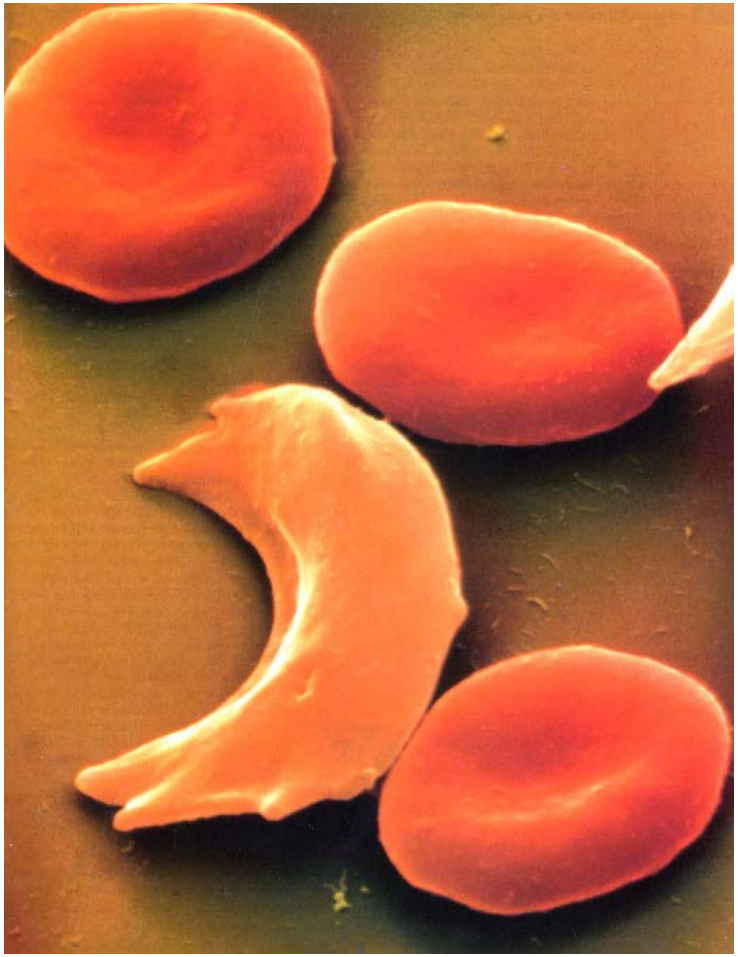
SCD: Genetic Basis

- An abnormal sickle-shaped haemoglobin (Hb-S).
- Genetic defect:
  - Substitution of valine for glutamic acid at 6th position of the beta globin chain.
- When deoxygenated:
  - Red cells undergo "sickling" due to polymerization of Hb-S.
- Subsequent repeated haemolysis.

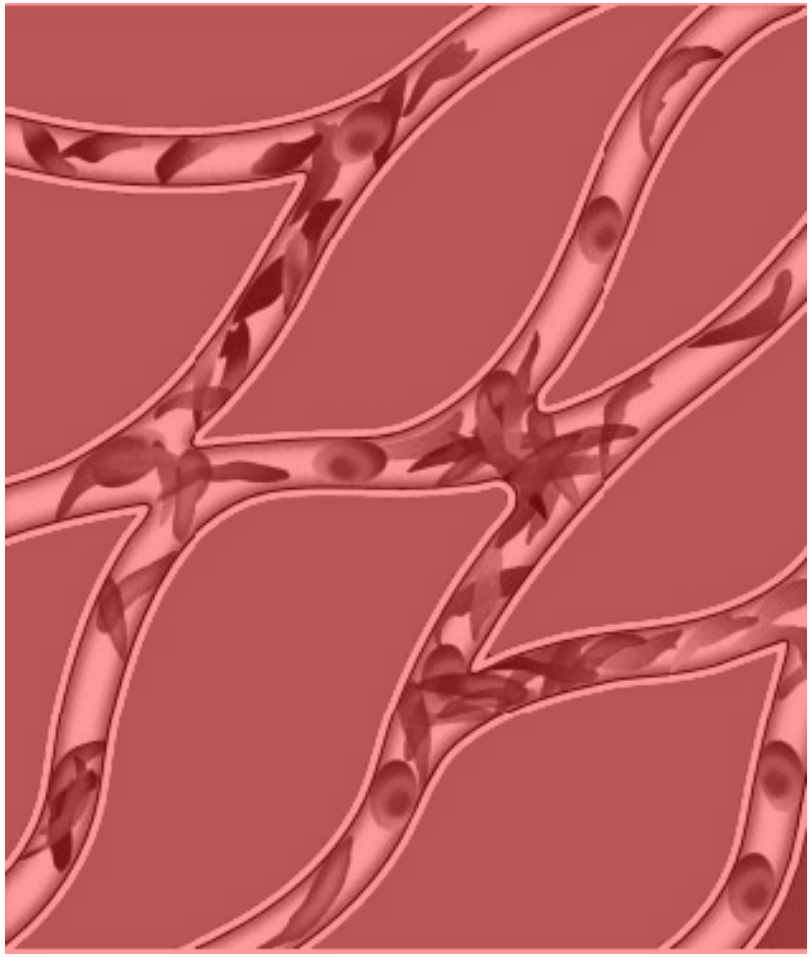




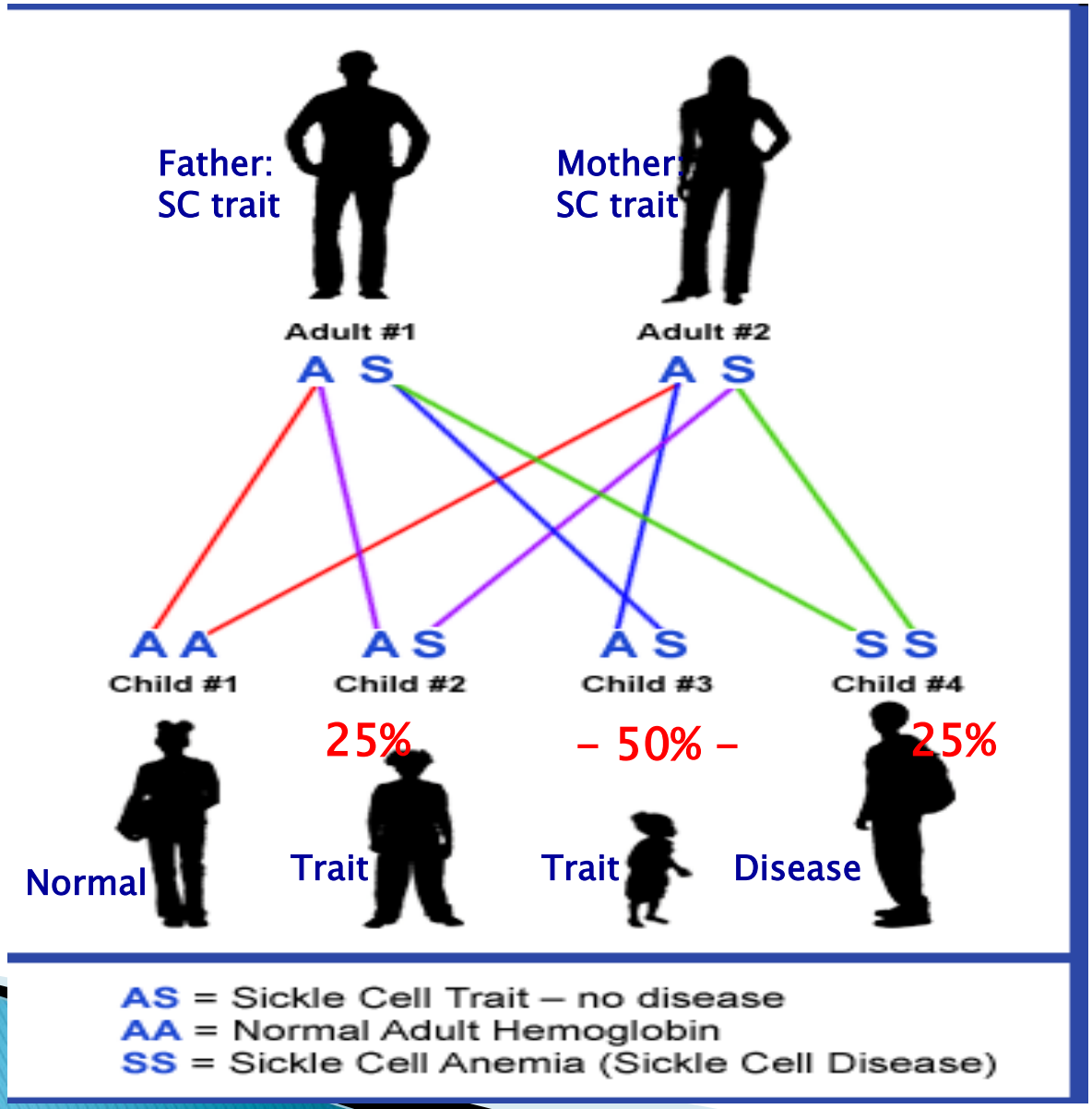
Sickle cell



Sickle cell microcirculation

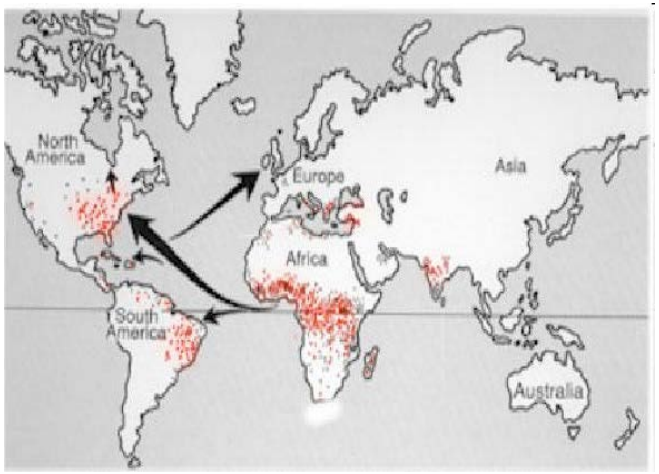


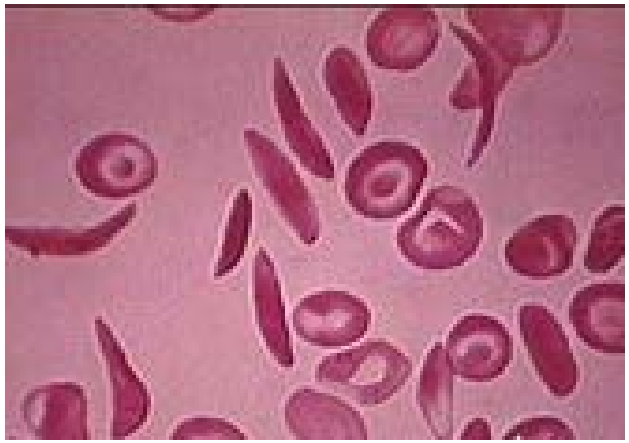
SCD: Inheritance



SCD: Epidemiology

- Prevalent in certain areas of the world:
  - Highly prevalent in malarial equatorial Africa
  - The sickle cell gene is also seen in:
    - North and South America,
    - UK,
    - The Caribbean,
    - Some Mediterranean countries
    - Middle east
    - Central India.





SCD: Clinical Manifestation

- **Quite variable and includes:**
  - Repeated painful vaso-occlusive crises,
  - Haemolytic episodes.
  - Sequestration crises.
- **Complications affect various systems mainly:**
  - Skeletal
  - Gastrointestinal
  - Spleen
  - Hepato-biliary
  - Cardiopulmonary
  - Central nervous system (CNS).

SCD: Aetiology of Clinical Manifestation

- **Chronic hypoxia may eventually damage:**
  - Heart: high output cardiac failure
  - Kidneys: chronic renal failure.

- **Susceptibility to infections with certain organisms:**
  - defective complement activation
  - asplenia or autosplenectomy.
  - impaired neutrophil function.

Sickle Cell Anemia Crisis

- **Vaso-occlusive crisis**:
  A vaso-occlusive **crisis** occurs when the microcirculation is obstructed by sickled RBCs, causing ischemic injury to the organ supplied. Occurs most commonly in bones and viscera organs

- **Sequestration crisis**
  These are caused by sickling within organs and pooling of blood, often with a severer exacerbation of anemia. Most common il lung, liver and the spleen

Sickle Cell Anemia Crisis (Cont.)

- **Aplastic crisis**
  may result from infection with parvovirus (B19) or folate deficiency and characterized by a sudden fall in hemoglobin and reticulocyte count.

- **Hemolytic crisis**
  Hemolytic crises are manifested by a sudden exacerbation of **anemia,** due to increased rate of haemolysis with a fall in haemoglobin level, patient usually present with severe jaundice but no pain. Hemoglobin falls, but reticulocyte count increases

SCD: Surgical Implications

- Surgery is associated with high morbidity & mortality.

- A special multidisciplinary care is needed in management of patients presenting to various surgical specialties.

SCD: Surgical Implications

Certain SCA crises may mimic surgical emergencies

SCA Crises	Surgical Condition
Vaso-occlussive crises	Acute Abdomen
Bone pain crises	Acute osteomyelitis
Acute chest syndrome	Pulmonary embolism
Acute splenic sequestration crises	Splenic abscess

It is important to recognize such conditions because surgeries may worsen them

SCD: Surgical Implications

- Surgical interventions are necessary for some complications of SCA. For example
  1. Splenectomy might be the best choice if more that two crisis of acute splenic sequestration occur.
  2. Skin grafting for chronic leg ulcers

SCD: Surgical Implications

- Specific attention should be given to SCA patients in the peri-operative period in order to prevent complications.

- Particularly important in procedures that are associated with ischemia or hypoxia.

SCD: Surgical Implications

- Adverse factors that may precipitate vaso-occlusive & sequestration crises:
  - **H**ypothermia.
  - **I**nfection.
  - **D**ehydration.
  - **A**cidosis.

SCD: Surgical Implications

- Additional risks in surgery are due to changes in:
  - Operating room temperature,
  - pH,
  - Oxygen tension,
  - Fluid volume.
  - Circulatory stasis
  - Suboptimal ventilation during surgery.

SCD: Preoperative Preparation

- Admitted 1-2 days earlier.
- Estimation of predicted operative risk & postoperative complications. To be based on:
  - Prior knowledge of the sickling history.
  - Severity of SCD activity: frequency and severity of complications.
  - PCV
  - Complexity of the surgical procedure.

SCD: Intraoperative Care

- Increase inspired oxygen concentration during surgery.
- monitor PO_2 by pulse oximetry.
- Avoid circulatory stasis:
  - adequate hydration.
  - prompt replacement of intraoperative fluid losses.
  - avoid acute hypovolemia.
- Avoid hypothermia or hyperthermia.
  - Keep the OR temperature near normal.

SCD: Postoperative Care

- Nursing in ICU is rarely needed.
- 24-hr Oxygen supplementation via mask.
- Adequate I.V fluid hydration.
- Thromboprophylaxis.
- Adequate analgesia.

Surgical Implications: conditions that may simulate surgical conditions or need surgery

- Abdominal Pain.
- Cholelithiasis.
- Splenic complications.
- Peptic ulceration.
- Ischaemic bowel.
- Leg ulcers.

Surgical Implications: Abdominal pain in SCD

- Common.
- Mimics many surgical emergencies.
- Careful clinical & radiological evaluation is essential.
- Avoid unnecessary surgical intervention.
- Acute pain often requires hospital admission for parenteral opioids & hydration.

Differential diagnosis of abdominal pain in SCD

Splenic causes

- Acute splenic sequestration crisis
- Splenic infarction
- Splenic abscess

Hepatobiliary causes

- Biliary colic
- CBD stones/polyp
- Acute cholecystitis
- Hepatic crisis
- Hepatitis
- Liver abscess

Other surgical conditions

- Acute appendicitis
- Acute pancreatitis
- Peptic ulcer disease
- Ischaemic colitis

Other causes

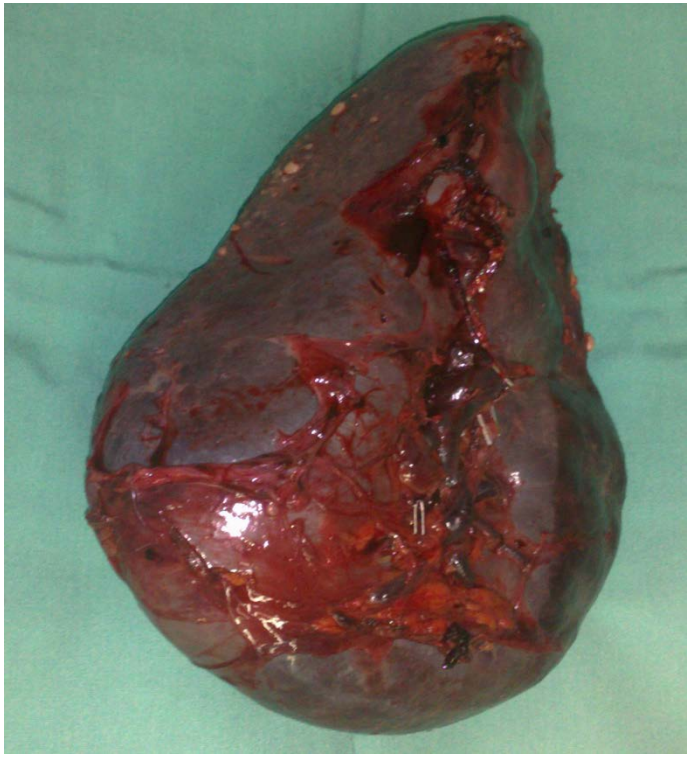
- Vasoocclusive crisis
- Acute chest syndrome
- Bone marrow infarction of vertebral bodies
- Nerve root entrapment
- Avascular necrosis
- Vertebral collapse
- Enlarged mesenteric lymph nodes
- Enlarged retroperitoneal lymph nodes

Splenic Complications

- Repeated splenic infarction.
- The spleen often becomes a fibrotic nodule.
- Patients with high Hb-F or thalassaemia:
  - have a persistently enlarged spleen
  - often affected with:
    - repeated acute sequestration crises.
    - Repeated infarctions.
    - infection of an infarcted area with abscess formation.

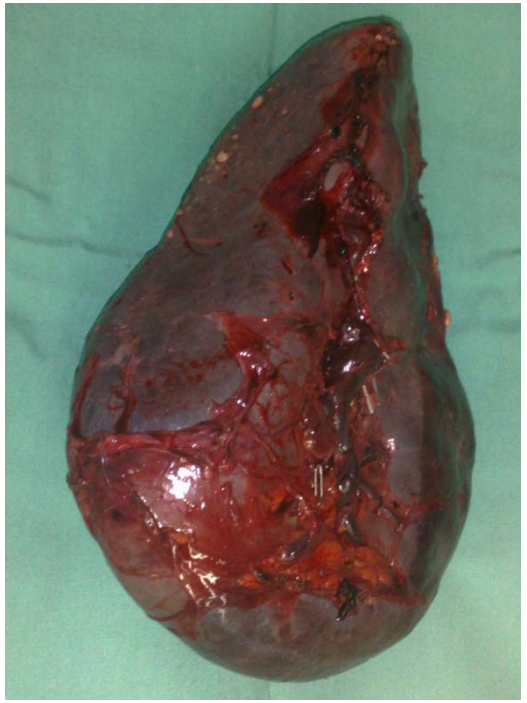
Indications for Splenectomy in SCD

- Recurrent acute splenic sequestration crises (ASSC).
- Troublesome hypersplenism with pancytopenia (anaemia, neutropenia & thrombocytopenia).
- Splenic abscess.
- Persistent massive splenomegaly: less common



Splenic complications: Acute Splenic Sequestration Crisis

- A life-threatening complication.



- The diagnosis is clinical:
  - sudden & massive enlargement of spleen.
  - rapid fall in Hb & Hct.

- Treatment:
  - **Minor crises:** resolve spontaneously
  - **Major crises:** require exchange blood transfusion.
  - 1 major or 2 minor episodes: **Splenectomy**.

Splenic complications: Splenic abscess

- SCD patients are at risk of developing splenic sepsis.
- Occurs secondary to infection of splenic infarcts.
- Ultrasound & CT Scan: crucial in the diagnosis.
- Treatment:
  - Ultrasound-guided percutaneous drainage.
  - Splenectomy (open or laparoscopic).

Hepato-biliary complications:

Cholelithiasis

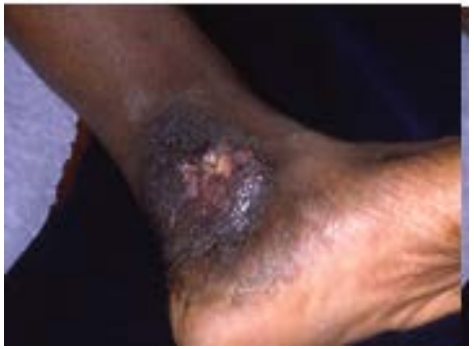
- High risk of developing pigmented gallstones.
- Chronic haemolysis.
- The incidence increases with age.
- Gallstones may be asymptomatic.

Gastrointestinal manifestations: Ischaemic colitis

- Occurs in young SCD patients.
- Diagnosis:
  - Sickling crisis.
  - Severe abdominal pain, rectal bleeding.
  - Has signs of peritonism.
- High fever, marked leucocytosis & generalized peritoneal signs suggest bowel infarction.
- Initial management:
  - NGT decompression.
  - Broad-spectrum antibiotics.
  - Haemodynamic support including exchange transfusion.
- Early surgical exploration if no improvement.

Leg Ulcers

- 10% of SCD patients will develop leg ulceration.
- Occur spontaneously or as a result of local trauma.
- No specific organisms.
- **Site:** majority located above the ankle, less commonly on the dorsum and rarely on the sole of the foot.



- The ulcer is treated conservatively by:
  - Regular daily wound dressings.
  - Bed rest.
  - Debridement
  - Topical ointments: zinc, steroids, antibiotics.
- Surgical procedures such as split skin & full-thickness grafts:
  - results have been disappointing.
  - a high recurrence rate: > 50%.

SCA: Orthopaedic manifestations

- Bone infarcts
- Osteomyelitis
- Marrow hyperplasia

Orthopaedic manifestations
Bone Infarcts

Sickling in bone marrow

↓

Stasis and sequestration

↓

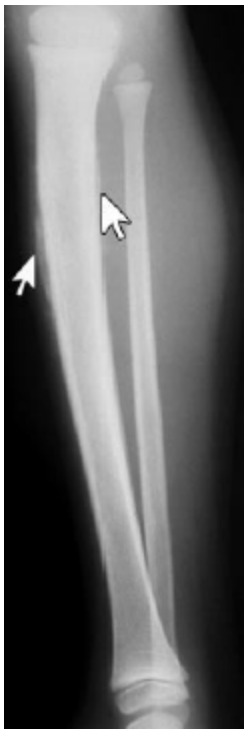
Ischaemia

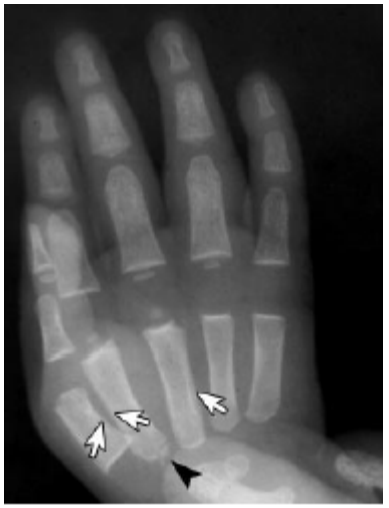
↓

Bone necrosis

SCA: Orthopaedic manifestations Bone Infarcts

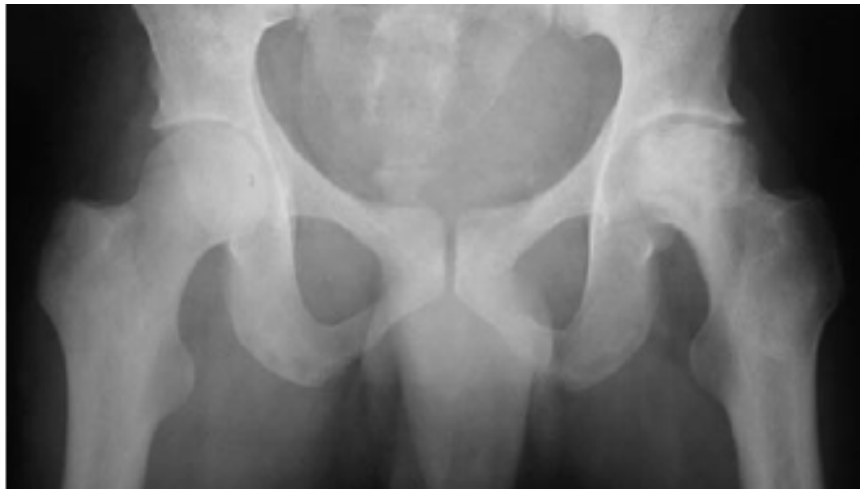
- Infants (6months-2years) presents with dactylitis or hand-foot syndrome
- Older children and adults presents with bone pain crises in diaphysis of long bones
- Avascular necrosis commonly seen in femoral and (less common) humeral heads





SCA: Avascular necrosis

- Also called osteonecrosis
- Most commonly affects the head of femur
- Less commonly; the humeral head
- AVN affects HbSC more than HbSS



SCA: Avascular necrosis

- About 50% of patients may develop AVN by the age of 35 years
- Commonly presents with limp, pain in the affected hip, and stiffness
- Collapse of the head leads to shortening

SCA: Osteomyelitis

Usually atypical because it may

- Affect any part of the bone
- Many bones simultaneously or sequentially
- Occur at any age
- They may also be associated with septic arthritis
- May be difficult to differentiate from bone infarcts
- Staph aureus is the most common organism.
- Salmonella is more common cause than in non-SCA patients

CONCLUSION

- SCD patients present with various surgical manifestations posing a formidable diagnostic & management challenges.

- Patients requiring surgical intervention need a comprehensive management plan including:
  - Adequate pre-operative planning of surgery
  - Special perioperative care.
  - Effective postoperative care and analgesia.

It is hoped that increasing awareness of the surgical manifestations of sickle cell disease and the use of preventive therapies will reduce the high morbidity and mortality associated with surgery in this high-risk group of patients.

Thank You