Surgery & Sickle Cell Disease

Sheraz Malik MB BS MRCS
Tutor

Paul Ofori-Atta MB. ChB, FRCS
President Motec Life UK
Lecture Outline

- Historical Background.
- Genetic Basis.
- Epidemiology.
- Diagnosis.
- Clinical Manifestation.
- Surgery & Anesthesia:
  - Preoperative Care.
  - Peroperative Care.
  - Postoperative Care.
- Conclusion.
SCD: Historical background

- Commonest genetic haematological disorder.

- **1670**: A history of the condition tracked back to 1670 in one Ghanaian Family.

- **1846**: Absence of a spleen in the autopsy of a runaway slave. *Southern Journal of Medical Pharmacology* 1846

- **1870s**: ‘*Ogbanjes*’ – “Children who come and go”. High infant mortality rate associated with SCD. *African Medical Literature*
SCD: Historical background

- **1910**: The first reported case was in a dental student from the West Indies. 
  Herrick JB. *Arch Intern Med* 1910

- **1922**: First use of the term 'sickle cell anaemia'.
  Mason VR. *JAMA* 1922

- **1949**: The genetic basis was demonstrated:
  - **Homozygous**: Sickle cell disease
  - **Heterozygous**: Sickle cell trait (carrier)
  Beet EA. *Ann Eur Gen* 1949
SCD: Genetic Basis

- An abnormal sickle haemoglobin (Hb-S).
- Genetic defect:
  - Substitution of valine for glutamic acid at 6\textsuperscript{th} position of the beta globin chain.
- When deoxygenated:
  - Red cells undergo “sickling” due to polymerization of Hb-S.
- Subsequent repeated haemolysis.
The susceptibility of red cells to sickle correlates well with the concentration of Hb-S.
SCD: Genetic Basis

Father:
SC trait

Mother:
SC trait

Normal Trait Trait Disease

25% 50% 25%

AS = Sickle Cell Trait – no disease
AA = Normal Adult Hemoglobin
SS = Sickle Cell Anemia (Sickle Cell Disease)
SCD: Epidemiology

- Prevalent in certain areas of the world:
  - highly malarial equatorial Africa
  - The sickle cell gene seen in:
    - North and South America,
    - UK,
    - The Caribbean,
    - Some Mediterranean countries
    - Arabian Peninsula:
      - Western region (African haplotype)
      - Eastern region (Asian haplotype).
    - Other Gulf States: Bahrain, Oman
    - Central India.
SCD is everywhere!

In recent years:

- Greater population mobility.
- SCD may be encountered anywhere in the world.
- Recognition of various manifestations important.
In the USA where there is no endemic malaria the prevalence of SCD in blacks is lower (2.25%) than in West Africa (4.0%), and is falling.

Without endemic malaria from Africa, the condition is purely disadvantageous and will tend to be bred out of the affected population.
SCD: Diagnosis

Normal Blood Smear

SC Blood Smear
SCD: Clinical Manifestation

- **Quite variable and includes:**
  - Repeated painful vaso-occlusive,
  - Haemolytic episodes.
  - Sequestration crises.

- **Complications affect various systems mainly:**
  - Skeletal
  - Gastrointestinal
  - Spleen
  - Hepato-biliary
  - Cardiopulmonary
  - Central nervous system (CNS).
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 neutrophils function
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.
Adverse Factors that Precipitate Vasoocclusive & Sequestration Crises

- Hypothermia.
- Infection.
- Dehydration.
- Acidosis.
General anaesthesia (GA) & surgical trauma add additional complications risk.

- Changes in
  - OR temperature,
  - pH,
  - Oxygen tension,
  - Fluid volume.
  - Circulatory stasis
  - Suboptimal ventilation during surgery.

Hb–S polymerizes in the capillaries with subsequent ischaemic infarcts in many tissues.
SCD: Surgical Implications

- Reported morbidity after major surgeries approaches 40%.

- This can be decreased by:
  - understanding the pathophysiology of SCD.
  - prophylactic preoperative blood transfusions.

SCD: Preoperative Preparation

- Admitted 1–2 days earlier.
- Estimation of predicted operative risk & postoperative complications:
  - Prior knowledge of the sickling history.
  - Severity of SCD activity: frequency & complications.
  - Actual Hb–S level: indicated by electrophoresis.
  - Complexity of the surgical procedure.
A decision on a preoperative transfusion regimen is made.

It reduces the perioperative risks associated with surgery & anaesthesia.

SCD: Preoperative Transfusion

Various Transfusion Regimens

**Conservative regimen:**
- correcting the anemia by simple top up transfusion irrespective of Hb–S concentration.

**Aggressive regimen:**
- Exchange Transfusion.
- used to reduce Hb–S to <35%.
- diminishes blood viscosity
SCD: Preoperative Transfusion

Randomized trials:

Conservative (simple) transfusion regimen:

- as effective as aggressive ones in decreasing perioperative complications.
- No differences in perioperative complications.
- Associated with half as many transfusion-associated complications.

Preoperative Transfusion Policy: Guidelines

- For minor surgical procedures:
  - Conservative regimen is advocated.

- Exchange transfusion is reserved for:
  - Those undergoing major surgery especially in presence of severe SCD as judged by:
    - frequent major crises
    - systemic SCD complications.
    - High Hb-S.

- Avoid over-transfusion.
SCD: Preoperative Care

- Avoid too much preoperative sedation.

- Administer prophylactic antibiotics.

- Adequate intravenous hydration:
  - especially on the night of the planned operation.

- Thromboprophylaxis:
  - LMWt or standard heparin.
  - except for patients who have received preoperative exchange transfusion.
SCD: Intraoperative Care

- Increase inspired oxygen concentration during surgery.
- Monitor PO2 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.

Firth PG, Head A. Anesthesiology 2004
Marchant W, Walker I. Paediatric Anaesthesia 2003
SCD: Postoperative Care

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

Adequate analgesia.

Postoperative Analgesia

- Effective postoperative pain control is essential but a daunting task.

- Successful pain control is often hampered by:
  - Misperceptions about the cause of the pain
  - Fear of addiction.

- Drugs:
  - **NSAI drugs**: excellent adjunct to opioids.
  - **Epidural analgesia**: minimize respiratory depression.
  - The use of **patient controlled analgesia devices** is effective for improved narcotic delivery.

SCD: Postoperative Complications

- Proper perioperative care minimizes postoperative complications.

- 2 most feared complications:
  - Acute chest syndrome.
  - Vasoocclusive crisis.
CONCLUSION: I

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

- Patients requiring surgical intervention need a comprehensive management plan including:
  - Preoperative blood transfusion.
  - Special perioperative care.
  - Effective postoperative analgesia.
CONCLUSION: II

- It is hoped that:
  - Increasing awareness of the surgical manifestations of SCD,
  - Better peri-operative patient care,
  - Adequate analgesia.

- will reduce the high Morbidity & Mortality associated with surgery in this high risk group of patients.