

## **CASE REPORT**

# **Sickle Cell Anaemia And The Anaesthetic Management**

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### **ABSTRACT:**

Sickle cell anaemia is a common hereditary, autosomal recessive, haemolytic anaemia in tropical Africa or West Indian states. It is also fairly common in Kingdom of Saudi Arabia. These patients require special and meticulous care during anaesthesia, e.g., avoidance of hypoxia, hypercarbia, acidosis, fever, hypothermia and dehydration. Successful management requires early detection and scrupulous technique.

**Key Words:** Haemoglobin, Haemoglobinopathies, Sickle cell, crisis.

### **CASE REPORT**

A six years male Saudi student was admitted for adenotonsillectomy at MH Al-kharj, Saudia Arabia. On preanaesthesia checkup, he was found to be sickle cell Positive. Systemic examination was unremarkable. His Hb was 11.1G% & Hct 35 % . on hemoglobin electrophoresis he had 60.7 % Hb-S [Hb.SS] . Blood group complete Picture, urine RE , serene chemistry, x-ray chest , acid base status were all within normal range . He was ASA – II with Mallampatti classification I and Wilson risk index O for airway assessment .

Patient was kept NPO after midnight but clear fluids were allowed till 4 am in the morning. Syp. Phenergan was given as premedication at night. To prevent dehydration a 5% D/W drip was started at 25 ml / hr at 6 am in the ward on the day of surgery . Operation was planned early in the morning.

Operating room was kept warm. Child was preoxygenated and anaesthesia was induced with propofol. A non-cuffed orotracheal tube was passed and secured properly in place. Anaesthesia was maintained with O<sub>2</sub>/N<sub>2</sub>O, halothane and fentanyl. Inj. atracurium was used as muscle relaxant and patient was ventilated on Servo ventilator 900C. Pulse, ECG, NIBP, ETCO<sub>2</sub>, SpO<sub>2</sub> and temperature were monitored intra-

operatively. Ringer's solution 100ml was given during operation, which lasted for about 45 min. Patient made an uneventful recovery. Supplemental oxygen was given for a prolonged period in the recovery room and it was continued for 24 hrs in the ward. Narcotics were avoided postoperatively. Patient was discharged on second post op day with advice for follow up which was uneventful.

### **DISCUSSION:**

Normal adult haemoglobin (Hb A) contains two alpha (a) and two beta (b) polypeptide chains. If the polypeptide chains are abnormal, so is the haemoglobin. The sickle cell haemoglobin Hb S arises from a mutation in the beta globin gene so that glutamic acid in the 6<sup>th</sup> position of beta chain is replaced by valine, whereas in HB C it is replaced by lysine. Alternatively, there may be defective production of qualitatively normal chains, resulting in a- thalassaemia (a 0 chain deficiency) or B-thalassaemia ( B chain deficiency ). In sickle cell disease (HbSS), the patient is homozygous for the abnormal gene whereas in sickle cell trait (HbAS), the patient is heterozygous for the gene.

The condition is most common in West Central Africa, North Saudi Arabia, and East Central India but has been described in Southern Mediterranean population. Incidence of HbSS in US blacks is under 1% and incidence of HbSS is 8-10%. In Saudi Arabia the overall incidence of sickle cell haemoglobinopathy is 20-30%.

### **SICKLING:**

When Hb is deoxygenated it becomes much less soluble and forms long crystals or tactoids, which distort the erythrocytes, producing the sickle shape. These sickle cells tend to aggregate in the capillaries and venules obstructing the flow of blood and so causing zones of infarction. The sickling phenomenon is dependent upon:

1. Percentage of Hb S in the red cells.



- Oxygen tension.
- pH.
- Nature of other haemoglobin present.

#### PERCENT OF HBS IN THE RED CELL

The higher the proportion of Hb S the more likely the cell to sickle. A simplified table of the percentages of the haemoglobin present in the various sickle cell syndromes is given in table.

#### OXYGEN TENSION

A red cell will sickle at a particular level of deoxygenation. Approximate values of PO<sub>2</sub> at which sickling

Genotype	% Hb-S	% Hb-C	% Hb-F	% Hb-A	Total Hb G/dl
SS	90-95	-	5-10	-	5-10
SA	25-45	-	< 3	55-75	normal
SC	50	50	< 3	-	10-14
S Bet Thal	70-100	-	5-10	0-30	8-14

occurs are as follows:

Sickle cell anaemia (SS)	40-50 mm Hg
Sickle cell Hb C disease (SC)	30 mmHg
Sickle cell trait (Hb SA)	20 mmHg

#### pH

A reduction in pH causes the Hb dissociation curve to shift to the right (Bohr effect) so that for a given PO<sub>2</sub> the haemoglobin is more deoxygenated. Any condition producing acidosis, whether it be respiratory or metabolic acidosis will cause sickling of red cells.

#### NATURE OF OTHER HAEMOGLOBIN PRESENT

Sickling is more likely to occur in the presence of Hb C than in the presence of Hb A or Hb F. In fact Hb F has been proved to have a protective role against sickling.

#### DIAGNOSIS OF SICKLE CELL SYNDROMES

- Anaemia with the presence of sickle cells or target cells.
- Sickledex Test. This test detects Hb S by pre-

cipitation.

3. Sodium metabisulphite. If RBC's which contain Hb S are made grossly hypoxic by suspending them in a sodium metabisulphite they will sickle.

4. Haemeoglobin electrophoresis. This is the definitive diagnostic test for sickle cell variants.

#### CLINICAL FEATURES

Sickle cell trait remains usually asymptomatic though sickling can occur under hypoxic conditions and there is an increased incidence of pyelonephritis, haematuria and renal papillary necrosis.

The sickle cell crises in three varieties of sickle cell disease may take several forms.

#### Haemolytic Crisis.

This is precipitated by infection and is associated with a sudden further reduction in the red cell lifespan. The lifespan in sickle cell anaemia (SS) may be as little as 7 days.

#### Infarctive Crisis.

This is the commonest presentation after trauma surgery. Sickling causes sludging of red cells within small vessels and leads to infarction especially of lungs, spleen and bones, though any organ may be affected, by causing microvessel occlusion.

#### Aplastic Crisis.

Marrow depression often associated with viral infection or folate deficiency causes an acute aplastic anaemia. This complication is relatively uncommon.

#### Sequestration Crisis.

There is enlargement of spleen with pooling of red cells. This type of crisis usually affects young children and infants and may necessitate immediate transfusion as large amounts of blood are sequestered in spleen as well as liver. A picture of acute hypovolaemic shock may be presented.

Clinical features of sickle cell anaemia (SS) which may be present include:

- \* History of bone and joint pains, osteomyelitis.
- \* Bossing of skull with overgrowth of maxilla.
- \* Leg ulcers.
- \* Enlarged Liver
- \* Jaundice due to haemolysis, gallstones.
- \* Haematuria.
- \* Priapism.



## ANAESTHETIC CONSIDERATIONS

Blood of all the African patients and those originating from countries bordering the Mediterranean should be screened for Hb S prior to anaesthesia; this does not apply to neonates. If Hb S is detected electrophoresis must be carried out. In an acute emergency there may not be time for electrophoresis, although very rapid electrophoresis techniques are available now. As mentioned above, anaemia and the presence of target cells are indicative of one of the sickle cell disease, rather than the trait.

### **SICKLE CELL TRAIT.**

No specific precautions are needed, although gross hypoxia may cause sickling and must be avoided at all cost.

### **SICKLE CELL DISEASE.**

Certain precautions should be taken in order to reduce the risk of anaesthesia to minimum. No patient should be anaesthetized during a crisis except life saving operations. Infection is treated and folate deficiency dealt with.

Pre-operative transfusion is needed if Hb is less than 6 g/dl. If blood transfusion is likely to be necessary during operative fresh blood should be cross-matched.

Patient is starved in the usual way but it is advisable to avoid dehydration. The use of sodium bicarbonate has been recommended in an effort to avoid acido-

sis.

For elective surgery, total red cell exchange is recommended, if possible by use of cell separator. The concentration of sickle haemoglobin should be less than 10 percent and the haemoglobin concentration is raised to normal values. Exchange transfusion in the third trimester of pregnancy is also valuable. This can simply be achieved by 2 pints mini exchanges at fortnightly intervals starting at 26-28 weeks of gestation.

Tourniquets and intravenous regional techniques are contraindicated. Preoxygenation is a must. During anaesthesia it is important to avoid hypoxia, hypercapnia, vasoconstriction, reduction in cardiac output and hypothermia. Moderate hyperventilation should be ensured. IV hydration is maintained. Frequent analysis of acid base status is required in Hb SS patients. Intraoperative crisis may present with changes in breathing pattern or blood pressure, or acidosis and hypoxaemia, which may be difficult to detect. Patients are not suitable for day case surgery.

Postoperatively patients should receive oxygen therapy for at least 24 hours. Analgesic drugs must be used with caution so as to avoid hypercapnia due to respiratory depression. Adequate hydration is maintained. Chest physiotherapy is important, as post op chest infection is a special risk in these patients. Early mobilization and other antithrombotic measures should be started as early as possible, because risk of thrombosis is much greater in these patients.



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### **TO WHOME IT MAY CONCERN**

When it's easy, make it difficult.  
When it's difficult, make it tough.  
When it's tough, make it hard.  
When it's hard, make it impossible.  
When it's impossible, then it's perfect.