

## Case Report

# Peri-Operative Management of a Patient with Sickle Cell Disease for Total Hip Replacement: Case Report

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**Received:** 18 August 2020; **Accepted:** 03 September 2020; **Published:** 02 October 2020

**Citation:** Bhuvanshyam J Bhaktavatsalam, Divya Iyer, Alexander Marfin. Peri-Operative Management of a Patient with Sickle Cell Disease for Total Hip Replacement: Case Report. Anesthesia and Critical Care 2 (2020): 079-083.

### Abstract

**Introduction:** Sickle cell disease is an inherited disorder, with multiorgan dysfunction resulting from vaso-occlusion and chronic haemolysis. Avascular necrosis of long bones is a known complication. Anaesthetists are frequently involved throughout perioperative period, which involves preoperative assessment, management of acute pain and prevention of postoperative complications.

**Case presentation:** We report a case of a 56 year old female patient with sickle cell disease who underwent total hip replacement for avascular necrosis of femoral head under spinal anaesthesia combined with general anaesthesia.

**Conclusion:** Meticulous perioperative care which includes careful planning, ensuring adequate oxygenation, hydration, tissue perfusion and pain control is vital in successful management of these patients.

**Keywords:** Sickle cell disease; Total hip replacement; Spinal anaesthesia; General anaesthesia

### 1. Introduction

Sickle cell disease (SCD) is an inherited haemoglobinopathy resulting from a mutation on chromosome 11. The mutation causes substitution of a single amino acid, valine for glutamate, at position 6 on the  $\beta$ -globin subunit of normal adult haemoglobin A, resulting in the formation of haemoglobin S (HbS) [1]. This makes HbS highly unstable and it precipitates out of solution when

in the deoxygenated state, forming the pointed, slightly curved 'sickle cells'. The greater the proportion of haemoglobin S in the cell, the greater is the propensity to sickle. The homozygous state results in aggressive form of SCD. The heterozygous carrier state or sickle cell trait results in the production of both haemoglobin A and S and is benign in nature. Other heterozygous states involving abnormal  $\beta$ -globin alleles such as  $\beta$ -thalassemia (HbS $\beta$ ), C (HbSC) and D-punjabi (HbSD) result in symptoms of SCD. It is most commonly seen in African, Mediterranean and south-west Asian population. The definitive test is haemoglobin electrophoresis, but in emergency cases, a rapid screening test, 'sickledex test' will detect HbS levels >10%; however, it is unable to distinguish SCD from sickle cell trait. Peripheral blood film may reveal sickle forms or an elevated reticulocyte count relative to the haemoglobin level.

The hallmarks of SCD are haemolysis and vasculopathy. This is characterized by intermittent, recurrent, acute episodes of severe pain, known as vaso-occlusive crises (VOC) or pain crises. Haemolytic anaemia is due to greatly reduced half-life of sickle cells when compared with normal red blood cells. Sickling is exacerbated by hypoxia, acidosis, cold, stasis, dehydration, infection, increased blood viscosity and stress [2]. Acute painful syndromes, severe anaemia, infections, acute chest syndrome, stroke, splenic sequestration and multiorgan failure causes increased morbidity and mortality. Surgeries such as cholecystectomy, splenectomy and orthopaedic procedures are often required. SCD and its specific complications increase the hazards of surgery as well as anaesthesia. About 7% of all deaths among patients with SCD are related to surgery [3].

## 2. Case Report

A 56 year old woman of Afro-Caribbean origin, diagnosed with avascular necrosis of right femoral head and was scheduled for total hip replacement. She was a known case of sickle cell disease (HbSC type) since childhood. She had a history of multiple episodes of vaso-occlusive crises requiring hospital admissions and has had multiple red cell exchange and blood transfusions in the past. In the past year, she developed right psoas abscess, which required admission to intensive care unit. Due to her disease process, she had poor physiological functional capacity. Her mobility progressively decreased due to worsening hip pain, and eventually she became bed bound. She was on increasing doses of morphine, tramadol, paracetamol and naproxen. She was on diazepam at night for anxiety.

On pre-operative assessment, her vital parameters were within normal limits and systemic examination was unremarkable. Airway examination revealed good mouth opening, Mallampati grade 1 and full range of neck movements. Her investigations were as follows: Hb 9.5 g/dL, HbS 45.4%, HbC 48.4%, HbA2 4%, HbF 1.1%; baseline coagulation profile, liver and renal function tests, electrocardiogram and chest X-ray were normal. On haematologist's advice, she had preoperative red cell exchange transfusion following which HbS dropped to 8.5%, HbC to 8.4% and Hb improved to 11.5 g/dL. She was started on incentive spirometry preoperatively. She was fasted from midnight and was started on intravenous fluids to avoid dehydration. Blood grouping and cross matching, with adequate blood availability was ensured prior to surgery.

Patient was extremely anxious and preferred to be asleep during surgery. Hence, the decision was made to give spinal anaesthesia first and then sedation/general anaesthesia. On arrival in the anaesthetic room, standard monitors were

applied with pulse oximetry, automated non-invasive blood pressure measurement and electrocardiography. Compound sodium lactate infusion through a fluid warmer was commenced via 18G cannula. Under all aseptic precautions, spinal anaesthesia was performed with the patient in the left lateral position using a midline approach at L3-4 interspace. 3 ml (15 mg) of 0.5% hyperbaric bupivacaine with 200 mcg of preservative free morphine was injected intrathecally. She was then turned supine and adequate level of sensory and motor blockade was achieved. Intravenous induction was done with fentanyl 100 mcg, propofol 150 mg and size 4 Igel was placed. Anaesthesia was maintained with sevoflurane. General anaesthesia with controlled ventilation was preferred over sedation in order to ensure good oxygenation and normocarbica. Right radial artery was cannulated and nasopharyngeal temperature probe was placed. Prophylactic antibiotics (Co-amoxiclav 1.2 g) and anti-emetics (dexamethasone 6.6 mg and ondansetron 4 mg) were given. Patient was catheterized to monitor urine output perioperatively and then secured in left lateral position for surgery. Protective pressure pads and forced air warmer were used. Intra-operatively, patient was hemodynamically stable and normothermic. Arterial blood gases (ABG) ruled out hypoxia, hypercarbia and acidosis. Surgery was uneventful with blood loss of 100 ml. She received total of 1.5 litres of IV fluids in total with an adequate urine output. At the end of the procedure, Igel was removed and she was transferred to high dependency unit (HDU) for postoperative care.

In HDU, patient was given oxygen via facemask till fully awake, and IV fluids were continued until oral intake was established. ABG was repeated and hourly urine output was monitored. Adequate pain control was achieved with multimodal analgesia which included morphine PCA (patient controlled analgesia) and continuation of regular paracetamol and naproxen. PCA was discontinued on

second day postoperatively and oral morphine was recommenced. Early mobilisation, thromboprophylaxis and chest physiotherapy/incentive spirometry was started. Repeat blood investigations on the following day showed Hb of 6.7 g/dL and 2 units of blood was transfused, with an aim to maintain Hb >8 g/dL. She was discharged on 4<sup>th</sup> postoperative day.

### **3. Discussion**

Orthopaedic disease affects a considerable proportion of SCD patients for which they often require surgical interventions. Common orthopaedic procedures reported include joint replacement, drainage of bone infections and correction of musculoskeletal deformities. The rate of sickle events after hip surgery is reported to be up to 19%. [4]. Management of such patients require multidisciplinary team approach with involvement of surgeons, anaesthetists, haematologists, acute pain team, physiotherapists and nursing staff.

The anaesthetist has specialized skills in pain management, ventilation, and critical care and hence, plays a pivotal role in the perioperative care of patients with SCD requiring surgery. Meticulous preoperative assessment should be carried out to ensure that the patient is maximally optimized for the surgery, to predict the risk of perioperative complications, and to plan for their optimal management. A detailed history and examination is required to assess disease severity (e.g. hospital admissions, number and timing of painful crises) and identify its precipitating factors. Signs of organ dysfunction should be sought and preoperative investigations should include chest X-ray, electrocardiogram, full blood count, renal and liver function tests, and coagulation profile. It is generally accepted to aim for a Hb of >10g/dL and HbS <30%. Routine preoperative red cell exchange transfusion to achieve this has been a standard practise for SCD patients undergoing major

surgery. However, data supporting this is mixed [5]. Hydroxyurea has been shown to decrease the frequency and severity of symptoms and of vaso-occlusive crises and has also been shown to reduce transfusion needs. It increases the formation of fetal haemoglobin (HbF), which confers some protection against sickling [6]. Avoidance of prolonged fasting combined with preoperative hydration may help prevent sickling. Thus, patients are often admitted at least 1 day prior to surgery. Chest physiotherapy has an important role in both pre and postoperative care.

The anaesthetic technique (general vs regional) and its impact on SCD complications remains controversial. Regional techniques are thought to be beneficial on the basis of improved analgesia, decreased blood loss, lower incidence of deep vein thrombosis, and the avoidance of airway manipulation and polypharmacy. However, it has been argued that this can be disadvantageous due to associated hypotension and the need for vasoconstrictors [7]. General anaesthesia with controlled ventilation may prove beneficial by avoiding hypoxia and hypercarbia, but has increased propensity to cause postoperative chest infections [8]. The choice of anaesthesia should be tailored according to the type of surgery and patient's general condition. During surgery, the goals of anaesthesia are to maintain oxygenation and hydration, avoid acidosis, and maintain normocarbida, normotension, and normothermia. Precautions should be taken to prevent venous stasis by careful positioning, avoidance of caval compression, and avoidance of prone position wherever possible. Occlusive orthopaedic tourniquets are not contraindicated, although this remains controversial [9]. The risks of a tourniquet are believed to be due to local hypoxia and acidosis leading to vaso-occlusion in the operative limb distal to the tourniquet; and that deflation of tourniquet can result in release of toxic metabolites and its associated systemic effects. Steps to reduce the risk include maintenance of hydration with

normal acid-base balance, the limb is exsanguinated well, and an experienced surgeon ensuring the shortest possible tourniquet time. Analgesia is an important component of any well-balanced anaesthetic technique. Multimodal analgesia comprising of paracetamol, non-steroidal anti-inflammatory drugs, opioids in conjunction with local and/or regional anaesthetic techniques plays a key role in achieving effective pain control.

SCD patients are at increased risk of postoperative complications such as painful vaso-occlusive crises, acute chest syndrome, stroke and infections. Predictors of postoperative complications in these patients are- type of surgery, increased age, frequency of complications and hospitalization, evidence of lung involvement, pregnancy, pre-existing infections and ethnicity [10]. Particular attention to oxygen supplementation, hydration, acid base regulation and analgesia is required and close monitoring in high dependency/intensive care setting is recommended, thus making day surgery inadvisable in these patients. Early ambulation, chest physiotherapy, thermoregulation and thromboprophylaxis also play a significant role in rehabilitation of these patients.

#### **4. Conclusion**

SCD has become a worldwide problem due to global shift of population and hence, remains an anaesthetic challenge all over the world. Our case highlighted the importance of multidisciplinary team approach and meticulous evaluation and planning at every stage to achieve a favourable outcome in the perioperative care of such patients. Prevention of sickling by avoiding the triggers remains the key in successful management of patients with SCD.

#### **Declaration of Patient Consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her

consent for her images and other clinical information to be reported in the journal. The patients understand that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial Support and Sponsorship

Nil.

### Conflicts of Interest

There are no conflicts of interest.

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