

# Anesthetic Management of a Patient with Sickle Cell Disease with Kochs Spine Posted for D5-D10 Instrumentation

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**Abstract:** Patients with sickle cell disease (SCD) may present to the anesthetist in different clinical settings like perioperative care, management of acute painful crisis and intensive therapy for acute respiratory failure. We describe the successful management of a 29-year-old male patient with SCD, case of Kochs spine posted for D5 to D10 decompression and instrumentation under general anesthesia. The importance of preoperative stabilization and careful anesthetic strategy is emphasized.

**Keywords:** Sickle cell disease, kochs spine, instrumentation, hbs, plasmapheresis

## 1. Introduction

Sickle cell disease (SCD) is a complex clinical entity characterized by an inherited chronic hemolytic anemia associated with variable number of acute painful vaso-occlusive episodes. Polymerization of hemoglobin-S (HbS) after deoxygenation is the fundamental molecular event that underlies the protean clinical manifestation of SCD.[1]. About 7% of all deaths among patients with sickle cell anemia are related to surgery.[2] Increased perioperative complications may result from vaso-occlusion after transient hypoxia, hypothermia, dehydration or acidosis. Inadequate post-operative pain of incision may reduce respiratory effort, leading to poor pulmonary toilet and relative hypoxia. We planned anesthesia and analgesia of the patient to avoid vaso-occlusive episodes and prevent complications.

## 2. Case Report

A 46-kg, 168-cm, 29-year-old man was posted for elective D5 to D10 decompression and instrumentation. He was diagnosed to have sickle cell anemia with homozygous trait 15 years back and hypertension 3 months back. On admission, she had complaints of fever on and off, generalized weakness, and jaundice. He was prescribed oral hydroxyurea, levetiracetam, allupurinol, vitamin and anti tubercular therapy. Antibiotic prophylaxis with intravenous (IV) ciprofloxacin and metronidazole was administered. He had recent history of one episode of convulsion and his computerized tomography scan showed transverse venous thrombosis. However, his magnetic resonance image (MRI) report was negative. He was not evaluated further for convulsion. He did not give any recent or past history suggestive of respiratory infection or complications, chest pain suggestive of cardiac involvement and renal problems suggesting impaired renal function. He was a known case of kochs spine with D5 to D10 involvement. On general examination, she was icteric and had stable vital parameters with regular pulse rate 90/min and blood pressure (BP) 110/86 mmHg. On per abdomen palpation, there was no visceromegaly. His investigations were as follows: Hb, 7.2 gm%; HbA2, 2.1%; HbF, 16.1%; HbS, 78.3%; baseline coagulation parameters, liver function tests, renal function

tests, blood sugar, electrocardiogram, X-ray chest, 2D echocardiogram and arterial blood gas (ABG) were within normal limits. Since his HbS was 78.3%, she underwent two partial exchange transfusions after which his HbS dropped to 36.6%, Hb was 11.3 gm%, packed cell volume (PCV) 32%, total count 8300/cu.mm and platelets 1,02,000/cu.mm.

The patient was started on incentive spirometry preoperatively. He was kept fasting after 12 midnight and was started on IV ringer lactate 100 ml/hour to avoid dehydration. His urine output was monitored and pulse oximeter showed 100% oxygen saturation. Right internal jugular vein was secured under local anesthesia after sedation with IV fentanyl 1  $\mu$ g/kg and midazolam 1 mg. The patient received oxygen by facemask. Right radial arterial line was secured and invasive blood pressure monitoring was used in view of long duration of surgery and massive fluid shift.

The patient received IV glycopyrrolate 0.2 mg, midazolam 1 mg, fentanyl 1  $\mu$ g/kg, ranitidine 50 mg and ondansetron 4 mg before induction. Anesthesia was induced with IV propofol and atracurium, and trachea intubated with 7.5 cuffed polyvinyl endotracheal tube. Utmost care was taken during induction to avoid hypotension and hypoxia. Baseline ABG done, which was normal. A nasopharyngeal temperature probe was passed for temperature monitoring and temperature was maintained at 36 $^{\circ}$ C  $\hat{a}$ c $^{\circ}$  37 $^{\circ}$ C. Anesthesia was maintained with 50:50 O<sub>2</sub>:N<sub>2</sub>O, isoflurane, IV atracurium and Fentanyl infusion. Sevoflurane was avoided as significant percent of these patients have impaired kidney function. The patient was mechanically ventilated using closed circuit with end-tidal carbon dioxide (ETCO<sub>2</sub>) monitoring. IV fluids were infused to maintain a central venous pressure (CVP) of 8-9 cms of H<sub>2</sub>O and urine output of 2 ml/kg/hour. Intraoperative blood loss was 400 ml which was replaced with 500 ml hydroxyethyl starch. Throughout surgery, the patient remained hemodynamically stable with pulse rate 64-78/min and systolic BP of 104-126 mmHg. Intraoperative repeated ABG was done to rule out hypoxia and acidosis. The patient was extubated on table and shifted to ICU for postoperative monitoring. He received oxygen supplementation at 6l/ min and fentanyl

infusion for 3 days post operatively. Regular ABG analysis was done to rule out hypoxia and acidosis. The patient was started on low molecular weight heparin as thromboprophylaxis and incentive spirometry on day 2 of postoperative period. Throughout his stay, hemodynamics remained stable and urine output was maintained between 50 and 100 ml/hour. On day 5 the patient was shifted to the ward.

### 3. Discussion

Vaso-occlusive phenomena and hemolysis are the clinical hallmarks of SCD, an inherited disorder due to homozygosity for the abnormal hemoglobin, that is, HbS. Vaso-occlusion results in recurrent painful episodes and a variety of serious organ system complications that can lead to disabilities and early death. Hydroxyurea and other agents have been used to increase the production of HbF, inhibiting HbS polymerization.[3] Induced hyponatremia has been used to reduce the HbS concentration, but was found to be impractical.[4] Alkalinization using magnesium glutamate, to increase oxygen affinity of Hb in the RBC, has been tried. Oral magnesium supplement reduces erythrocyte dehydration, reducing the cellular concentration of HbS in SCD patients.[5]

Acute chest syndrome is one of the most serious complications of SCD, with a mortality rate of 10%, but its pathogenesis is not clearly understood.[6] Progressive fibrosis has been detected in children with multiple episodes of acute chest crisis.[7] It is difficult to differentiate respiratory symptoms due to bacterial infection from acute chest syndrome. So, it is always advisable to start patients on broad-spectrum antibiotics in the perioperative period. Functional hyposplenism makes these patients susceptible to streptococcal infection and amoxicillin has good activity against streptococcal pneumonia. To prevent pulmonary complications, prophylactic continuous positive airway pressure and incentive spirometry should be started.[8]

Proper planning and optimal perioperative preparation is a key to successful management of SCD patients. Adequate hydration to decrease the viscosity of blood, control of infections and getting the hemoglobin levels normal and PCV between 30% and 35% is essential. Many of these patients have impaired kidney function due to renal medulla infarction which may interfere with their ability to maintain fluid and electrolyte balance during periods of stress. Preoperative need for exchange transfusion depends on the general condition of the patient and the type of surgical procedure. Exchange transfusion is generally recommended before major surgical interventions in order to minimize sickling and reduce the circulating HbS concentration below 30%.[9] In our patient, partial exchange transfusions done before surgery reduced the HbS level from 78.3 to 36.6% which drastically improved the perioperative outcome.

### 4. Conclusion

In conclusion, meticulous anesthetic management in the form of avoiding acidosis, hypoxia, hypothermia, hypovolemia, maintaining normocarbia, good intraoperative and postoperative pain relief with epidural infusion,

postoperative thromboprophylaxis, postoperative oxygen therapy with inspired concentration up to 40%, chest physiotherapy, nebulization, incentive spirometry with early mobilization and regular ABG monitoring played an important role in improving the patient outcome. Postoperative monitoring and pain relief play a vital role in avoiding pulmonary complications.

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