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# **Case Reports**

# Case of anesthetic management of a patient with sickle cell disease for pericardiectomy: anterior pericardiotomy and avoiding cardiopulmonary bypass

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#### ARTICLEINFO

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#### Abstract

Abstract: Patients having sickle cell disease can land up to the anesthetist for various purposes like surgery, acute pain management and intensive care for acute respiratory failure. Our case was of a 20-year-old female patient presented with anemia and symptoms of constrictive pericarditis. Following investigations, which were also suggestive of sickle cell disease, she was planned for pericardiectomy under general anesthesia. We performed anterior pericardiectomy rather than a complete pericardiotomy to avoid cardiopulmonary bypass and to decrease the chance of postoperative complications.

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#### Introduction

**S**ickle cell disease (SCD) is a beta globin chain hemoglobinopathy characterized by chronic hemolytic anemia along with intermitent acute painful vaso-occlusive episodes, respiratory compromise and multi organ failure. Perioperative mortality is almost 7% in patients with sickle cell anemia in elective surgeries. The perioperative complications is mainly due to vaso-occlusion following episodes of hypoxia, hypothermia, dehydration or acidosis. Anterior pericardiectomy rather than a complete pericardiaectomy and avoidance of cardiopulmonary bypass was planned to prevent deleterious effects of cardiopulmonary bypass (CPB) and its possible resulting complications.

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Case report: Our case was a 20-year-old woman with a height of 152 cm and weight of 45 kg scheduled for elective pericardiectomy for chronic constrictive pericarditis. She had sickle cell anemia with homozygous trait diagnosed since 7 years. Her major complaints were shortness of breath on exhertion, pedal edema, facial puffiness and jaundice since last 1 year. She was under oral anti tuberculosis therapy for a course of 6 months. She was currently under hydroxyurea, folic acid and lasilactone once daily. She did not give any recent or past history suggestive of vaso- occlusive crisis. On general examination, she was icteric and had pedal edema and facial puffiness. Her vitals were stable with pulse rate of 72/min and regular, blood pressure of 90/60 mmHg, respiratory rate of 16/min and room air oxygen saturation 98%. On abdominal examination fluid thrill and shifting dullness was positive suggestive of ascities. Her lab investigations showed a hemoglobin of 8.0 gm%; platelets count 67000/ cu mm, total bilirubin 106 mmol/l and direct bilirubin 38 mmol/l. Her other coagulation tests, renal function tests, blood sugar, electrocardiogram, X-ray chest, were within normal limits. 2D echocardiogram showed dilated inferior venacava with inspiratory collapsibility <50%, thickened pericardium, moderate mitral regurgitation, moderate tricuspid regurgitation and other features suggestive of effusive constrictive pericarditis. Pre operatively she underwent three units of blood transfusion and her post transfusion hemoglobin was 11.7% and platelets count was 90000/cu mm.

The patient was kept nil per oral after 12 midnight and intravenous ringer lactate was started at the rate of 100 ml/hour to avoid dehydration. Patient was taken to the operation room and ECG and pulse oxymeter was attached. Patient was given oxygen supplementation by face mask. Forced air warmer was used in order to prevent hypothermia. Invasive blood pressure was measured by cannulating right radial artery with a 20 gauge arterial cannula. Central venous cannulation was done at the right internal jugular vein under local anesthesia and dopamine was started at 5 micro grams per kg/min.

The patient was given intravenous (IV) antibiotics Cefazolin 1 gm and flucloxacillin 1 gm. She also received IV midazolam 1 mg, fentanyl 2  $\mu$ g/kg, ranitidine 50 mg and chlorpheneramine 12.5 mg before induction. Anesthesia was induced with IV ketamine 50 mg and atracurium 30 mg and oro tracheal intubation was done with 7.0 mm ID cuffed polyvinyl endotracheal tube. Patient was kept in mechanical ventilation with standard ventillatory settings. FIO2 was kept 60% throughout the surgery. Dopamine dose was titrated in order to maintain mean arterial pressure of more than 70 throughout the surgery. A nasopharyngeal temperature probe was kept and temperature was maintained at 36°C-37°C. Anesthesia was maintained with isoflurane and IV atracurium as needed. Warm intravenous ringer lactate was infused as a maintenance fluid. Anterior pericardiectomy was

performed. Intraoperative urine output was 230 ml and blood loss was about 400 ml. Intraoperatively, the patient was hemodynamically stable with pulse rate 72-84/min and MAP of 70-84 mmHg. Repeated ABG was done intraoperatively to rule out hypoxia and acidosis. The patient was shifted to the intensive care unit and was extubated after 2 hours as per fast-track extubation protocol. Oxygen supplementation was given with a face mask at 5 l/ min and regular arterial blood gas analysis was done to rule out hypoxia and acidosis. Adequate warming of the IV fluids and patients bed was done in order to prevent hypothermia. Analgesia was maintained with intravenous paracetamol 1 gm 6 hourly and tramadol 50 mg 8 hourly. Incentive spirometry was started on day 2 of surgery. Her hemodynamics remained stable throughout her ICU stay and dopamine infusion was gradually tapered off on day 2 of surgery. Mediastinal drain, central venous catheter and arterial line were removed and the patient was shifted to the ward on the 4<sup>th</sup> day of surgery.

**Discussion:** Sickle cell disease is a hemoglobinopathy due to abnormal hemoglobin, HbS leading to vaso-occlusive phenomena and hemolysis which results in recurrent painful episodes and other serious systemic complications leading to disabilities and early death. Various pharmacologic measures like Hydroxyurea and other agents are used for increasing the production of HbF and inhibiting the HbS polymerization. Our patient was under hydroxyurea and we continued the drug in the perioperative period.

Surgery can itself precipitate the sickling phenomenon mainly due to hypoxia, hypothermia, hypovolemia and pain. So proper planning has to be done in order to prevent the likely complications. The viscosity of blood has to be decreased and hematocrit has to be between 30-35%, so adequate perioperative hydration has to be done. Infection control and exchange transfusion has to be done prior to major surgeries to minimize the HBS level to less than 30%. In our case adequate hydration and blood transfusion was done to maintain the optimal hemoglobin and hematocrit percentage. The perioperative complications is mainly due to vaso-occlusion following episodes of hypoxia, pain, hypothermia, dehydration or acidosis<sup>ii</sup>. We could maintain normothermia, normocarbia, normal PH and optimal oxygenation throughout the perioperative period. The use of cardiopulmonary bypass can provoke sickling itself due to hypoxia, acidosis or hypothermia. In our case, anterior pericardiectomy rather than a complete pericardiaectomy and avoidance of cardiopulmonary bypass was performed to prevent deleterious effects of CPB and its resulting complications.

In conclusion, perioperative management by avoiding acidosis, hypoxia, hypothermia, hypovolemia, and optimal pain management prevents the unwanted complications and improves the patient outcome. Besides cardiopulmonary bypass itself can provoke sickling phenomenon, so whenever possible avoidance of CPB

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in cardiac surgical patients can be beneficial. Anterior pericardiotomy without cardiopulmonary bypass if applicable may be a viable option.

**Consent for publication:** A consent for scientific publication of the disease condition without disclosing the identity was obtained from the patient.

**Conflict of interest**: The author has filled the ICMJE conflict of interest form and has nothing to disclose.

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