Case Report

Perioperative considerations in a sickle cell patient undergoing cardiopulmonary bypass

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ABSTRACT

An 11-year-old child, a known case of sickle cell anaemia with a history suggestive of sickling crisis in the past was scheduled for surgical pulmonary valvotomy. Pre-operative blood transfusion and hydroxyurea were administered. Pre-operative blood transfusion is indicated in sickle cell disease patients to raise the haematocrit level and lower sickle haemoglobin (HbS) levels. Before the start of cardiopulmonary bypass (CPB), exchange transfusion was performed to reduce HbS level and raise adult haemoglobin level. Hypothermia was prevented by employing normothermic CPB.

Key words: Autotransfusion, cardiopulmonary bypass, haemofiltration, hydroxyurea, sickle cell disease

INTRODUCTION

Sickle cell disease (SCD) is a form of haemoglobinopathy caused by the abnormal genetic substitution of glutamine by valine in the haem portion of the haemoglobin molecule. Sickle haemoglobin (HbS) tends to form polymers and aggregates when deoxygenated and can lead to vasoocclusion and organ ischaemia (1). Cardiopulmonary bypass (CPB) is commonly associated with hypothermia, hypoxia, hypoperfusion, and acidosis, which are pre-disposing factors that can trigger a profound sickle cell crisis. When such patients undergo cardiac surgery with CPB, they need special precautions and management to prevent fatal vaso-occlusive episodes. There are no guidelines for management of sickle cell patients during cardiac surgery, but only case reports are mentioned in the literature.

CASE REPORT

An 11-year-old child (weight - 23 kg, height - 122 cm, body surface area - 0.89 m^2) presented with a history of shortness of breath while playing, jaundice, and joint pain. She was scheduled for balloon pulmonary valvotomy. She was a known case of sickle cell anaemia with a history suggestive of sickling crisis in the past, which was managed by blood transfusion. Patient was started on hydroxyurea 250 mg/day (oral). On admission, patient's Hb was 8 g/dL. Haemoglobin assessment by electrophoresis revealed HbS of ≅70.80% and adult haemoglobin (HbA) of ≅30.60%. Echocardiographic evaluation showed severe pulmonary stenosis. She was transfused whole blood on 2 consecutive days before proceeding for valvotomy. Pre-procedure Hb improved to 10.2 g/dL. Initial percutaneous balloon pulmonary valvotomy was attempted but it was not successful. Surgical repair was planned. On the day before the surgery, one unit whole blood was transfused and before induction, Hb electrophoresis values were obtained, which showed HbS ≅30.3%, foetal haemoglobin (HbF) ≅12.1%, and HbA ≅49.9%.

Cardiopulmonary bypass circuit was primed with 1300 mL of Ringers solution, 3 units of fresh donor red cells (collected within 24 h), 1 unit fresh frozen plasma, 60 mL albumin 20%, 100 mL mannitol and 50 mL 7.5% sodium bicarbonate. After the circuit was primed, the fluid was circulated through oxygenator to achieve adequate oxygenation and warming of blood. Analysis of prime fluid showed PaO_2 of 310 mm Hg, haematocrit of 24% and potassium, 3.0 mmol/dL.

After systemic heparinisation with 3 mg/kg of heparin, aortic and bicaval cannulation was performed. Before

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the start of CPB, exchange transfusion was planned to reduce HbS level and raise HbA level [Figure 1]. As 500 mL autologous blood was drained, systolic arterial pressure dropped to 50 mm Hg. Venous drainage was stopped temporarily, and prime fluid was infused until haemodynamics got stabilised. The drainage was slowly restarted, and volume of 1400 mL of autologous blood was collected with simultaneous infusion of priming fluid. The collected blood was discarded. After exchange transfusion, Hb electrophoresis showed HbS ≅ 6.5%, HbF ≅ 3.2%, and HbA ≅ 78.2%. CPB was initiated with flows at 2.3-2.5 L/min at normothermia [Table 1]. Cardiotomy suction blood was discarded. Continuous haemofilteration was performed during bypass. Venous reservoir level was maintained by adding fresh blood to keep haematocrit level near 25%. Patient was weaned from bypass by inotropic support of adrenaline 0.02 μg/kg/min. As the patient was off the bypass, remaining blood in the reservoir was discarded. Modified ultrafiltration was done after the discontinuation of CPB. Post-CPB, patient was maintained normothermic using warm fluid, warm air mattress, and at room temperature. After heparin reversal, Hb was 8.4 g/dL and one unit fresh blood was transfused. The child was shifted to intensive care unit with stable haemodynamics. Post-operative Hb was 10.2 g/dL and HbS 10% [Table 2]. Post-operative analgesia and sedation was provided by dexmedetomidine infusion and intravenous paracetamol. There was no episode of veno-occlusive complications post-operatively. Patients recovery was uneventful and extubation was performed within 4 h of surgery.

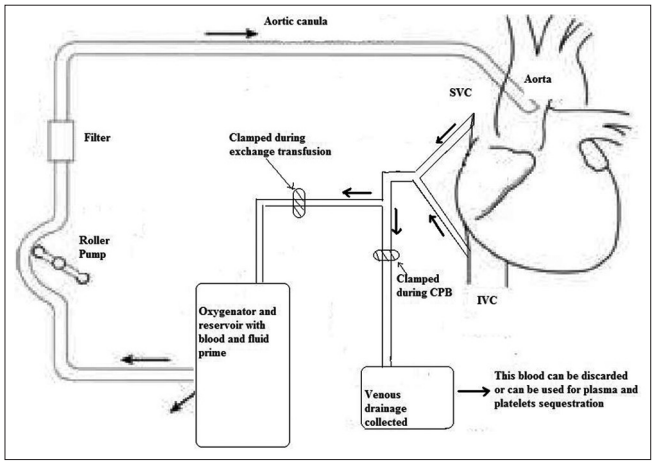

Figure 1: Cardiopulmonary bypass circuit showing arrangements for exchange transfusion

Table 1: CPB data

CPB data
Flow rate - 2.3-2.5 L/min
Mean blood pressure - 60-70 mmHg
Temperature - 36-36.5°C
Activated clotting time >500 s
CPB time - 36 min
Aortic cross clamp time - 26 min

CPB – Cardiopulmonary bypass

DISCUSSION

Sickle cell disease is an autosomal recessive condition that results from a single nucleotide polymorphism in the β-globin gene. Under the right conditions haemoglobin SS molecules with this point mutation can polymerise, stiffening the erythrocyte membrane and distorting the cellular structure to the characteristic sickle shape. This change of shape alters cellular transit through the microvasculature. Certain circumstances such as hypoxia, hypothermia, acidosis or diminished blood flow can lead to aggregation, vascular occlusion and thrombosis. Chronically, SCD can give rise to multiorgan damage secondary to haemolysis and vascular obstruction.[1]

Reduced life expectancy and a tendency for complications in sickle cell trait or disease can negatively affect the likelihood of survival after open heart surgery. Yousafzai *et al*. have reported heart valve surgery and surgery for congenital heart diseases performed safely in patients with SCD or sickle cell trait with acceptable outcome and survival rates.[2]

Bocchieri *et al*. described a method of complete intraoperative exchange transfusion in the operating room that reduces the haemoglobin S (HbS)

Table 2: Laboratory parameters in the peri-operative period

Parameters	Baseline preoperative	Pump prime before exchange transfusion	On CPB	After heparin reversal	Post-extubation
pH	7.42	7.65	7.51	7.45	7.38
pCO_2 (mm Hg)	39	21	31	37	45
pO_2 (mm Hg)	95	310	357	363	247
HCO_3^-	25.6	30	24.7	25.7	25.4
O2 Saturation (%)	99	100	100	100	100
Haemoglobin	11.8	7.4	7.3	8.4	10.2

CPB – Cardiopulmonary bypass

level to <5%, without pre-operative exchange transfusions. Plasma and platelet fractions separated intra-operatively from the patient's native red cell mass were used, in addition to hemoconcentration while on CPB, to effectively reduce the red cell and clotting factor transfusion requirements after the procedure.[3] Usman *et al*. suggested minimally invasive, warm, beating heart approach to try and minimise the risk of sickling due to CPB, low-flow states, cold cardioplegia and aortic cross-clamping.[4] Maddali *et al*. have reported case of elective coronary artery bypass graft surgery in a SCD patient using CPB techniques following pre-operative transfusions to increase the haemoglobin A levels to above 60%.[5] In this case, we used pre-operative blood transfusion and hydroxyurea. Pre-operative blood transfusion is indicated in SCD patients to raise the haematocrit level and lower HbS levels.[6,7] Hydroxyurea is the main treatment in symptomatic sickle cell patients because of its beneficial effects.[8] Hydroxyurea increases HbF, which prevents sickling and also has beneficial effects on red blood cells hydration and vascular wall adhesion.There is no consensus on safe values of HbS in patients undergoing surgery, but some reports suggest HbS should be decreased to <30% for major surgeries[9] or <5% for cardiac surgeries prior or at the time of operation.[10] In our case, even though HbS was 30.3% before surgery, we decided to do pre-operative exchange transfusion to promote perfusion, lower the incidence of peri-operative sickling crisis and perioperative multiorgan failure. Exchange transfusion is known to decrease HbS, increases pre-operative HbA, haematocrit, and cellular oxygen delivery.[2] The classic pre-disposing factors for sickling include exposure to cold stress, dehydration, infections, hypoxia, inflammatory cascades, and acidosis.[2,11] These conditions are common in cardiac surgery.

Hypothermia is prevented by employing normothermic CPB, use of warm air blankets and maintaining operating theatre temperature. We avoided hypoxia by hyperoxygenating pump prime, maintaining haematocrit above 24% by using fresh donor blood (collected within 24 h) and keeping venous oxygenation saturation above 80%. Frequent monitoring of arterial blood gas is needed to maintain pH in normal range. In our case, warm induction of cardioplegia with oxygenated crystalloid solution was used. Advantages of this technique[6] are delivery of high PaO_2 to myocardium, improved distribution of cardioplegic solution due to decreased viscosity and flushing of HbS from the myocardium, thereby reducing the risk of localised anoxia.

Use of continuous haemofiltration and modified ultrafiltration reduces the inflammatory response and improves lung compliance.[6] We did not use cell saver and autotransfusion was not done during or after the surgery. Cardiac surgery constitutes major stress, especially in paediatric patients, so it is strongly recommended to give the patients adequate analgesia and sedation.[2,11] Simultaneously we need to remember the respiratory depressant effects of opioids. Hence, we used dexmedetomidine infusion and intravenous paracetamol for analgesia.

CONCLUSION

Thorough pre operative stabilisation of the patient with respect to Hb and use of initial exchange transfusion, warm cardioplegia and maintenance of blood gases enable CPB to be conducted safely in sickle cell patients.

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