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Mitral Valve Replacement in a Patient with Sickle Cell Disease Using Perioperative Exchange Transfusion

David Chabot, MS, CCP, LP; Robin Sutton, MS, CCP, LP

Department of Perfusion Technology, Rush University Medical Center, Chicago, Illinois

Abstract: Sickle cell disease is a genetic hemoglobinopathy in which a significant number of red blood cells carry hemoglobin-S as opposed to normal red blood cells that contain hemoglobin-A. Under certain conditions such as hypoxia, acidosis, and hypothermia, the red blood cells containing hemoglobin-S will sickle, leading to occlusion of the microvasculature. As such, patients with sickle cell disease present unique challenges during heart surgery using cardiopulmonary bypass (CPB). After conducting a literature review, we discovered that the exact hemoglobin-S level for conducting cardiac surgery with CPB is not known. However, a hemoglobin-S level <30% is considered safe for conducting CPB. The following case report will discuss these challenges and present a patient with sickle cell disease undergoing a mitral valve repair. Management of this patient involved exchange transfusions both preoperatively and intraoperatively.
Keywords: sickle cell disease, cardiopulmonary bypass, exchange transfusion, hemoglobin-S, mitral valve replacement. *JECT. 2008;40:275–277*

Sickle cell disease is one of the most prevalent genetic disorders in the United States, affecting ~50,000 Americans per year (1). Sickle cell disease is a form of sickle cell hemoglobinopathy that occurs in a homozygous recessive form versus patients with the sickle cell trait, which is in the heterozygous form (2). Patients with sickle cell disease carry predominantly hemoglobin-S (HbS) in their blood instead of hemoglobin-A (HbA), which is found in normal healthy adults (3). These HbS cells are structurally fragile and tend to aggregate and assume a sickle shape. Sickling of red blood cells can lead to a vaso-occlusive crisis, leading to vascular occlusion and organ ischemia. Conditions such as oxygen saturation <85%, acidosis, hypothermia, and capillary stagnation can lead to this crisis state (2,3). Because these conditions can be present in a patient using cardiopulmonary bypass (CPB), patients with sickle cell disease who require the use of CPB present a unique challenge. In this paper, we present a patient with sickle cell disease requiring mitral valve replacement and the use of an intraoperative exchange transfusion immediately before CPB.

CASE REPORT

A 41-year-old woman with known sickle cell disease presented to Rush University Medical Center with chest pain, palpitations, shortness of breath, and mitral valve prolapse. She was found to have mitral valve regurgitation and an ejection fraction of 45–50%. She was scheduled for elective mitral valve replacement. The patient was 165 cm tall and weighed 59 kg, resulting in a body surface area (BSA) of 1.64 m^2 and was found to have an HbS level of 87%. The decision was made to perform an exchange transfusion using packed red blood cells (PRBCs) and fresh frozen plasma (FFP) both in the pre-operative period and immediately before initiation of CPB.

On the day before surgery, the patient underwent an exchange transfusion of PRBCs and FFP. After this procedure, her HbS level dropped from 87% to 29%. However, this information was not available to the surgical team on the day of the operation, so the decision was made to perform a second exchange transfusion intraoperatively.

Our standard CPB circuit was used. This consisted of a hollow fiber oxygenator (SX-18; Terumo, Ann Arbor, MI). A heparin-coated circuit (Medtronic, Minneapolis, MN) was used with a centrifugal type arterial pump (Delphin; Terumo), an arterial line filter (Affinity; Medtronic), and 4:1 blood cardioplegia system (Sorin Group BCD, Arvada, CO). A hemoconcentrator (HPH 400; Minntech, Minneapolis, MN) was also placed in the circuit. An inline blood gas monitoring device (CDI 500; 3M Health Care, St. Paul, MN) was used for continuous monitoring of arterial and venous blood gases, venous oxygen saturation, and hemoglobin and hematocrit.

The circuit was adapted for the exchange transfusion by

Address corresponence to: David Chabot, MS, CCP, LP, Rush University Medical Center, 1653 W. Congress Parkway, Suite 625, Chicago, IL 60612. E-mail: David_L_Chabot@rush.edu
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adding a ½–½–⅜″ Y connector within the venous line. The two ½″ ends of the Y were connected to the standard venous line of the circuit. The ⅜″ end of the Y was connected to ⅜″ tubing and attached to a separate cardiotomy reservoir (Figure 1).

Our circuit was initially primed with 1300 mL of Plasmalyte-A (Baxter, Deerfield, IL), 10 meq of sodium bicarbonate, 5000 units heparin, and 135 mL of 20% mannitol. After priming and debubbling the circuit in our standard fashion, excess crystalloid fluid was drained from the circuit, and 750 mL of PRBCs and 750 mL of FFP were added to the venous reservoir. An additional 3000 units of heparin, 20 meq of sodium bicarbonate, and 1 g of calcium chloride were added to normalize the prime.

Once the chest was opened and the heart was exposed, the patient was systemically heparinized with 22,000 units of heparin. A 7-mm aortic cannula (Sarns soft flow; Terumo) was placed in the ascending aorta. Venous drainage was accomplished through bi-caval cannulation with 28- and 30-Fr single stage venous cannulas (Edwards, Irving, CA) placed in the superior vena cava and inferior vena cava, respectively.

Once an activated clotting time of >480 seconds was achieved, the exchange transfusion was initiated. With the venous line clamped distal to the Y connector, the line to the separate cardiotomy reservoir was opened, allowing the patient's blood to drain. Simultaneously, blood from the heart lung machine was transfused to the patient to maintain the patient hemodynamically stable. After an exchange transfusion of 1500 mL was achieved, the patient was placed on CPB. The sequestered blood was processed with a Cell Saver 5 type red cell processing device (Hemonetics, Braintree, MA). Platelet-rich and platelet-poor plasma was separated. Eight hundred milliliters of platelet-poor plasma was returned to the patient while on CPB. The platelet-rich plasma was transfused to the patient after CPB termination. Sequestered red blood cells were discarded.

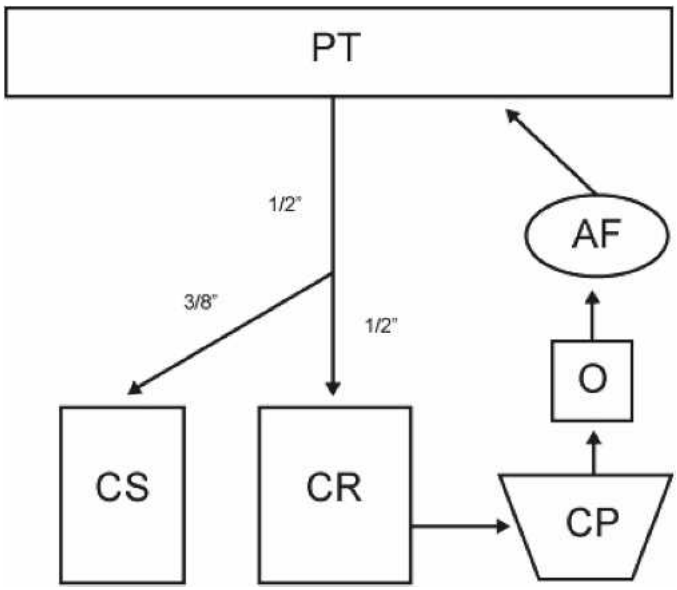

Figure 1. Diagram of CPB circuit modifications for exchange transfusion. AF, arterial line filter; CR, cardiotomy reservoir heart lung machine; CS, Cell Saver reservoir; CP, centrifugal pump; O, oxygenator; PT, patient.

Mild hypothermia was used, and the patient was cooled to a bladder temperature of 34°C. Antegrade and retrograde cardioplegia cannulas were placed. An aortic cross-clamp was placed, and the heart was initially arrested with 2770 mL antegrade warm blood-crystalloid cardioplegia in a 4:1 ratio. Twenty-two hundred milliliters of warm retrograde cardioplegia was given, at which time the decision was made to switch to a cold solution. Subsequent doses of cold retrograde cardioplegia were given at 10- to 20-minute intervals. A 29-mm mechanical heart valve (St. Jude, St. Paul, MN) was implanted. CPB flows were maintained at a minimum cardiac index of 2.3 L/min. Venous saturation was kept >70%, and no acidosis occurred during the CPB period. One unit of PRBCs was given during the bypass run. The patient was re-warmed to a bladder temperature of 36°C. The aortic cross-clamp was removed after 110 minutes. After resumption of normal sinus rhythm, the patient was ventilated and successfully weaned from CPB. Total CPB time was 191 minutes.

After surgery, the patient was transferred to the surgical intensive care unit and was extubated the same day. The patient was discharged to home on postoperative day 5.

DISCUSSION

This case showed that patients with sickle cell disease can successfully undergo cardiac surgery with the use of CPB. Because this patient underwent a pre-operative exchange transfusion that reduced her HbS levels from 87% to 29%, the second exchange transfusion in the operating room could have been avoided if the surgical team was made aware of that information, because our goal was to achieve an HbS level of <30%. However, without this information, we felt that the second exchange transfusion would have increased our margin of safety during CPB.

The safe level of HbS for CPB has not been established; however, there is literature stating that HbS levels of <30% are considered safe for elective cardiac surgery (4,5). Other measures the perfusionist can undertake is aggressive treatment of acidosis, maintaining high flows, and reducing areas of stagnant blood within the CPB circuit. During this case, high flows were maintained with a cardiac index of at least 2.3 L/min. These high flows helped us maintain a venous saturation of >70% without acidosis. Had any acidosis occurred, this would have been aggressively treated with sodium bicarbonate. Although the literature states that saturations of ~85% should be

maintained to prevent sickle cell crisis, the authors felt that, after transfusing 1 unit of PRBCs and achieving an hematocrit (HCT) of 25% with high flows, additional transfusions of PRBCs were not necessary (2). The decision to arrest the heart with warm blood cardioplegia was made because it was believed that flushing the myocardium with the warm solution would wash out the remaining HgS, thus reducing the risk of myocardial ischemia and occlusive crisis. The initial warm solution would also improve the distribution of the cardioplegia solution by having a lower viscosity. This technique was previously described in the literature (6). The use of the Cell Saver to separate the RBCs and plasma and further produce both platelet-rich and platelet-poor plasma was beneficial in having plasma with the patients clotting factors available, as well as a supply of platelet-rich plasma after termination of CPB to help achieve hemostasis. This case shows that, with advance planning on the part of the surgical team, patients with sickle cell disease can safely and successfully undergo elective cardiac surgical procedures.

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