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Case Reports

Sickle Cell Hemoglobin C Disease Patient Undergoing Coronary Artery Bypass Grafting with Complete Exchange Blood Transfusion during Cardiopulmonary Bypass

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Abstract: Sickle cell disorders are associated with increased risk of sickling and vaso-occlusive complications when undergoing cardiopulmonary bypass (CPB) surgery. Monitoring of certain parameters such as venous and arterial oxygen content, hematocrit, acid base homeostasis, and body temperature are required for a superior outcome. Furthermore, perioperative exchange transfusion has a positive effect on the outcome of surgery and on the survival of patients undergoing heart surgery. Avoiding intraoperative hypoxia and hypothermia, and minimizing hemoglobin S (HbS) and hemoglobin C (HbC) levels with exchange transfusion make bypass surgery relatively safe with enhanced outcomes in these cases. The exact HbS level for conducting cardiac surgery with CPB is not known, however, a HbS level <30% is considered safe for conducting CPB. By using a "discard" cardiotomy reservoir and priming the oxygenator reservoir with donor blood, we were able to reduce the intraoperative circulating HbS and HbC levels to less than 15% and sequester the plasma and clotting factors from the discarded blood using intraoperative plasmapheresis.
Keywords: sickle cell anemia, cardiopulmonary bypass, exchange transfusion, Hemoglobin SC disease. *J Extra Corpor Technol. 2018;50:117–9*

OVERVIEW

Individuals with sickle cell disease (SCD) have abnormal hemoglobin, called hemoglobin S (HbS) *or* sickle hemoglobin, in their red blood cells (RBCs). People who have SCD inherit two abnormal hemoglobin genes, one from each parent. In all forms of SCD, at least one of the two abnormal genes causes a person's body to make HbS. SCD is an autosomal recessive condition represented by two copies of the gene creating a homozygous condition. Sickle cell trait (SCT) presents with one copy of the gene in a heterozygous condition.

RBCs that contain hemoglobin A (HbA) are disc shaped (like a doughnut without a hole). This shape allows the cells to be flexible so that they can move through large and small blood vessels to deliver oxygen (1–5).

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HbS can form stiff rods within the red cell, changing it into a crescent or *sickle* shape. Sickle-shaped cells are not flexible and can stick to vessel walls, causing a blockage that slows or stops the flow of blood (Figure 1). When this happens, oxygen can't reach nearby tissues creating a vaso-occlusive crisis. Sickle cells can't change shape easily, so they tend to burst apart or *hemolyze*. Normal RBCs live about 90–120 days, but sickle cells last only 10–20 days.

The red cell sickling and poor oxygen delivery can also cause organ damage. The lack of tissue oxygen can cause attacks of sudden, severe pain, called *pain crises*. Over a lifetime, SCD can harm a person's spleen, brain, eyes, lungs, liver, heart, kidneys, penis, joints, bones, or skin.

Most adults contain 96–98% HbA. Patients with SCD have almost no HbA and 70–98% HbS. SCT patients usually present with less than 50% HbS.

When such patients undergo cardiac surgery with cardiopulmonary bypass (CPB), they need special precautions and management to prevent fatal vaso-occlusive episodes. Patients with SCD who require cardiac surgery are at risk of a potentially fatal sickling crisis, which may be induced by hypothermia, hypoxia, acidosis, or low-flow states. Preoperative

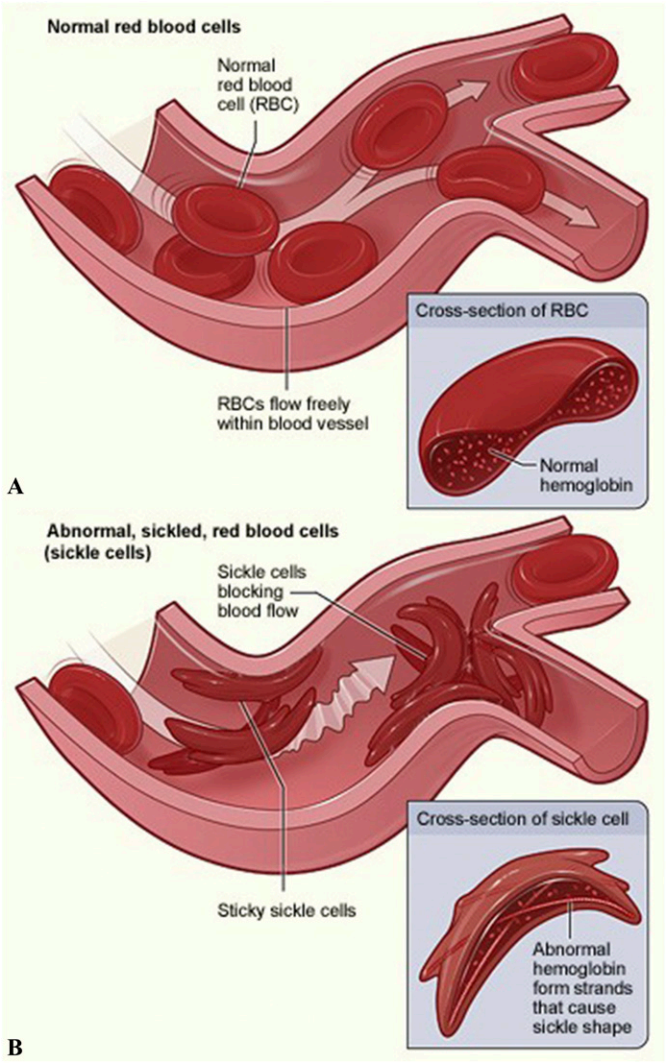

Figure 1. Sickle cell cross section.

exchange transfusions may reduce HbS levels to less than 30%, which is generally accepted as a safe level. Exchange transfusion, generally performed 24 hours or more before the operation, requires planning and is by nature not completely effective in removing HbS and replacing it with normal hemoglobin.

DESCRIPTION

A 61-year-old African American female with hemoglobin SC disease presented with coronary artery disease. Her body surface area was 1.84 m^2 with an approximate circulating blood volume of 5,200 mL. She was a known case of sickle cell anemia with a history suggestive of sickling crisis in the past. The patient had no blood antibodies and had never been transfused. Upon admission, hemoglobin electrophoresis was performed which revealed HbS—47.3%, hemoglobin C (HbC)—45.3%, and HbA—3.3%.

Case Discussion

On arrival into the operating room, the patient was put under anesthesia, prepared, and draped in a normal fashion. One unit of autologous blood was removed and sequestered using the Xtra cell saver (LivaNova, Houston, TX) to separate the plasma and red cells. The plasma was separated into platelet poor plasma (PPP) and platelet rich plasma (PRP). The PPP was sequestered and saved in an effort to preserve clotting factors and readministered to the patient after bypass. The PRP was applied to the sternotomy in combination with topical thrombin upon closing the chest. The patient's red cells were discarded.

The circuit consisted of the Capiox FX 25 oxygenator and cardiotomy (Terumo Cardiovascular Group, Ann Arbor, MI), Revolution centrifugal pump (LivaNova), and the Vanguard 4:1 cardioplegia set (LivaNova). The Hemostasis Management System Plus (Medtronic, Minneapolis, MN) was used for heparin management throughout the case. The CPB circuit was primed with 1,200 mL of Plasmalyte solution, four units of fresh donor red cells (collected within 3 days to ensure low K+), two units fresh frozen plasma, 400 mL autologous plasma, 200 mL albumin 25%, 1 g $CaCl_2$, 75 mEq 7.5% sodium bicarbonate, and 30,000 units heparin for a total prime volume of 3,800 mL. After the circuit was primed, the fluid was circulated through the oxygenator to achieve adequate oxygenation and warming of blood. Analysis of prime fluid was optimized showing PaO_2 greater than 400 mmHg, hematocrit of 22%, potassium 5.5 mmol/dL, and activated clotting time greater than 1,000 seconds.

After the patient was systemically heparinized with 25,000 units of heparin, the aorta and atrium were cannulated. At the initiation of CPB, exchange transfusion was performed to reduce HbS and HbC levels and raise adult hemoglobin level.

Cell saver was not used until after the initiation of bypass, after exchange transfusion had taken place to reduce HbS collection and reinfusion.

Two venous reservoirs were connected with a Y-connector to the venous line of the CPB system to enable the perfusionist to sequester 65–70% of the patient's blood volume before CPB was initiated. The venous line was drained until 3500 mL of autologous blood was removed and collected into the "discard" venous reservoir. The blood prime was infused simultaneously ensuring maintenance of hemodynamic stability. The collected blood was then sequestered using the cell saver to separate the red cells and plasma. The red cells were once again discarded and the plasma was saved (Figure 2).

CPB was initiated with flows maintained greater than a 2.2 L/min cardiac index and mild hypothermia at 34

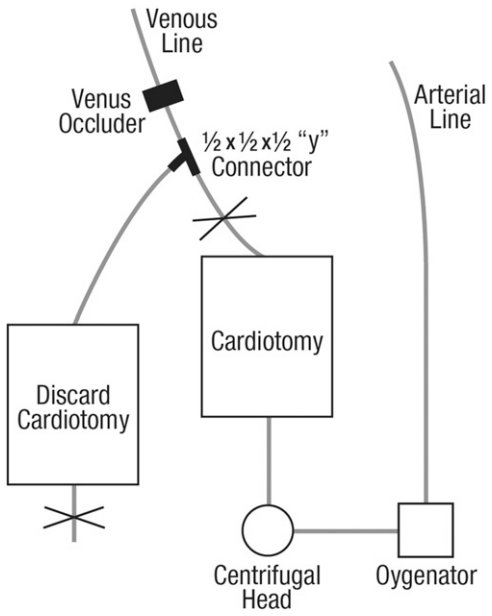

Figure 2. Sickle cell perfusion setup.

degrees was induced. The heart was arrested with an antegrade induction of warm, 4:1 blood cardioplegia followed by cold, 6°C blood cardioplegia. Subsequent doses of cold, retrograde cardioplegia were performed every 20 minutes or less. Continuous hemofiltration was performed during bypass. An additional unit of autologous pRBCs and 1,400 mL of autologous plasma was added during the bypass run. Venous saturation was maintained greater than 75% and pO2 was maintained greater than 350 mmHg. Blood gases were optimized to maintain hematocrit levels greater than 20% and a pH of 7.4. A warm dose of blood cardioplegia was administered before removal of the cross clamp. The patient was successfully weaned from bypass, and heparin was reversed with protamine.

After decannulation, the remaining blood in the reservoir was sent to the cell saver to be processed. This was tended to as any standard case as the remaining blood was hemoglobin A. Anesthesia administered cell salvaged blood, the remaining autologous plasma as well as two units of platelets, two units of FFP, two units of pRBCs and 700 mL of washed cell saver blood from the pump circuit. Once all products were administered, a final blood gas was drawn before the patient left the room reflecting hemoglobin of 7 g/dL.

Blood samples were collected after bypass and sent off for hemoglobin electrophoresis to be performed at an outside laboratory. The results were received within 3 days and reflected HbS $\cong$ 13.1%, HbC = 12.6%, and HbA $\cong$ 70.9%.

There was no episode of vascular occlusive complications postoperatively. The patient's recovery was uneventful, and extubation was performed within 5 hours of surgery. The patient was ambulating in the unit on postoperative day 1. No further blood products were administered in the ICU. The patient was discharged home on postoperative day 7.

COMMENT

We describe a method of intraoperative exchange transfusion with an attempt to lower the HbS and HbC levels while salvaging the patient's plasma and clotting factors from the discarded blood. The steps and techniques we used reduced the chance of a sickling event and enhanced our procedure.

Other measures the perfusionist can undertake to optimize the bypass run include reducing stagnant blood, avoiding cell saver and sucker usage until exchange transfusion is complete, maintaining high blood flows, maintaining high pO2 levels, and treatment of acidosis. During this case, flows were maintained with a cardiac index of at least 2.2 L/min. These high flows helped us maintain a venous saturation of >75% without acidosis. The use of the cell saver to separate the pRBCs and plasma was beneficial in having the patient's clotting factors available. This case shows that, with advance planning on the part of the surgical team, patients with SCD can safely and successfully undergo elective cardiac surgical procedures.

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