Case Report

Cardiopulmonary Bypass in the Sickle Cell Anemia Patient using Profound Hypothermia and Circulatory Arrest: A Case Report

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ABSTRACT

A homozygous sickle cell anemia patient undergoing a pulmonary thromboendarterectomy required the use of profound hypothermia and circulatory arrest. Reports of sickling crises have been documented under conditions of hypoxemia, acidosis, hypothermia, hypovolemia, and blood trauma. This patient's management included preoperative and intraoperative exchange transfusion, increased blood flow rates and optimizing blood gas values to prevent the sickling environment.

The pulmonary thromboendarterectomy surgery was successful in reducing pulmonary hypertension in this sickle cell patient. Using these techniques, no adverse sickling effects resulted from the profound hypothermia and circulatory arrest.

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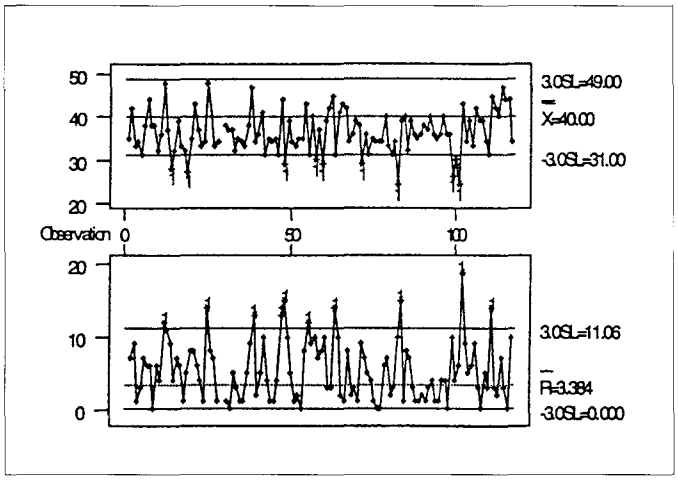
INTRODUCTION

Sickle cell anemia is a genetic disorder where the abnormal hemoglobin, hemoglobin S (Hg S), can deform to a sickle shape under conditions of hypoxemia, hypoperfusion, acidosis, hypothermia, or blood trauma. The result of this process is vessel occlusion and organ dysfunction (1). As the life span of a sickle cell anemia patient increases with improved health care, more need cardiac surgery using cardiopulmonary bypass (CPB) (2). Reports of open heart surgery in these challenging patients have identified the need for therapeutic intervention for the best outcomes (3-11).

Management of this patient included preoperative and intraoperative exchange transfusion, increased blood flow rates, optimizing blood gas values, and reducing blood trauma. This is the first reported successful use of profound hypothermia and circulatory arrest on the sickle cell anemia patient without detrimental sequelae.

Figure 1: Modifications to ECC



ECC = Extracorporeal circuit

CASE REPORT

In May of 1997, a 41-year-old African American female, BSA 1.56 m^2, was admitted with significant dyspnea on exertion, and was scored New York Heart Association class III. She had been diagnosed with homozygous sickle cell anemia since childhood and had painful crises until 1981, when her family moved from Haiti to New York. Episodes of shortness of breath had occurred over the past fifteen years and worsened over the past three years. Her differential diagnosis was pneumonia. After she was diagnosed with a pulmonary embolism and medicated with warfarin, she was referred for further evaluation.

Evidence of pulmonary hypertension on admission was confirmed upon completion of the following tests. Right heart catheterization demonstrated a right atrial pressure of 3 mmHg, pulmonary artery pressure of 70/20 mmHg, wedge pressure of 11 mmHg, thermodilution cardiac output of 5.04 L/min, and a pulmonary vascular resistance of 429 dynes/sec/cm^{-5}. Echocardiogram revealed right atrial and right ventricular enlargement. After two minutes of walking on a treadmill at 1 mi/hr, her arterial oxygen saturation decreased from 93% to 89%. Arterial blood gases on room air while resting were pH 7.45, pO_2 75 mmHg, and pCO_2 33 mmHg. Hematocrit (Hct) was 33.4% with hemoglobin electrophoresis revealing an Hb S of 88.5%. On her ventilation/perfusion scan, no perfusion was noted to the superior segments of the right lower lobe. Perfusion defects were noted in the left upper and lingular lobes. Pulmonary angiogram confirmed the absent perfusion in the right superior segment and "web" defects in the left lower and superior lingular lobes. Diagnosis of chronic pulmonary thromboembolic disease with pulmonary hypertension was made, and she was scheduled for a pulmonary thromboendarterectomy (PTE).

One day prior to surgery, a preoperative exchange transfusion was performed using the Cobe Spectra[a]. This is a continuous flow centrifugal system. The desired results can be programmed in for automated exchange of the erythrocytes. The final Hb S percentage, a calculation by the Cobe Spectra, was derived by the following formula:

$$\text{Hb S\% final} = 1 - \frac{\text{PRBCV x Hct replaced}}{\text{TBV x Hct initial + PRBVC x Hct replaced}}$$

where Hb S% = hemoglobin S percentage, PRBCV = packed red blood cell volume, Hct = hematocrit, and TBV = total blood volume.

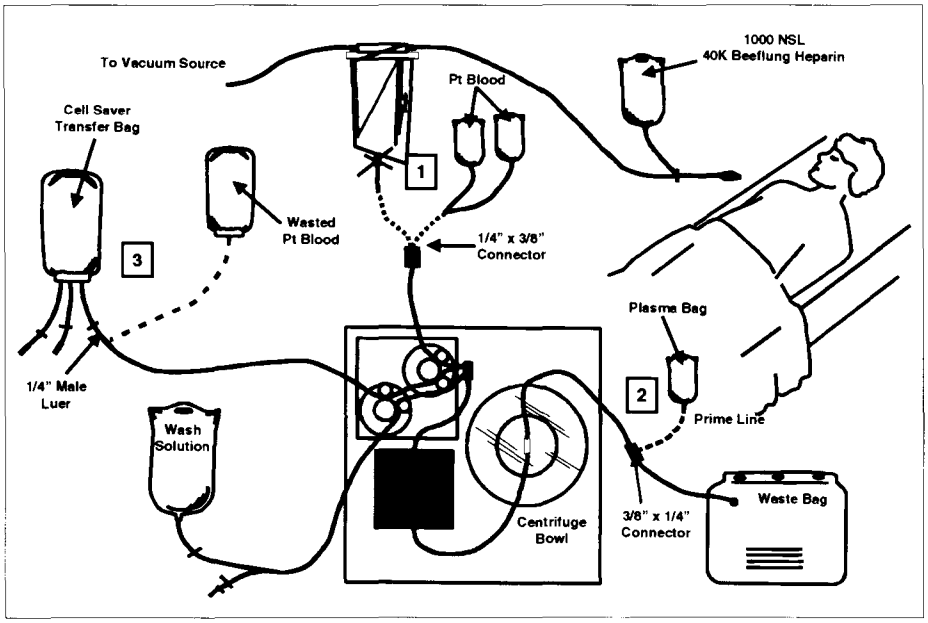
Eight units of packed red blood cells were used to increase the Hct from 28% to 33% and to reduce the Hb S from 88.5% to 12%. When the patient was brought to the operating room, preparation for CPB was followed in the usual fashion.

The use of a roller pump, closed membrane oxygenator system, left atrial and pulmonary vent, cardiotomy suction, and myocardial cooling jacket were standard components of the setup. Modifications were made in the venous line of the CPB circuit and the autotransfusion circuit. The venous drainage line modification was made for gravity exsanguination of the patient's blood. Figure 1 shows the 1/2" x 1/2" luer connector cut into the 1/2" venous line, close to the inlet of the venous reservoir for the perfusionist's ease of access. A double-spiked prime line with empty solution bags was connected by way of a 1/4" male luer adaptor and stopcock to the in-line 1/2" luer connector. The collected blood was processed through an autotransfusion device[b] for plasma sequestration and disposal of the red

a COBE Spectra, COBE Blood Component Technology, Inc., Lakewood, CO 80126

b Haemonetics Plus, Haemonetics Inc., Braintree, MA 02184

Figure 2: ATS Setup



ATS = autotransfusion; K = x 1000; NSL = normal saline liter; Pt = patient; 1 = cardiotomy; 2 = waste bag; 3 = reinfusion bag

cell component. Figure 2 shows the changes to the autotransfusion circuit at the cardiotomy, waste bag, and reinfusion bag. This made two separate circuits, one for blood collected during the exchange transfusion and the other for blood collected at the operative field. For the exchange transfusion, the cardiotomy reservoir outlet 1/4" tubing stub was clamped, and a 1/4" by 3/8" connector was placed in the intake line to the prime line with the patient's collected blood in the solution bags. The waste outlet line collected the salvaged plasma using a 1/4" x 3/8" connector to a spike line into a transfer bag, while the original waste bag was clamped off. The reinfusion bag was separated and clamped off. Again, a 1/4" x 3/8" connector to a spike line into a transfer bag was used to collect the exsanguinated red cell volume for disposal.

After heparinization and at the beginning of CPB, the perfusionist performed the intraoperative exchange transfusion. The patient's blood was gravity drained from the venous line into the empty solution bags. Adequate blood pressure support was maintained by transfusion of pump volume in equal amounts to the sequestered volume. Additional fresh frozen plasma, albumin, and crystalloid volume were added to the standard 1850 ml crystalloid/colloid pump prime. The components acted to flush the patient's body of her own blood. The process took an estimated 20 minutes to remove 4000 ml of patient blood volume, 3000 ml prior to CPB, and 1000 ml on initiation of CPB. Full CPB was initiated when the blood pressure became unstable. The patient's blood was processed in the manual mode and 2500 ml plasma was sequestered for later use. The patient's red blood cell volume was discarded.

Standard CPB protocol for pulmonary thromboendarterectomy, as previously described, was followed in preparation for periods of circulatory arrest (12). The patient was cooled to a core temperature of 20°C, maintaining no more than 10°C difference between blood and core temperatures. The first blood gas analysis, taken when the patient was at a rectal temperature of 35.5°C, shortly after initiation, revealed a metabolic acidosis (pH 7.355, pCO_2 33, pO_2 328, base deficient -7), venous saturation of 70%, and an Hct of 9%. Sodium bicarbonate and 500 ml of packed red blood cells were given to correct the pH to 7.50 and the Hct to 18%. Blood flow rates were kept at a 2.4 L/min/m^2 cardiac index or greater throughout the case. Phenytoin, sodium pentothal, and ice bags around the head were used for protection of the brain.

When the patient's core temperature reached 20°C, the cross clamp was placed on the aorta and 1000 ml of 4:1 blood cardioplegia was given. A cooling jacket around the heart was used for additional myocardial protection. Surgically, the intimal layers were dissected and removed from the right and left pulmonary arteries with circulatory arrest periods of less than 20 min for each side. After 16 min of circulatory arrest for endarterectomy of the right pulmonary artery, CPB was reinstated for 47 min as the right pulmonary artery was repaired and closed. This allowed the venous saturation to be restored to maintain at greater than 90%; the pH returned to 7.41-7.46 and the core temperature to 20°C.

After the second arrest period for endarterectomy of the left pulmonary artery, warming was begun using the 10°C temperature difference maximum between blood and core temperature. Four units, 1000 ml, packed red blood cells and the patient's plasma were given back through the pump circuit. Hemoconcentration was employed, removing 5000 ml of extracellular fluid. This increased the Hct from 18% to 29%. After the cross clamp was removed, a spontaneous normal sinus rhythm resulted. At a core temperature of 37°C, the patient was separated from bypass without difficulty. The arterial blood gas values just prior to termination were: pH 7.48, pCO_2 38 mmHg, pO_2 253 mmHg, base excess +5, and Hct 29%. Total cardiopulmonary bypass time was 252 min, with a cross clamp time of 113 min and a circulatory arrest time of 33 min.

The postoperative course for this patient with homozygous sickle cell anemia was standard for PTE patients. She was extu-

bated on the second postoperative day and transferred out of the intensive care unit. Her postoperative hemodynamic values were: pulmonary vascular resistance 184 dynes/sec/cm^{-5}, pulmonary artery pressure 39/13 mmHg, central venous pressure 7 mmHg, and cardiac output 6.36 L/min. No sickle cell crises or other complications were noted.

DISCUSSION

Improved health care has helped the sickle cell anemia patient live longer. As a result, more sickle cell patients are coming to the operating room needing cardiac surgery with the use of cardiopulmonary bypass. A rare and select group of these patients will require a PTE using profound hypothermia and circulatory arrest for treatment of their chronic pulmonary thromboembolic disease. The reported incidence of pulmonary embolism resulting in chronic pulmonary hypertension is 0.5% to 4% (13). The small subset of this percentage that is sickle cell patients is unknown. There are many reports of the use of cardiopulmonary bypass in sickle cell anemia patients, mostly for valvular repair (2-11). The PTE procedure differs in the use of profound hypothermia and circulatory arrest. Dangers of sickling and hemolysis, during and after CPB, have been reported to be increased in areas of stagnant blood flow and impaired microcirculation. Both of these conditions occur during circulatory arrest.

Reducing the Hb S to less than 30% of the total hemoglobin has been suggested as a level where sickling is hindered and undesired effects are not seen (14, 15). The current recommendation is to reduce the Hb S to 5% and treat the anemia just before CPB (2, 16). The benefit of this therapy is improved oxygen carrying capacity of the blood. This is accomplished by correcting the anemia and deficiency of Hb A, while also suppressing the production of Hb S. The preoperative exchange transfusion allows for an increased margin of safety during the induction of anesthesia and prior to CPB (1, 3).

Perfusion management emphasized optimizing tissue perfusion by minimizing the environment where sickling has been shown to occur. By keeping the blood flow rate at a cardiac index of 2.4 L/min/m^2 or greater, a short capillary time was achieved. The venous saturation was 70% or greater throughout the case. Oxygenation was closely monitored, especially after surgery when atelectasis and reperfusion edema may occur. Phenylephine was not used to help ensure optimal tissue perfusion.

To our knowledge, the use of profound hypothermia and circulatory arrest for a patient with homozygous sickle cell disease has not been described in the literature. The effect of hypothermia on the sickle cell patient is not clear. Hypothermia does increase blood viscosity and presumably sickling. Malfa and Steinherdt report that hypothermia has little effect on Hb S and may inhibit sickling (17). The preoperative and intraoperative exchange transfusions made possible the optimal patient conditioning for this surgery based on our present knowledge. More research is clearly needed on the effects of hypothermia and circulatory arrest in the sickle cell anemia patient.

Blood trauma, as always, should be avoided, since hemolysis increases sickling. Blood and air interface was minimized by use of a closed venous reservoir, regulating the speed of the pulmonary and left atrial vents, and cardiotomy suction. The cardiotomy suction was not used until after the intraoperative exchange transfusion was complete. Theoretically, use of a heparin-bonded circuit could reduce thrombus formation and inflammatory response (18). However, one was not used for this case due to the urgency of the sickle cell patient's condition and the standard CPB circuit had already been prepared.

More patients with sickle cell anemia are having successful cardiac surgery using CPB. This homozygous sickle cell patient had profound hypothermia with circulatory arrest to endarterectomize chronic thrombi from her pulmonary arteries. Her operation was successful in reducing her pulmonary hypertension. Profound hypothermia and circulatory arrest did not appear to have adverse sickling effects.

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