JECT. 2001;33:243–244
The Journal of The American Society of Extra-Corporeal Technology

Case Report

Cardiopulmonary Bypass with Deep Hypothermic Circulatory Arrest for a Patient with Sickle Cell Anemia: A Case Report

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Presented at the 39th International Conference of the American Society of Extra-Corporeal Technology, March 22–25, 2001, Miami, FL

Abstract: A 36-year-old sickle cell anemia patient undergoing a pulmonary thromboendarterectomy required the use of cardiopulmonary bypass incorporating deep hypothermic circulatory arrest. Being aware of reported incidences of sickling crises, a team of the surgeon, anesthesiologist, hematologist, and perfusionist met to devise a plan of treatment. Treatment included preoperative and intraoperative exchange transfusion, optimal blood gas management, and increased blood flows during bypass. The surgical procedure was performed and was successful in reducing pulmonary hypertension, incorporating a team approach and utilizing these techniques. No incidence of adverse sickling events was observed during this procedure. **Keywords:** sickle cell anemia, deep hypothermic circulatory arrest, cardiopulmonary bypass, pulmonary thromboendarterectomy. *JECT. 2001;33:243–244*

The sickle cell anemia patient poses a particular problem for perfusionists. The conditions that are likely to induce sickling, including anesthesia, hypothermia, hypoxemia, acidosis, and blood trauma, are inherent aspects of cardiopulmonary bypass and circulatory arrest. The goal of the procedure is to minimize the risks of a sickling crisis while achieving the goals of this surgical procedure.

SC sickle cell disease is present in about 1% of the black population in the United States. Hemoglobin (Hb) S and HbC are present in these patients in equal amounts and they lack all ability to produce the normal HbA. A decreased solubility of HbS when deoxygenated can cause vaso-occlusion, hypoxia, and tissue damage in the microvasculature, all conditions that can lead to further sickling crises as well as thromboembolism. As a result of recurrent thromboembolic events, pulmonary hypertension is considered to be a relatively common secondary disease in older sickle cell patients compounded by further hypoxemia and increased incidence of sickling crises (1, 2).

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Received March 25, 2001; accepted October 21, 2001.

DESCRIPTION

The patient was a 36-year-old black female with SC sickle cell disease, chronic thromboembolic disease, pulmonary hypertension, and repeated sickling crises. The patient's worsening right heart failure resulted in increased sickling crises requiring partial exchange transfusions approximately every 2 weeks. At the time of her admission, her Hb levels were HbS 20% and HbC 18% having received a partial exchange transfusion 1 week earlier. The goal determined by the team was to reduce her combined HbS and HbC levels to less than 10% before cardiopulmonary bypass (Table 1).

The patient was taken to the operating room where appropriate monitoring lines were placed, and she was intubated, prepped, and draped in the usual sterile fashion. A Carmeda-coated bypass circuit consisting of a Maxima Plus oxygenator, a closed venous reservoir, a cardiotomy reservoir, an arterial filter, a Biomedicus centrifugal pump, and a venous saturation monitor (Medtronic, Minneapolis, MN) were in place. The circuit was modified for the final exchange transfusion with insertion of a 1/2 in × 1/2 in × 1/4 in connector into the venous line, which was then attached to an empty 3-L IV bag (Figure 1). The circuit was primed with Plasmalyte-A (Baxter Healthcare Corp., Deerfield, IL) and recirculated while eight units of homologous blood were sequestered in the cardiotomy

Article available at https://ject.edpsciences.org or https://doi.org/10.1051/ject/2001334243

Table 1. Laboratory hemoglobin levels.

	HbS Level	HbC Level
Admission (26 days preop)	20%	18%
Day of surgery	9%	11%
4 days postop	4%	4%

reservoir. A median sternotomy was performed, and the patient was systemically heparinized for a target activated clotting time (ACT) of 450 s. The venous cannulas were placed in the inferior and superior vena cava. The arterial cannula was placed in the ascending aorta.

Directly before the initiation of bypass, approximately 6 L of blood were slowly removed into the empty volume bags while replaced with crystalloid prime through the aortic line. When sufficient volume was removed, bypass was initiated, and the homologous blood was added to the circuit, bringing the hematocrit to 30%. Using a 10°C gradient, the patient was cooled to 18°C and then the aorta was cross clamped and 1 L of cold antegrade blood cardioplegia was given.

Both pulmonary arteries were opened and thromboendarterectomy performed. The procedure required three short cardiac arrest periods lasting 10, 22, and 24 min. Between each arrest period, full flow was returned for at least 10 min or until the venous saturation returned above 90%.

The right pulmonary artery was opened and explored first. The surgeons found and excised a minimal amount of plaquing and obstruction. In the left pulmonary artery, a larger amount of thrombus was found and removed, particularly from the lower lobe. The pulmonary artery was closed, the aortic cross clamp was removed, and the rewarming process began. The patient was rewarmed using a 10°C gradient, and particular effort was made to maintain higher flows during rewarming to prevent any sickling problems. The patient was removed from cardiopulmonary bypass without incident.

DISCUSSION

The case demonstrates the ability of the surgical team to safely use cardiopulmonary bypass for patients with sickle cell anemia. The main challenge is to prevent sickling exasperated by hypothermia, hyperoxygenation, blood trauma, and decreased blood flow rates (3–5). As medical management and technologies improve lifespans in sickle cell patients, cardiopulmonary bypass procedures such as these will become a more common aspect of our profession.

Reducing HbS and HbC levels was key in safely using CPB for this patient. Govoni and Vichinsky, in separate

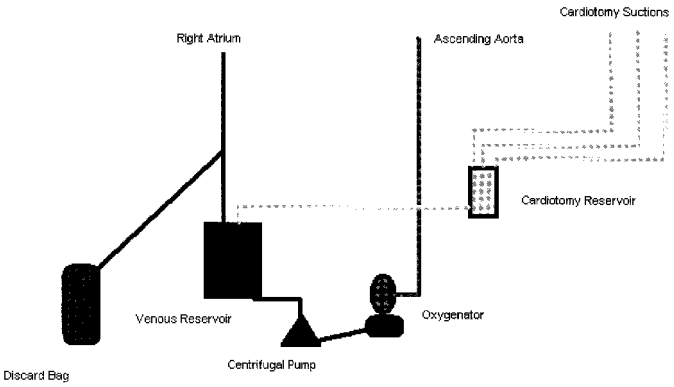

Figure 1. Circuit Diagram

studies, have suggested reducing the HbS to less than 30% of the total hemoglobin as a safe level where sickling is slowed down and the untoward effects are minimized, thus improving the oxygen-carrying capacity of the hemoglobin (6, 7, 8).

Should we be presented with another case of this nature, the authors feel it would be best to incorporate the CDI (Terumo Cardiovascular Systems, Ann Arbor, MI) blood gas monitor in the circuit. This would allow for better control of the pO_2 throughout all phases of the procedure.

The point that cannot be overstressed is the importance of teamwork and a proactive approach to the procedure. The ability to perform CPB on this patient without incident is a result of preplanning and communication among all members of the medical team.

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