
Exchange Transfusion Prior to Cardiopulmonary Bypass in Sickle Cell Anemia

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Introduction

During cardiopulmonary bypass (CPB) the maintenance of optimal cellular perfusion is an important goal. The conduct of CPB is complicated when the patient has sickle cell anemia, because perfusion may be jeopardized by sickle cell crisis and the attendant vaso-occlusion. The sickle cell crisis cycle¹ starts with hypoxia and sickling of cells and leads to vaso-occlusion and thrombosis. Several factors including acidosis and hypoxia may lead to the phenomenon. Critical maintenance of arterial pH and PaO₂ are needed to prevent the sickling of cells containing abnormal hemoglobin, precipitating the vaso-occlusive and thrombotic crises.² Hypothermia, a technique used often during CPB to increase safety margins and reduce metabolic requirements, causes sludging and sickling, and is usually avoided. The destructive effects of the sickling, crisis upon the cardiovascular and respiratory systems, kidney, liver, and brain should be readily apparent.³

Several preoperative and intraoperative procedures have been previously reported to prevent the onset of a sickling crisis. Hydration and alkalinization⁴, simple blood transfusions⁵, partial exchange transfusion^{6,7}, and maintenance of oxygen tension in the blood² have all been recommended.

This case report demonstrates a new technique for cardiopulmonary bypass that will eliminate the special management considerations recommended for the sickle cell disease patient requiring open heart surgery.

Case Report

History. The patient, a black female (MUSC No. 324298-07309) was first admitted during May of 1978 at 9 months of age to the Medical University of South Carolina for pneumonia. During a second admission to MUSC on August 10, 1978, she was diagnosed as having (1) sickle cell anemia, (2) ventricular septal defect (VSD), and (3) possible hemolytic or aplastic crisis. Because of bilateral ventricular hypertrophy (BVH), congestive heart failure (CHF), and suspected VSD, catheterization was performed providing the following data: cardiac index (CI) of 9.2 L/M²/min, left to right shunt with Qp/Qs of 2.2, LV pressure of 100/12 mm Hg. The remaining hemodynamic values were unremarkable. Post catheterization diagnosis was atrial and ventricular septal defects (ADS and VSD), and anomalous return of the left superior pulmonary vein to the superior vena cava. Surgical repair was recommended, but was postponed several times due to various complications: tonsillitis, otitis media, severe anemia with possible sickle cell crisis, and bacterial pneumonia.

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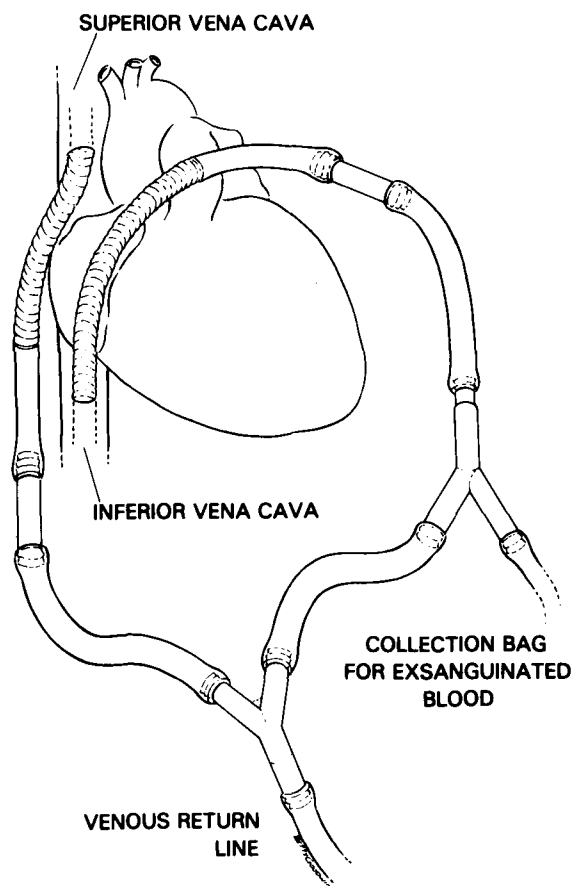


FIGURE 1. Venous drainage circuit used in cardiopulmonary bypass procedure. Strategically placed tubing clamps divert the flow of blood to the appropriate sites in the required sequence.

The patient was readmitted to MUSC on July 30, 1979. Repeat catheterization showed a $CI = 10.0$ L/M²/min, $Qp/Qs = 0.83$ indicating shunt reversal, LV pressure = 110/10 mm Hg, $Rp/Rs = 0.9$, Ao pressure = 110/70 mm Hg, main pulmonary artery pressure = 110/40 mm Hg, and hemoglobin = 9.0 g/dl. Because of the low hemoglobin, transfusion of 120 cc's of packed red cells (PRC) resulting in a hemoglobin of 11.4 g/dl was performed with volume control of diuretics. Due to rapid progression of her disease, she was scheduled for surgery.

Procedure. On the day of surgery, August 4, a new method of managing the sickle cell patient during CPB was utilized. The bypass system consisted of a 0.8 M² Sci-Med oxygenator, a Bentley Q220F cardiotomy reservoir, a Pall arterial line filter, a Travenol Pediatric Mini-Prime Heat Exchanger, and a Travenol Modular Heart-Lung Machine. The bypass circuit prime consisted of 1 unit fresh frozen plasma (FFP), 2 units PRC, 1600 cc's Lactated Ringer's solution, 40 mEq



FIGURE 2. Haemonetics Cell Saver, Haemonetics Corporation, Natick, Massachusetts.

NaHCO₃, and 5000 units of sodium heparin. The patient was heparinized (300 units/kg) and cannulated in the usual fashion. As CPB was instituted, all of the patient's blood was diverted via the venous return lines to a venous "tap" line, as shown in Figure 1. Approximately 700 cc's of the sickle cell blood was thus, directed into a reservoir bag. CPB proceeded routinely, including moderate hypothermia with esophageal and rectal temperatures of 27° and 32° centigrade respectively. Bypass remained unremarkable. The patient's blood was separated into red blood cells and platelet rich plasma (PRP) by a Haemonetics* Cell Saver (Figure 2). This PRP was collected from the Haemonetics Cell Saver at the site normally used for discard. The plasma fraction was saved, stored at room

* Haemonetics Corporation, Natick, Massachusetts.

TABLE I
Laboratory Data

Determination	Preop	Post pump	After autologous plasma fraction	3 hours post pump (1 FFP)	24 hours (platelet-48 hrs)
Platelets (thous/mm ³)	206	65	63	14	69
Fibrinogen (mg/dl)	253	47	115	177	
PT (sec)	14.2		15.4		16.7
PTT (sec)	25.1		34.7		38.1
Hgb (g/dl)	14.2	6.1	5.1	9.7	11.0

temperature, and reinfused immediately post bypass. The concentrated sickle cells were discarded. After termination of CPB, the circuit contents were fractionated in the normal fashion by the Haemonetics Cell Saver. Approximately 250 cc's of washed cells were given back to the patient in the operating room.

Immediately post pump, the patient was devoid of sickle cells. The platelet count and fibrinogen concentration were 65,000/mm³ and 47 mg/dl, respectively. The hematocrit was 19%. (See Table I.) After infusion of the autologous plasma fraction and 25 mg of protamine, the platelet count remained essentially unchanged, but the fibrinogen increased to 115 mg/dl. At three hours post pump and following the addition of 1 unit of FFP, the fibrinogen level increased to 177 mg/dl; the platelet count was 14,000/mm³; and the hemoglobin was 9.7 g/dl.

A twenty-four hour microscopic examination for sickle cells was negative. Poikilocytosis was noted, but this cell irregularity is commonly found post CPB. Hemoglobin electrophoresis showed a sickle hemoglobin concentration ([Hgb S]) of 18% on the day of discharge (10 days post operation).

A total of 900 cc's of PRC were administered to the patient during her entire hospital stay. This amount includes preoperative, pump prime, and post operative blood usage. (See Table II.) The post operative period was unremarkable with the chest tube drainage totaling 180 cc's.

Discussion

The critical substitution of one amino acid for another in hemoglobin is the molecular basis for sickle cell disease. The substitution in the protein moiety of hemoglobin, causes, at deoxygenation, precipitation and molecular deformation to occur, which subsequently results in the sickling of the red blood cell. The sickling/pO₂ relationship depends upon the concentration of sickle hemoglobin. In sickle cell disease, there is a [Hgb S] of 90% or greater and significant sickling at a pO₂ of 40 mm Hg, whereas, in the sickle cell trait there is a lower [Hgb S], <50%, and increased sickling is exhibited at a pO₂ of 30 mm Hg or below⁸. A sickle hemoglobin concentration of less than 40% will maintain normal microcirculation⁹.

Sickle hemoglobin has an O₂ dissociation curve that is shifted to the right, thus, has a decreased oxygen affinity. The avoidance of further oxygen dissociation curve shifts which accentuate a decrease in oxygen tension at tissue level and increased sickling is the basis for many of the management techniques advocated and used with the sickle cell patient. Thus, acidosis and lower levels of 2,3 diphosphoglycerate (2,3 DPG) should be prevented. Finally, hypothermia increases oxygen-hemoglobin affinity, but causes an elevated viscosity which potentiates vaso-occlusive crises and should be avoided.

After considering the aforementioned management

TABLE II
The Place and/or Time Blood Products were Used

Place and/or time	Blood product given
Pump prime	1 unit FFP
Pump prime	2 unit PRC
Post pump (12:00 Noon)	1 unit FFP
SICU (5:30 p.m.)	1 unit FFP
Patient room	85 cc PRC

recommendations and complexities, it was decided that the elimination of the causative factor would result in safer and simpler bypass. Total exchange transfusion removes nearly all of the sickle hemoglobin and allows for routine patient care during CPB.

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