A Technique for Mitral Valve Surgery in Sickle Cell Disease

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INTRODUCTION

Cardiac surgery in the patient with sickle cell disease (HbSS) presents several challenges, as many of the techniques used to safely perform cardiopulmonary bypass and to ensure myocardial protection may precipitate sickling and the complications thereof. Several strategies exist to prevent sickling during cardiac surgery requiring cardiopulmonary bypass. A case of a patient with HbSS requiring mitral valve repair is presented, along with a discussion of the approach to this case.

Keywords: Mitral valve surgery, sickle cell disease

CASE REPORT

A 15-year old Trinidadian male with sickle cell disease was diagnosed with acute rheumatic fever at age 11 years. He was treated for his acute presentation but did not receive antibiotic prophylaxis. Two years later, he developed exertional dyspnoea and palpitations on strenuous exercise. He was referred to a cardiologist and severe mitral regurgitation was diagnosed and confirmed by transthoracic echocardiography. He was referred to the Queen Elizabeth Hospital, Barbados, for mitral valve surgery. On clinical examination, he was jaundiced with hepatomegaly, pulse rate 88 beats per minute, regular, with the apex beat in the sixth left intercostal space in the anterior axillary line. There was a grade 5/6 pansystolic murmur loudest at the apex and radiating to the axilla, along with a palpable P2. Electrocardiogram confirmed sinus rhythm with a partial right bundle branch block. His haematocrit was 22.6%, haemoglobin (Hb) 7.0 g/dL.

On the day prior to surgery, he was transfused with two units of packed red blood cells. In the operating room, monitoring lines were placed which included a right radial arterial line, a right internal jugular vein Swan Gantz catheter, urethral catheter, rectal temperature probe and a transoesophageal echocardiogram probe. A partial exchange transfusion of two units of blood was performed. Transoesophageal echocardiogram confirmed Type 2 mitral regurgitation with excessive leaflet motion at A2.

A median sternotomy was performed and heparin 300 IU/kg given. Standard aortic and bicaval cannulation was performed and cardiopulmonary bypass commenced. A small right atriotomy was performed and a self-inflating retrograde coronary perfusion cannula was sited in the coron-

ary sinus under direct vision and sutured in place with a 4/0 prolene purse string suture. A vent catheter was placed in the proximal ascending aorta. The aorta was cross-clamped and retrograde warm blood coronary perfusion commenced. A left atriotomy was made after dissection in the interatrial groove, and the mitral valve examined. There was prolapse of the A2 segment of the anterior mitral valve leaflet with elongated chordae. There was no thickening of the leaflets or subvalvular apparatus, and no mitral annular calcification. There was annular dilation and a roughened area on the posterior leaflet corresponding to the mitral regurgitant jet. His mitral valve repair consisted of plication of the elongated primary chordae with 4/0 prolene sutures, and placement of a 36 mm rigid mitral annuloplasty ring. He was easily separated from cardiopulmonary bypass and decannulated without incident. Transoesophageal echocardiogram confirmed trivial mitral regurgitation post repair.

Carbon dioxide was used on the field throughout the procedure. A continuous infusion of milrinone was also used for the duration of the procedure. The rectal temperature was maintained between 37 and 37.3 °C throughout the procedure. Cardiopulmonary bypass time was 129 minutes, with an aortic cross-clamp time of 64 minutes, and a retrograde coronary perfusion time of 63 minutes.

Postoperatively, he did not require transfusion. Intermittent positive airway pressure noninvasive ventilation was used to treat atelectasis in the immediate postoperative period, and a right thoracocentesis was performed on postoperative day four and 500 mL of bloody fluid obtained. He was discharged on postoperative day eight.

DISCUSSION

The focus during surgery in patients with HbSS is prevention of hypothermia, hypoxaemia, acidosis, dehydration and low flow states, to prevent sickling in the perioperative period. The use of preoperative blood transfusion, exchange transfusions and one volume whole blood exchange transfusion to lower circulating levels of HbS and correct anaemia has been described (1–3). Cardiac surgery in this population presents unique problems as hypothermia, the cornerstone of myocardial protection, can cause vasoconstriction, sludging of cells with increased capillary transit time and increased oxygenation extraction, resulting in sickling.

In order to assess the mitral valve with some tone still in the papillary muscles, it is our practice to cool to 25 °C on cardiopulmonary bypass and allow the heart to fibrillate and then examine the mitral valve prior to cardioplegic arrest. As we wished to avoid hypothermia in this patient, we altered our strategy with the mitral valve being assessed on the beating empty heart. This confirmed the transoesophageal echo-

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cardiogram assessment of the anatomical cause of the mitral regurgitation.

There are cases of patients with sickle cell trait and disease successfully undergoing cardiopulmonary bypass with moderate hypothermia and cold cardioplegic cardiac arrest, and one report of a pulmonary thromboendarterectomy performed in two patients with deep hypothermic circulatory arrest, after exchange transfusion (4, 5). It was felt that the safer option in our setting with limited blood products was to maintain normothermia.

The use of retrograde blood coronary perfusion provided for myocardial oxygenation and provision of substrate, and may also flush sickled cells out of the coronary circulation. A similar technique utilizing antegrade coronary perfusion has also been described (6). We wanted to avoid any coronary malperfusion that might result from aortic root distortion by the mitral retractor with an antegrade aortic root perfusion technique, hence the use of retrograde perfusion.

The use of cardiopulmonary bypass with an in-line blood gas monitoring system allowed for continuous monitoring of arterial and mixed venous oxygen saturations, the latter being a surrogate of adequate tissue oxygenation. Haematocrit and acid base status were also monitored continuously throughout the bypass run. Prior to and post-separation from cardiopulmonary bypass, volume status was assessed by the pulmonary arterial diastolic and central venous pressures and assessment of left ventricular end diastolic volumes on transoesophageal echocardiogram.

The patient underwent a preoperative partial transfusion to reduce the percentage of circulating Hb S, and to raise his haematocrit prior to cardiopulmonary bypass. As our laboratory is unable to quantify the percentage of sickle cells, it was not known what percentage of circulating Hb S he had either pre or post-transfusion. In the operating theatre, to further reduce the circulating level of Hb S, two units of the patient's blood were removed prior to heparinization and replaced with two units of banked blood.

Reports of cardiac surgery in sickle cell patients without perioperative blood transfusion have been published (4). This avoids any chance of transfusion reactions. In our setting with limited blood bank resources, avoiding exchange transfusion in the future would be advantageous. A further modification of the blood management strategy that could be considered is the processing of the patient's removed units of blood to obtain plasma and platelets for retransfusion postoperatively (7). This will be of greater significance in more complex surgeries with longer cardiopulmonary bypass times, or if moderate hypothermia or circulatory arrest is necessary.

The preoperative decision to repair the mitral valve was confirmed both by transoesophageal echocardiogram and on inspection of the valve. The chordal elongation with normal leaflet morphology, absence of annular calcification without shortening or thickening of the subvalvular apparatus leant itself to a relatively easy repair.

CONCLUSION

The technique utilized in this patient resulted in a smooth operative course with no sequelae of sickle cell disease. The key components were normothermia, partial exchange transfusion, avoidance of cardiac arrest, an efficient, accurate operation and continuous monitoring. As sickle cell disease is common in this region, there are likely to be more cases requiring cardiac surgery, and also more complex cases. Given our limited blood bank resources, harvesting of platelets from the patient's blood after exchange transfusion would be useful, especially in cases requiring hypothermia or circulatory arrest.

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