Stage 1 Palliation for Hypoplastic Left Heart Syndrome Without the Use of Allogeneic Tissue, with Reduced Allogeneic Blood Product Exposure: A Case Report

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Abstract: In the 30 years since Norwood described the palliative procedure for hypoplastic left heart syndrome (HLHS), many modifications have been described which have increased the survival rate of children born with this lesion. We describe further modifications which result in reduced cardiopulmonary bypass time, no cooling or circulatory arrest time, and decreased banked blood exposure. A 16-day-old infant with HLHS undiagnosed during pregnancy presented for stage 1 palliation incorporating the Mee modification, Sano right ventricle to pulmonary artery conduit, dual arterial cannulation of the innominate artery and descending aorta, single venous cannulation of the right atrium, and a bypass prime volume of 130 mL. Anticoagulation and hemostasis were monitored with the Hepcon HMS Plus Hemostasis Management System (Medtronic USA, Minneapolis, MN). Bypass commenced at normothermia. A 5.0 Gore-Tex shunt was placed for the Sano Shunt, and the aortic arch was repaired without use of homologous tissue or synthetic material using a modification of the Mee technique. Bypass time was 92 minutes with a 10 minutes cardiac ischemic time. Modified ultrafiltration (MUF) was performed for 12 minutes and heparinization was reversed with protamine. There was no significant bleeding and no indication to transfuse clotting factors. The patient’s only allogeneic donor exposure was 350 mL of red blood cells during bypass necessary to achieve a post MUF hematocrit of 50% per our current institution policy for cyanotic infants. Using modified surgical and perfusion techniques along with low prime bypass circuits can result in reduced cross clamp and bypass times as well as a decrease in blood donor exposure. Hypothetical benefits include reduced operating room, ventilation, intensive care unit, and hospital times, improved neurodevelopmental outcomes, and an overall reduction in the cost of care for infants with HLHS. Keywords: cardiopulmonary bypass, hypoplastic left heart syndrome, low prime, Mee modification, Sano. JECT. 2011;43:258–260

OVERVIEW

In the 30 years since Norwood described the palliative procedure for hypoplastic left heart syndrome (HLHS), many modifications have been described which have increased the survival rate of children born with this lesion (1). First-stage palliative mortality for HLHS has fallen dramatically over the past decade, while that for second- and third-stage procedures remained stable. The cumulative operative mortality for three-staged repair has been reported to have fallen from 39–24% (2). First-stage palliative mortality has fallen significantly from 43–18% accounting for much of the overall reduction in operative mortality (2). Many have credited the increase in survivability to improved understanding of the single ventricle physiology, better preoperative and post operative care, superior cardiopulmonary bypass equipment, and techniques such as modified ultrafiltration (3). Alternatively, surgical techniques such as a right ventricular to pulmonary shunt instead of the historic right modified Blalock-Taussig shunt and utilization of the Mee modification for arch reconstruction have also been implicated in the much improved outcomes for the primary stage of reconstruction (4,5).

We describe further modifications which result in reduced cardiopulmonary bypass time, minimal cooling, no circulatory arrest time, and decreased banked blood exposure.
DESCRIPTION

A 1-day-old 3 kg baby presented to the hospital in extremis with a postnatal diagnosis of aortic atresia and dysfunctional hypoplastic left ventricle with right ventricle-dependent systemic flow. The patient presented to the hospital after spontaneous closure of the ductus arteriosus with acidosis, compromised perfusion, end organ dysfunction, and potential neurologic compromise. After prostaglandin initiation, hemodynamics gradually stabilized. End organ injury also stabilized (i.e., the patient was extubated, feeding, and had appropriate renal and mesenteric function) and at 2 weeks of age the baby presented to the operating theater for stage 1 palliation for HLHS.

Surgical Procedure

Through a midline sternotomy, the anterior pericardium was opened in the central portion with a punch and a 5 mm thin-walled Gore-Tex tube was sutured to that opening with running Prolene. The anterior pericardium surrounding the shunt was then harvested and fixed in 1.6% glutaraldehyde for 5 minutes. The pericardial tissue was then used on the distal end of the right ventricle to pulmonary artery shunt for anastomosis to the pulmonary arteries. The innominate artery was cannulated with an 8 fr Research Medical Aortic Arch Cannula (Edwards Lifesciences, Irvine, CA). A 16 fr right angled Research Medical venous cannula was inserted into the right atrium. The heart was retracted superiorly and the descending aorta was exposed and cannulated with an 8 fr Research Medical Cannula arterial cannula. The pulmonary artery branches were snared down and cardiopulmonary bypass was initiated. The patient was cooled to 32 degrees centigrade. The ductus arteriosus was doubly suture ligated and divided in its midportion. The main pulmonary artery was divided just before the arterial branches and the ductus stump was excised. A clamp was applied to the descending aorta beyond the isthmus. The left subclavian and left carotid arteries were snared and the mid transverse arch was snared. Adequacy of perfusion to the head was monitored continuously using a right radial arterial pressure line along with continuous non-invasive monitoring of cerebral saturation using the Somanetics INVOS 5100 monitor (Somanetics Corp., Troy, MI). The isthmus was excised. The posterior aspect of the descending aorta was spatulated longitudinally and the anastomosis was started to the greater curvature of the distal transverse arch. The anterior aspect of the descending aorta was then incised longitudinally for a few millimeters. The main pulmonary artery was correspondingly incised longitudinally and an anastomosis was begun, leaving space below the arch for the pulmonary artery bifurcation. When this was sutured up to the previously placed snare, the ventriculostomy was created for placement of the shunt. The atrial cannula was removed and using sucker bypass, the interatrial septum was excised. The atrial cannula was reinserted and regular venous drainage reinstated. A snare was applied behind the innominate artery cannula and a longitudinal incision was made on the underside of the transverse arch and down to the ascending aorta. A 2 mm Ostial Edwards Cardioplegia cannula was inserted in the ascending aorta and antegrade cardioplegia was administered. The cardioplegia cannula was removed. The incision in the aorta was continued down to the level of the pulmonary valve sinuses. The anastomosis across the underside of the transverse arch was then completed. No patch material was used. The arch was then deaired and the snares were removed restoring perfusion to the heart, which subsequently resumed spontaneous sinus rhythm. The patient was then rewarmed to 36 degrees centigrade and the proximal end of the Sano was doubled and sutured to the right ventriculostomy. With that complete, the patient was separated from cardiopulmonary bypass. Hemodynamics were appropriate with a balanced pulmonary blood flow to peripheral blood flow to give an SpO₂ of greater than 75% and a mean arterial pressure greater than 45 mmHg. Echocardiography verified the above findings without unexpected surgical complications. After modified ultrafiltration, heparin was reversed with protamine; dosing was calculated using the Medtronic Heparin Management System (Medtronic Inc., Minneapolis, MN). Decannulation and hemostasis were achieved without the use of random donor banked blood products. Atrial and ventricular pacing wires were placed and the patient was intermittently atrially paced during slow sinus activity. Atrioventricular conduction was intact. A trial of the chest closure resulted in slightly impaired hemodynamics and so decision was taken to leave the chest open.

Cardiopulmonary Bypass

The heart-lung machine used was the Sorin SV with the DMS data acquisition system (Sorin Group, Arvada, CO). The oxygenator used was the Terumo Baby FX (Terumo Cardiovascular, Ann Arbor, MI). The arterial line consisted of 1/8" polyvinyl chloride (PVC), the venous line of 3/16" PVC, and 3/16" boot tubing with Sorin Smart Coating. The Sorin CSC14 with blood to crystalloid ration of 4:1 was used for cardioplegia. Baxter Plegisol (Irvin, CA) was used as the crystalloid component with 70 mEq KCl and 15 mEq NaHCO₃ added. The Terumo HCOS5 hemofilter was used with 1/8" PVC tubing with Sorin Smart Coating on the inlet and outlet. The prime consisted of Baxter PlasmaLyteA as the crystalloid component, 15 mEq NaHCO₃, 120 mL washed packed red blood cells, 75 mL 25% human albumin, 5 mg/kg methylprednisolone, 25 mg CaCl₂, and 100 mg/kg tranexamic acid. The prime was hemofiltered down to a total of 180 mL prime volume. The prime volume includes the hemofilter and blood cardioplegia device which are crystalloid primed before use. When the arterial-venous loop is separated and tubing is
shortened to adequate length, the prime volume is further reduced giving a final static prime volume of the circuit of 130 cc. A prime blood gas was obtained to ascertain physiologic balanced acid/base and electrolytes with a starting hematocrit of 32%.

Two arterial cannulas were used and both were 8 fr to balance circulation to the head and the lower body. Two arterial pressure monitoring lines were placed, one in the right radial artery and one in the right femoral artery to adequately monitor the balance of perfusion throughout the procedure. Bypass was initiated and a Cardiac Index of 3.0 L/min/m² was targeted as adequate perfusion. Perfusion pressures above 35 mmHg were deemed appropriate for both the upper body and head pressure monitored using a right radial arterial line and for the lower body monitored using a femoral arterial line, making sure to not overflow the head. Should there be a flow distribution issue where the head is receiving more flow than the lower body, the surgeon places a partial occlusion tubing clamp to the tubing on the innominate artery cannula, increasing the resistance and redirecting flow to the lower body. Hematocrit was kept above 30% and was maintained using only 1 unit of packed red blood cells throughout the procedure.

Conventional ultrafiltration and zero balanced ultrafiltration were used throughout the procedure and the replacement fluid used was 2/3 buffered normal saline (1 L normal saline buffered with 30 mEq NaHCO₃) and 1/3 .45 buffered saline (1 L .45 saline buffered with 30 mEq NaHCO₃) to maintain normal sodium and chloride levels as well as remove any excess potassium from cardiopulmonia. Cardiopulmonary bypass time was 92 minutes, with a cross clamp time of 10 minutes and sucker bypass time of 1.5 minutes.

Comment

Pulmonary overcirculation through a systemic-pulmonary shunt has been one of the major causes of early death after the Norwood stage 1 procedure. To avoid this lethal complication, a right ventricle to pulmonary shunt in first-stage palliation of hypoplastic left heart syndrome has been widely used and is referred to as the Sano shunt (5). This modification to the historical Norwood stage 1 has arguably been given credit for the significant improvements in the outcomes in the past 10 years. Combining this technique with the Mee modification allows one to reconstruct the aortic arch with little patch material and without homograft material (1,4). In doing so, one may realize a stage 1 palliation without the use of homogenous graft material that may increase the immune response, which has the potential to cause marked immunologic sensitization, which may complicate potential future heart transplantation, if required (6). Using new generation, lower prime extracorporeal devices allows prime volume reduction, reduces surface area blood comes in contact with, and has the potential to reduce the number of blood product donor exposures as was seen here (7). Utilizing two arterial cannulas, one placed in the innominate artery and one in the descending aorta, just above the diaphragm, allows one to have two separate flow systems where the transverse aorta can be isolated, allowing for continuous flow to all the organs of the body during the aortic arch reconstruction of HLHS patients. By maintaining continuous perfusion, deep hypothermic circulatory arrest may be avoided thus preserving platelet and clotting factor function, decreasing or eliminating the need for clotting factor transfusion necessary to maintain hemostasis, decreasing the amount of time on bypass, and reducing the overall operating room time. Utilizing the Medtronic Heparin Management System has also been shown to reduce the amount of blood products needed during open heart procedures (8).

CONCLUSION

Using modified surgical and perfusion techniques along with low prime bypass circuits can result in reduced cross clamp and bypass times as well as a decrease in blood donor exposure. Hypothetical benefits include reduced operating room, ventilation, intensive care unit, and hospital time, improved neurodevelopmental outcomes from the potential shorter cardiopulmonary bypass times, reduced bank blood donor exposure, and an overall reduction in the cost of care for infants with HLHS.

REFERENCES