Reconstruction of the Aorta and Pulmonary Artery During Heart-Liver Transplantation in an Adult Congenital Patient

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Central Figure Legend

Patient with Fontan failure underwent a heart-liver transplant and aortic reconstruction.

Central Message

Aortic and pulmonary artery reconstruction can be safely performed during heart-liver transplantation for patients with Fontan failure and complex aortic pathology.
Case report

The patient provided written informed consent prior to publication of this report. Human subject research exemption was approved by IRB protocol ID #69579 on March 14th, 2023.

A 30-year-old female with hypoplastic left heart syndrome and coarctation of the aorta presented to our institution with Fontan failure. Her cardiac palliation included a Damus-Kaye-Stansel procedure (DKS) at 3 months of age, a bidirectional Glenn at 9 months, Fontan fenestration at 18 months, Fontan fenestration closure at 19 months, balloon angioplasty of a coarctation at 12 years, and coarctation stenting at 17 years of age. In total, the patient had three sternotomies. After an episode of ventricular tachycardia, she received an implantable cardioverter-defibrillator. Her Fontan failure was complicated by ventricular arrhythmias, poor exercise tolerance, fluid retention, chronic kidney disease, and Fontan-associated liver disease with portal hypertension, ascites, and varices. Given her disease progression, she was listed for combined heart-liver transplantation. In addition to extensive pulmonary and chest wall collaterals, she had aneurysmal dilatation of the ascending aorta to 5.9 cm x 4.9 cm. Extensive repair of the pulmonary artery (PA), as expected after Fontan circulation, would be accompanied by aortic reconstruction requiring a period of circulatory arrest.

When appropriate organs became available, the patient was taken to the operating room for a deceased donor combined heart-liver transplant. 5-French sheaths were placed into the right and left common femoral veins, left common femoral artery, and right internal jugular vein. An end-to-side anastomosis was performed to attach a 6mm Hemashield platinum perfusion graft to the small right axillary artery. Perfusion was established through the graft by a 3/8 to 1/4 connector.

A sternotomy continuous with a midline laparotomy incision was made and extended below the umbilicus. Dense mediastinal adhesions were carefully lysed with electrocautery.
Concurrently, the liver was mobilized. The superior vena cava (SVC) was cannulated with an 18-French cannula using sterile Seldinger technique. The right common femoral vein was cannulated with a 25-French multistage cannula inserted to the junction of the inferior vena cava (IVC) and right atrium. The patient was placed on cardiopulmonary bypass (CPB) and cooled. Upon reaching 28°C, circulatory arrest was initiated with retrograde cerebral perfusion (RCP). RCP is the authors’ preferred method of performing circulatory arrest and was used to flush out any debris while the initial dissection was performed. The heart and Fontan graft were excised, and the heart was removed.

After CPB was stopped, the aorta was divided just distal to the DKS. The heart and the Fontan graft were excised. The right main pulmonary artery defect from the Fontan connection was reconstructed with a Bovine pericardial patch using a running 5-0 polypropylene suture. Then, the innominate artery was clamped, and the patient was transitioned to selective antegrade cerebral perfusion (SACP) for cerebral protection by clamping the right innominate artery. Simultaneously, the SVC snare was released and the clamp on the SVC tubing was removed, thus halting RCP. The aortic resection was carried out using a peninsula-style technique. A 28mm single side-arm Gelweave graft was used for the hemiarch. There was vigorous blood return to the left atrium due to aortopulmonary collateral vessels, so the left atrial and pulmonary artery anastomoses were performed while the patient was on SACP (Figure 1).

The native pulmonary artery was opened and the atretic PA branches sized with a 25 mm Freestyle sizer. An appropriate graft was subsequently selected to reconstruct the PA. Then, the donor aorta to graft anastomosis was completed. After placing the patient in a steep Trendelenburg position, CPB was resumed via the side-arm graft, the body was de-aired, and the cross-clamp was re-applied to the graft, thus ending circulatory arrest. The aortic anastomosis was completed in an
end-to-end fashion while de-airing the heart. Then, the cross-clamp was removed, and the heart was reperfused. The IVC anastomosis was completed in an end-to-end fashion. The Glenn was then ligated at the right pulmonary artery, and the proximal end was controlled with a running 5-0 polypropylene suture. The SVC anastomosis was completed, the heart was de-aired, pacing wires were placed, and the root vent was removed.

Due to the complexity of the case, the liver transplant was performed after reperfusion of the heart, but while still on cardiopulmonary bypass. The IVC cannula was withdrawn below the infrahepatic vena cava to allow clamping of the IVC above and below the liver. A 21-French Medtronic venous cannula was placed into the portal vein and connected to a Y in the bypass circuit. The hepectectomy was completed and the allograft liver implanted in a standard bicaval fashion. Reperfusion of the liver on CPB was uneventful. The heart exhibited delayed graft function. An intra-aortic balloon pump (IABP) was placed, followed by initiation of veno-arterial extracorporeal membrane oxygenation (ECMO). The chest and abdomen were left open with the patient on VA ECMO to allow recovery of the transplanted organs. The CPB time was 365 minutes, circulatory arrest time was 87 minutes, RCP time was 8 minutes, SACP time was 79 minutes, the allograft total ischemic time was 429 minutes for the heart, and the allograft warm ischemic time was 46 minutes.

The abdomen was closed on post-operative day (POD) 1, and the chest was closed on POD 4. The IABP was removed after 8 days, and the patient was weaned from ECMO at POD 12. She was discharged on POD 64. As of six months post-transplantation, the patient is alive, had no episodes of rejection, no major adverse cardiac events, and no re-hospitalizations.

Discussion
This report describes for the first time the use of circulatory arrest during a combined heart-liver transplant and aortic reconstruction for a patient with failed Fontan physiology and an aortic aneurysm (Figure 2). Our patient had hypoplastic left heart syndrome and aortic coarctation, with a very complex past surgical history. Due to the significant advancements in the fields of pediatric cardiology and cardiothoracic surgery, patients with congenital heart disease are surviving to adulthood, with a burgeoning population of young adults presenting with failed Fontan physiology and the sequelae of end-stage organ failure. This provides a unique opportunity and challenge for cardiac surgeons, especially when these patients present with complex aortic pathology and require transplantation. However, there is relatively little published on combined heart-liver transplantation for these patients and even less published detailing how aortic pathology should be addressed. While considerable surgical planning is routine for these patients, unexpected complexity or complications require improvisation. In this case, the heart-liver transplant was complicated by extremely dense mediastinal adhesions and the need for reconstruction of the aorta and pulmonary artery. Accordingly, with the additional time required for circulatory arrest and the complicated reconstruction of the PA and aorta, the liver was kept in ice and implanted after reperfusion of the heart to prevent a long warm time, while maintaining CPB to rest the heart and protect it from reperfusion injury.

Circulatory arrest, implementation of both RCP and SACP, can be safely used during combined heart-liver transplantation in cases where patients have pathology of the aorta and pulmonary artery. These techniques utilized during the heart-liver transplant allowed safe reconstruction of the pulmonary artery and hemi-arch replacement.
Figure Legends

Figure 1: Photographs of the recipient’s heart and an intra-operative photograph. a) Anterior side of the patient’s heart. b) Posterior side of the recipient’s heart. c) Patient’s native aortic valve; the patient underwent the Damus-Kaye-Stanel procedure at 3 months old. d) Patient’s native pulmonary valve; the patient underwent the Damus-Kaye-Stanel procedure at 3 months old. e) Intra-operative photograph after the aortic reconstruction, pulmonary artery reconstruction, and donor’s heart was implanted. The liver transplant team prepares to implant the donor liver.

Figure 2: Pre-operative and post-operative echocardiograms and pre-operative CT angiograms. a) A two-dimensional transthoracic echocardiogram with color flow taken 7-months prior to transplantation. The systemic RV (purple arrow) and hypoplastic left ventricle (white arrow) can be visualized, as well as flow through the DKS and aorta. b) A two-dimensional transesophageal echocardiogram taken the day of surgery showing the system right ventricle (purple arrow) and hypoplastic left ventricle (white arrow). c) A two-dimensional transesophageal echocardiogram taken three weeks after transplantation, showing a well-functioning left ventricle, right ventricle, and aortic root. d) Pre-operative computed tomography (CT) angiogram showing the 58.6 mm x 48.9 mm aortic aneurysm. e) Pre-operative reconstruction from CT angiogram showing the patient’s anatomy.
References


