New Transplant Technique With Hemiazygos Continuation and Interrupted Inferior Vena Cava

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In the presence of left superior vena cava, cardiac transplantation is performed at the risk of left superior vena cava flow obstruction. In patients with hemiazygos continuation with interrupted inferior vena cava, the patency of left superior vena cava is vital. We introduced a new surgical technique to use a Fontan connection including the native central pulmonary artery as the systemic venous return. The pulmonary artery anastomosis was performed distal to the superior vena cava anastomosis sites. We used this treatment approach successfully to perform cardiac transplantation in two patients who developed severe heart failure after Fontan completion.


Cardiac transplantation in the presence of the left superior vena cava (SVC) has been managed with the technique of maintaining left SVC connection to the coronary sinus, left SVC to donor innominate vein anastomosis, or left SVC and donor right atrial appendage anastomosis [1, 2]. In the presence of hemiazygos continuation with interrupted inferior vena cava (IVC), patency of the left SVC is crucial after cardiac transplantation. This report presents a new cardiac transplant technique to manage the left SVC flow to the donor right atrium using a Fontan circuit in two patients.

Technique

In the first case (patient 1), a 9-year-old boy has the diagnosis of heterotaxy syndrome with bilateral SVC, interrupted IVC, hemiazygos continuation, common atrioventricular valve, and pulmonary atresia. He had a systemic to pulmonary shunt as a neonate and had bilateral bidirectional cavopulmonary anastomosis at the age of 19 months. At the age of 5 years, he had fenestrated extracardiac Fontan completion using an 18-mm polytetrafluoroethylene (PTFE) conduit (Fig 1A). Two years after the Fontan procedure, the patient developed severe ventricular dysfunction with an elevated end-diastolic pressure of 23 mm Hg. He was listed for cardiac transplantation. At the time of transplantation, branch pulmonary arteries were divided distal to the SVC insertion sites (Fig 1B). The proximal pulmonary artery sites were closed using a bovine pericardial patch. The donor right atrium was anastomosed to the PTFE conduit. Donor branch pulmonary arteries were recovered up to the first branching points (Fig 1C). The recipient left pulmonary artery was anastomosed directly to the donor left pulmonary artery. The posterior wall of the recipient right pulmonary artery was anastomosed directly to the donor right pulmonary artery. Owing to the long course of the right pulmonary artery, an additional donor distal ascending aortic wall patch was required for the anterior part of the right pulmonary artery reconstruction. Left atrium and ascending aortic anastomosis were performed in the regular fashion.

In the second case, (patient 2) a 26-year-old woman has the diagnosis of heterotaxy syndrome with left SVC, interrupted IVC, hemiazygos continuation, common atrioventricular valve, and single ventricle. At the age of 22 years, she had nonfenestrated extracardiac Fontan completion using a 22-mm PTFE conduit. Four years after Fontan completion, she developed left lung pulmonary arteriovenous fistula and had Fontan revision to intracardiac Fontan to bring the hepatic factor to the left lung. Eight months after Fontan revision (Fig 2A), she developed severe ventricular dysfunction with an elevated end-diastolic pressure of 26 mm Hg. She was listed for cardiac transplantation.

At the time of transplantation, the branch pulmonary arteries were divided distal to the left SVC insertion sites bilaterally (Fig 2B). The proximal pulmonary artery sites were closed using a bovine pericardial patch on the left side, and the right side was closed primarily. The donor right atrium was anastomosed to the PTFE Fontan conduit. The central pulmonary artery was created using a 16-mm PTFE tube from the initial Fontan conduit to the distal left pulmonary artery (Fig 2C). The donor main pulmonary artery was anastomosed to this PTFE tube with end-to-side technique. The left atrium

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and ascending aortic anastomosis were performed in the regular fashion.

Postoperative evaluation with echocardiography and cardiac catheterization showed no stenosis of the Fontan conduit, right atrium, or branch pulmonary arteries and no significant tricuspid valve regurgitation in either case (Fig 3).

**Comment**

The left SVC obstruction can be managed with a stent or balloon at the time of elective catheterization in the majority of cases after cardiac transplantation in the presence of right SVC [1, 3]. In the patient with hemi-azygos continuation with interrupted IVC, the patency of

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**Fig 1.** (A) Systemic venous Fontan circulation was shown in this diagram. (B) The systemic venous return was kept using Fontan connection, and branch pulmonary arteries were divided distal to the superior vena cavae anastomosis sites. (C) The central pulmonary arteries were reconstructed using donor central pulmonary arteries.

**Fig 2.** (A) Systemic venous Fontan circulation was shown in this diagram. (B) The systemic venous return was kept using Fontan connection, and branch pulmonary arteries were divided distal to the superior vena cavae anastomosis site. (C) The central pulmonary arteries were reconstructed using a 16-mm PTFE tube. The donor main pulmonary was anastomosed to this PTFE tube. (PA = pulmonary artery; PTFE = polytetrafluoroethylene.)
the left SVC is crucial because of the lower body including the kidney vein draining to the left SVC through the hemiazygos vein. Vricelli et al [1] reported that 3 of 15 patients with situs inversus developed left SVC obstruction after left SVC to donor innominate vein anastomosis or innominate vein to donor SVC anastomosis. If the left SVC is anastomosed to the donor innominate vein in the setting of single ventricle palliation, the risk of left SVC obstruction can be higher because of the large size of the ascending aorta. This new technique avoids left SVC to donor systemic vein anastomosis and decreases long-term concern for left SVC obstruction.

For the first patient, in order to gain growth potential, the donor branch pulmonary arteries were used to reconstruct central pulmonary arteries. If the donor heart cannot be recovered with the branch pulmonary arteries, the central pulmonary artery can be managed with a synthetic tube or xenopericardial roll. However, the use of these materials creates a high potential for branch pulmonary artery stenosis after outgrowth. Because the second patient is an adult, a PTFE tube was used for central pulmonary artery reconstruction. Because reconstructed branch pulmonary arteries are located anterior to the native central pulmonary artery, the branch pulmonary stenosis is a reasonable concern. Neither of our cases showed any signs of branch pulmonary artery stenosis by echocardiography and cardiac catheterization postoperatively.

Because right atrial to Fontan conduit anastomosis is more similar to biatrial anastomosis compared to bivacal anastomosis, postoperative tricuspid valve function is a concern. In our series, neither patient had significant tricuspid regurgitation postoperatively. Long-term follow up is warranted.

References