Orthotopic Heart Transplantation in a Patient with Persistent Left Superior Vena Cava

Alexander Weymann, MD, Bastian Schmack, MD, Matthias Karck, MD, PhD, and Gabor Szabo, MD, PhD

We would like to discuss this rare case of an orthotopic heart transplantation in a recipient with an unknown history of persistent left superior vena cava.

Introduction

The persistent left superior vena cava (PLSVC) is a very rare, congenital venous heart abnormality with general prevalence of 0.3% and 1.5%–10% in patients with other congenital heart defects.1,2) PLSVC was first described in 1850 as a venous heart anomaly.3) PLSVC represents a part of the embryonic sinus venosus which usually closes and becomes the coronary sinus during normal development. In 90%, the PLSVC drains into the coronary sinus and then continues into the right atrium; the remainder drain to the left atrium, causing a right-left-shunt.4) In 80% of all cases, there is a right superior vena cava (SVC) present which drains into the right atrium, allowing the condition to go unnoticed.4,5) Other related deformities include a missing right SVC or left vena azygos, causing the right cranium and the right arm to drain through the innominate vein into the PLSVC.6) The secondary coronary sinus dilation can stretch the AV-node and the bundle of His resulting in cardiac arrhythmias or a partial mitral valve obstruction.7)

Case Report

A 45-year-old man presenting with dilative cardiomyopathy, a history of mechanical mitral valve replacement, and simultaneous dacron-patch closure of a high-seated membranous ventricular septal defect received a heart transplantation in our department.

Routine hemodynamic evaluations showed pulmonary hypertension with a mean pulmonary pressure of 40 mm Hg. Additional echocardiography showed severe tricuspid valve insufficiency and a severely impaired left ventricular ejection fraction of 10%, prior to the heart transplantation.

Through a routine midline sternotomy, the connection for cardiopulmonary bypass was established via arterial cannulation of the ascending aorta. The SVC and inferior vena cava (IVC) were directly cannulated. After tying off both vena cavae, an unexpected PLSVC was discovered. The PLSVC drained into the dilated coronary sinus and even appeared to be larger than the right SVC (Fig. 1A). We could not identify any communication through the innominate vein between the left and right side of the upper body. The PLSVC was pinched off with a tourniquet and cut transversally 2 cm away from the connection to the coronary sinus. The heart explantation then commenced routinely, with the dorsal part of the left atrium. The aorta and pulmonary artery were then severed, allowing adequate cuff-material for the anastomosis. Recipient and donor atria were prepared for atrial-to-atrial anastomosis. Following the excision of the right atrium, the atrial cuff was almost entirely preserved. The implantation of the donor organ began with the left atrium using the continuous suture technique. Afterwards, we performed an end-to-end anastomosis of the donor’s SVC.
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with the recipient’s isolated PLSVC (Fig. 1B and 1C). Following the incision ranging from the IVC to the SVC of the donor’s right atrium, the recipient’s preserved right atrial cuff was anastomosed. The remaining vessels were routinely anastomosed. Cardiopulmonary bypass time was 205 minutes, respectively. The patient presented in the intensive care unit with a stable sinus rhythm and minimal chest tube drainage. The postoperative trajectory was without complications, and the patient received standard immunosuppression. A postoperative angiographic examination showed regular drainage through the PLSVC.

A slew of innovative techniques had already been documented to preserve a PLSVC during orthotopic heart transplantation. The use of prosthesis between the right atrium and the innominate vein, as well as the ligation of the PLSVC, allows for conventional heart transplantation. The disadvantage of using this technique is the higher risk of a prosthesis-related infection due to the immunosuppression.8) Other techniques require the prior knowledge of this anomaly in order to prepare a segment of the donor’s SVC long enough to use for different anastomotic techniques.9) Another technique is to mobilize the PLSVC and anastomose the donor’s SVC by tunnelling through the sinus transversus.10)

In conclusion, the presented technique is shown to be simple and without complications while reducing heart lung machine time. Following this method, heart transplantation technique can be done routinely, allowing, to the benefit of the recipient, a shorter ischemic time span. Moreover, our technique preserves the PLSVC and does not require the use of prosthetic material. The recipient’s

A: The illustration shows the recipient’s opened chest cavity following heart explantation. Notable is the drainage of the persistent left superior vena cava into the right atrium through the coronary sinus close to the inferior vena cava. PLSVC indicates persistent left superior vena cava and SC sinus coronarius.

B: The schematic drawing depicts the completed anastomosis of the donor’s superior vena cava and the recipient’s persistent left superior vena cava. The recipient’s right atrial cuff remains intact and the persistent left superior vena cava can be preserved. PLSVC indicates persistent left superior vena cava and SVC superior vena cava.

C: Intraoperative view showing the key-anastomosis of the donor’s superior vena cava and the recipient’s persistent left superior vena cava. PLSVC: persistent left superior vena cava; SVC: superior vena cava.
right atrial cuff remains intact, and the donor’s SVC is anastomosed with the recipient’s PLSVC. Using the aforementioned operative technique, the preoperative knowledge of this anomaly has been rendered non-essential.

**Disclosure Statement**

None.

**References**


