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CASE REPORT

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Heart transplantation using a donor with partial anomalous pulmonary venous connection and atrial septal defect

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Abstract

Over the last decade, the shortage of donors has led to increased waiting time prior to transplantation and its related mortality. Therefore, extended criteria for donor hearts have been proposed. In this report, we describe a successful transplantation despite a diagnosis of partial abnormal pulmonary venous return associated with an atrial septal defect sinus venosus and persisting left-sided superior vena cava. Knowledge in congenital cardiac disease can broaden the definition of 'marginal' donor hearts and allow their use without increasing the risk of transplantation.

Keywords: Congenital heart disease • CHD • Organ donor management • Transplantation

INTRODUCTION

Current management of heart failure patients and limited donor availability has resulted in an increased waiting time for patients listed for cardiac transplantation [1], resulting in the development of 'extended donor criteria' [2]. Among those, the use of donor with cardiac congenital malformation such as simple atrial septal defects (ASDs) has been advocated. More complex congenital abnormalities are currently regarded as contraindication to transplantation.

In this report, we describe the use of a donor heart with partial abnormal pulmonary venous return, ASD-SV and a persisting left superior vena cava.

CASE REPORT

Our recipient was a 56-year-old male with a diagnosis of non-ischaemic dilated cardiomyopathy.

Pretransplant evaluation confirmed the absence of pulmonary hypertension (PVR 2UW, TPG 3 mmHg). The donor was an 18-year-old man who suffered a spontaneous intracranial bleeding. Donor–recipient weight and height were perfectly matched (100 vs 96 kg, 180 vs 178 cm). The donor was haemodynamically stable, without inotrope. Donor electrocardiogram revealed a sinus rhythm, QRS duration of 83 ms, and a normal ST segment. At transthoracic echocardiography, the right and left atria were not dilated, right ventricle was neither dilated nor hypertrophic, the left ventricle dimensions were normal (end-systolic diameter 31 mm, end-diastolic diameter 64 mm), with a left ventricle fraction shortening of 52%.

Prior to harvest, the cardiac team inspected the heart. First, a left superior vena cava was identified. Further dissection revealed that all pulmonary veins from the right lung drained into the right superior vena. Lungs were declined by the thoracic retrieval team. The right ventricle was not enlarged. Heart explantation was performed harvesting a long segment of the right superior vena cava together with a cuff of each pulmonary vein. The left superior vena cava was divided proximal to its drainage into the coronary sinus. *Ex situ* heart examination revealed a 1-cm-large sinus venosus ASD. Seen from the superior caval vein orifice, the septal crest of the ASD was clearly overriding, thereby reducing the superior vena cava-right atrium pathway.

Prior to transplantation, back-table reconstruction consisted of left superior vena cava stump closure. The azygos vein and the pulmonary veins cuffs on the right superior vena cava were similarly closed with running sutures. To allow a broader superior vena cava-right atrium pathway, the atrial septal wall was divided inferiorly over 2 cm and secondarily closed with a large heterologous pericardial patch (Fig. 1). The back-table time was 30 min. Graft ischaemic time was 224 min. The patient postoperative course was uneventful. No supra-ventricular arrhythmia occurred during hospitalization, and post-transplantation electrocardiogram showed a sinus rhythm with normal atrio-ventricular conduction.

Echocardiography at discharge revealed no residual shunt at the atrial level and laminar flow in the superior vena cava. Angio-

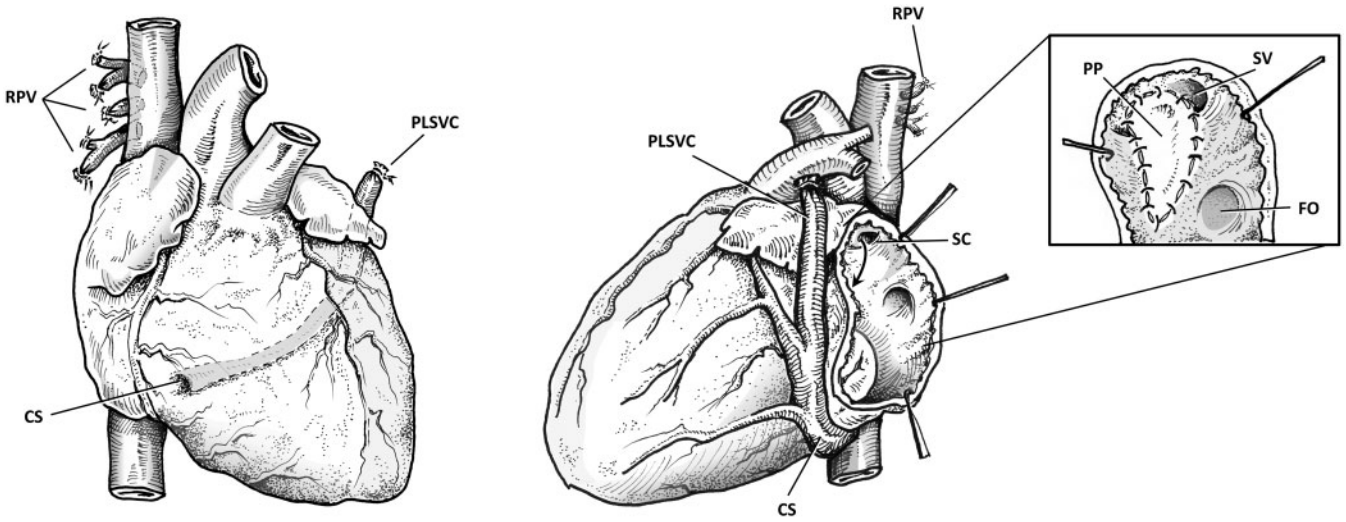

Figure 1: Anatomy of the variant heart transplant, in anterior (left) and postero-lateral (right) views. PLSVC: persistent left superior vena cava (ligated); RPV: right pulmonary veins ending in the SVC (ligated); SV: sinus venosus; SC: septal crest; FO: fossa ovalis; CS: coronarys sinus.

CT scan at 3 month confirmed unobstructed systemic venous pathways (not shown).

DISCUSSION

To palliate donor's shortage, many centres have extended their criteria for donor heart acceptance [3]. However, most congenital cardiac abnormalities (with the exception of minor defects such as simple ASDs) are regarded as contraindication to transplantation.

In our patient, none of the congenital cardiac abnormalities were detected during donor evaluation. The abnormal pulmonary venous return, the ASD sinus venosus and the persisting left superior vena cava were diagnosed at the time of retrieval.

Moons *et al.* [4] recently reported a transversal nationwide study on prevalence of congenital heart disease in the general population. Overall prevalence was 8.3‰. Among the most frequent diagnosis, ventricular septal defects (2.7‰) and 'ostium secundum' atrial septal defects (1.5‰). Some of those congenital heart diagnosis will remain undetected throughout life.

The ASD sinus venosus is a rare variant of atrial septal defect (0.03‰) that affects other area than the fossa ovalis (ASD secundum-type). Most often located at the superior vena cava junction, the defect is related to the orifice of the right superior pulmonary vein which drains directly into the superior caval vein. In our patient, all three pulmonary veins drained into the superior vena cava. In term of pathophysiology, most abnormal pulmonary venous return result in significant left-to-right shunts, with subsequent right-sided structures dilatation. In our patient, review of the cardiology report with a focus on signs of chronic volume overload revealed no sign of dilatation of either atrial or ventricular structures. It is likely that the overriding of the superior vena cava on the atrial septal crest, as identified after explantation, resulted in some streaming effect, allowing an even distribution of the blood flow from the superior vena cava into both atria.

In adults, sinus venosus defects can be difficult to visualize by transthoracic echocardiography so that when other abnormal anatomic findings are present, transoesophageal echocardiography should be performed to infirm intra-cardiac anomalies [5].

This case report illustrates the importance of complete assessment at the time of donor evaluation, as well as the importance of the surgical exploration. Medical and surgical expertise in congenital heart anomalies should allow for a broader definition of potential donor hearts and their use without increasing the risk for the recipients.

Conflict of interest: none declared.

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