Aortic arch and pulmonary artery reconstruction during heart transplantation after failed Fontan procedure

Ajay J. Iyengar, Varun J. Sharma, Yves d’Udekem and Igor E. Konstantinov*

Cardiothoracic Surgery, Royal Children’s Hospital, University of Melbourne, Murdoch Childrens Research Institute, Melbourne, Australia

* Corresponding author. Cardiothoracic Surgery, Royal Children’s Hospital, Flemington Road, Parkville 3052, Australia. Tel: +61 3 9345 5200; fax: +61 3 9345 6001; e-mail: igor.konstantinov@rch.org.au (I.E. Konstantinov).

Received 10 November 2013; received in revised form 18 December 2013; accepted 30 December 2013

Abstract

Recent significant improvement in surgical management of patients with univentricular palliations has resulted in an increasing number of patients with complex univentricular heart repairs surviving to heart transplantation. Heart transplantation in these patients can be challenging and may require extensive vascular reconstruction during transplantation. Herein, we describe the surgical reconstruction in a complex patient that was required prior to heart transplantation.

Keywords: Fontan procedure • Hypoplastic left heart syndrome • Congenital heart disease • Heart transplantation • Aortic root aneurysm

INTRODUCTION

Owing to improved surgical outcomes of patients with univentricular palliations, particularly those with hypoplastic left heart syndrome (HLHS), an increasing number of patients with univentricular physiology requires heart transplantation. As a result, the technical complexity of heart transplantation in congenital heart disease has increased during recent decades. Herein, we describe a patient with failed Fontan circulation, previous multiple procedures and aneurysmal dilatation of the neoaorta, who required complex reconstruction of the great vessels to make heart transplantation possible.

CASE DESCRIPTION

A 16-year old boy with HLHS presented with intractable heart failure, protein losing enteropathy and chyloptysis 3 years after Fontan completion. Prior palliations undertaken were neonatal Norwood procedure with modified Blalock-Taussig shunt, bidirectional cavopulmonary shunt with concomitant repair of a neoaortic root aneurysm and left pulmonary artery stenosis using homograft arterial patches at 1 year of age, transcatheter stenting of the left pulmonary artery at 12 years of age and, finally, extra-cardiac Fontan procedure with an 18 mm Gore-Tex conduit and 4 mm fenestration at 14 years of age. Due to persistent chylothorax, he required bilateral thoracotomies for pleurodesis after the Fontan procedure. Before transplantation, coiling of aortopulmonary collaterals and right lung arterio-venous malformations had been performed to attempt reduction of elevated pulmonary arterial pressures. However, these did not result in clinical improvement.

At the time of transplantation, the neoaortic root was severely enlarged and densely adherent to the previously stented left pulmonary artery (Fig. 1A). Bleeding from the aneurysmal neoaortic root necessitated en bloc resection and replacement of both pulmonary arteries from hilum to hilum (Fig. 1B), utilizing 14 and 10 mm Gore-Tex conduits (W.L. Gore & Associates, Newark, DE, USA), and the entire aortic arch (Fig. 1C), utilizing an 18 mm Gelweave prosthesis (Vascutek Terumo, Scotland, UK). The reconstruction was performed during 43 min of deep hypothermic circulatory arrest at 18°C. The total cardiopulmonary bypass (CPB) time was 351 min, aortic cross-clamp time 106 min and donor heart ischaemic time 109 min. It took ~7 h to perform sternotomy, divide adhesions, cannulate for CPB, excise the recipient heart and perform all vascular reconstructions before the donor heart could be implanted. The harvesting of the donor heart was appropriately postponed and coordinated with the progress of the recipient’s vascular reconstructions.

Following these reconstructions, transplantation was performed in a standard fashion. The patient remained in the hospital for 3 months due to immunosuppressive complications, including multifocal leukoencephalopathy, acute renal failure, interstitial pneumonia requiring tracheostomy and neutropenia. Subsequently, he recovered and is well 6 months after transplantation.

COMMENT

Early mortality after heart transplantation in the setting of univentricular congenital heart disease was initially 29% [1, 2]. Recent reports show improvements in short- and long-term outcomes [2, 3], likely due to technical improvements. However, very few descriptions of technical solutions exist [4], and some teams still reject such patients for consideration of transplantation due to anticipated technical difficulties. The surgeon’s efforts during heart transplantation after failed univentricular palliation are frequently hampered by the...
enormous technical challenges posed by exposure, cannulation and previous reconstructions. This case illustrates that these challenges may be overcome. Despite anomalies of both great vessels, complete reconstruction was possible under deep hypothermic circulatory arrest, enabling the remainder of the transplantation to proceed in a standard fashion. By utilizing prosthetic material to reconstruct the recipient’s great vessels and coordinating the timing of retrieval with the donor harvesting team, we were able to minimize the ischemic time of the donor heart. For this reason, even if the great vessels from the donor are available for harvesting, we prefer to reconstruct these structures in advance with prosthetic materials. Our current preference for reconstruction of the pulmonary arteries is to use Gore-Tex, although its long-term superiority over allograft or pericardium for the treatment of pulmonary artery discontinuity is untested [5].

In patients who have had multiple previous operations, transplant surgeons must be aware of the variety of reconstructive methods available to them. They must also be aware of the time taken to perform the complex dissection and reconstructions in order to plan organ retrieval and minimize donor heart ischemic time, as was achieved in this case. It is thus crucial that heart transplantation in these patients be performed by surgeons with expertise in the management of complex congenital heart disease.

ACKNOWLEDGEMENTS

The authors thank Bill Reid, medical illustrator, for his assistance in producing the figures in this publication. The authors acknowledge the support provided to the Murdoch Childrens Research Institute by the Victorian Government’s Operational Infrastructure Support Program.

Funding

This work was supported by postgraduate scholarships for Ajay Iyengar from the National Health and Medical Research Council & Heart Foundation of Australia [APP1038802] and Royal Australasian College of Surgeons [Foundation for Surgery Catherine Marie Enright Kelly and Eric Bishop Scholarships]. Yves d’Udekem is a Career Development Fellow of The National Heart Foundation of Australia [CR 10M 5339].

Conflicts of interest: none declared.

REFERENCES