Surgical Management of Giant Pulmonary Artery Aneurysm in Patients with Severe Pulmonary Arterial Hypertension

Jay Gajera[1], Atsuo Doi[2], Daniel Niewodowski[1], Shivanand Gangahanumaiah[2], Greg Snell[2], David Kaye[1], Tim Joseph[1], and David McGiffin[2]

[1]Alfred Health
[2]Alfred Hospital

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Abstract

Patients with pulmonary arterial hypertension (PAH) may develop large pulmonary artery aneurysms (PAA) which may be complicated by rupture, dissection or intravascular thrombus formation. These patients were traditionally considered for heart-lung transplantation but more recently, there have been reports of successful lung transplantation with reconstruction of the pulmonary artery (PA). We present two patients who underwent successful transplantation for PAH with giant PAA. One patient had end stage PAH and right pulmonary artery atresia complicated by a giant main PAA. This patient underwent bilateral sequential lung transplantation with concurrent pulmonary artery reconstruction. Another patient had end stage PAH with giant PAA on a background of D-transposition of the great arteries who had a Mustard repair at 9 months of age. This patient underwent heart-lung transplantation. Both heart-lung transplantation and lung transplantation with reconstruction of the pulmonary should be considered as a treatment option for patients with PAH with PAA.

1 INTRODUCTION

Patients with pulmonary arterial hypertension (PAH) may develop large pulmonary artery aneurysms (PAA) which may be complicated by rupture, dissection or intravascular thrombus formation. These patients were traditionally considered for heart-lung transplantation but more recently, there have been reports of successful lung transplantation with reconstruction of the pulmonary artery (PA).

We present two patients who underwent successful transplantation for PAH with giant PAA using two different strategies. Furthermore, we review previous literature and describe the treatment strategy for PAH with PAA.

2 CASE REPORT

2.1 Case 1

A 41-year old man with end stage severe PAH and right pulmonary artery atresia and giant main PAA (8 x 8 x 9cm) underwent bilateral sequential lung transplantation (figure 1). Concurrent (PA) reconstruction was planned as described in the literature.[1-5] His right lung derived its blood supply from collaterals arising from the bronchial arteries and mediastinal vessels. The entire main PA as well as the right and partly left PA were reconstructed using donor descending aorta and a bovine pericardium tube. The donor right PA did not have enough length to reach the reconstructed proximal right PA, and therefore the right PA was extended using a bovine pericardial tube (figure 2). This bovine pericardial tube was brought behind the superior vena cava and ascending aorta (figure 3).

Post operatively the PA pressures remained elevated and CT Pulmonary Angiogram (CTPA) demonstrated a mechanical cause due to kinking of the anastomosis between the bovine pericardial tube and the reconstructed

proximal right PA (figure 4). At reoperation, a posterior shelf had formed due to redundancy in the length of donor aorta and this was excised and reanastomosed. The remainder of his post-operative course was uneventful, and he was discharged home day 33. He is progressing well, without activity limitations seven months after transplantation.

2.2 Case 2

A 42-year-old man with end stage pulmonary hypertension and giant PAA (9 x 10 x 10cm) underwent heart and lung transplantation (figure 5). He had a background history of D-transposition of the great arteries post Mustard repair performed at age 9 months, with a baffle leak and left to right shunt. Heart-lung transplantation was performed through a clamshell incision as described previously [6-8]. The donor hila were positioned anterior to the phrenic nerves. [7, 8] Due to significant coagulopathy, the chest was packed and then closed the next day. There were no major post-operative complications and he was discharged home day 37. The patient is doing well twenty months after transplantation.

3 DISCUSSION

The argument for heart-lung transplantation for patients with pulmonary arterial hypertension (with or without a giant PAA) is no longer compelling with the knowledge that even the most adversely remodeled right ventricle, such as those preoperatively requiring inotropic or VA ECMO support, will reverse remodel after lung transplantation alone [9]. Most centers try to avoid heart-lung transplantation when possible, due to the shortage of donor organs as well. In the most recent analysis of the International Society of Heart Lung Transplantation (ISHLT) registry the 1,3 and 5 year survival in heart-lung transplantation for PAH was 87.3%, 68.6 and 61.8 respectively. In comparison the 1,3 and 5 year survival for all lung transplant recipients for PAH transplanted between 2008 and 2015 were 80.7%, 63.4% and 55.2% respectively [10]. Consequently, the decision for heart-lung transplant in PAH is reserved for patients with severe left ventricular failure or those with anticipated difficulty in repairing complex congenital heart disease [11-13]. Case 2 required heart-lung transplantation due to the underlying complex congenital heart disease and severe PH with PAA.

In PAH patients the prostacyclin analogue intravenous epoprostenol has been found to significantly increase transplant free survival [10, 14, 15]. Intravenous epoprostenol is reserved for patients with WHO class IV symptoms, where it is used in combination with one or both of an endothelin receptor antagonist and a phospho-di-esterase 5 inhibitor [16, 17]. In addition to initiating epoprostenol therapy, early referral of patients with WHO class III or IV symptoms to a transplant center is appropriate given their higher rate of disease progression and mortality [16].

However, with PAA in the setting of PAH, an increase in size is independent of normalized pressures [18], and progression of dilation and chest pain are indications for urgent transplant due to the risk of dissection and rupture. The rare complication of PAA poses substantial problems for lung transplantation and traditionally, combined heart-lung transplantation has been recommended as the treatment of choice. Sole lung transplantation is being increasingly recognized as an alternative in experienced lung transplant centers. In a relatively large 12-year series, Schwarz et al. reported 7 of 127 patients with PAH presented with severe PAA in their 12 year series. Their institutional experience with a lung-only strategy, where donor PA trunk was procured along with the lungs, and the main PA was reconstructed along with the implantation of the right lung. The left lung was then implanted and the left PA was reconnected to the main PA. Their series demonstrates feasibility of this approach in this complex group of patients [19]. Shayan et al. reported a case where the donor pulmonary trunk was procured with the lungs [20]. They implanted the lung allograft bloc by positioning it posterior to the heart and anastomosing the trachea instead of separate bronchial anastomoses. Force et al. and Noda et al. described techniques where donor thoracic aorta was fashioned to replace the recipient PAA which we employed in Case 1. It is important to carefully assess the pulmonary arterial and right ventricular pressures following replacement of the pulmonary arteries, especially in cases where extensive reconstruction was required.

4 CONCLUSION

Patients with PAH and giant PAA may be challenging and potentially carry high surgical risk. These patients should be evaluated by an experienced heart and lung transplantation centers. Strategies can be variable depending upon underlying disease, etiology, center dependent expertise, and resources available. In those patients who are refractory to medical therapy, transplantation may be the only management option. Multidisciplinary consensus should be obtained to offer patients individualised treatment options. Replacement or reconstruction of the pulmonary trunk is a feasible option for those who do not require heart-lung transplantation.

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Nil

Disclosures

The authors of this manuscript have no conflicts of interest to disclose as described by the American Journal of Transplantation.

Figure Legends

Figure 1. Case 1 Preoperative CT - axial (top), coronal (bottom)

Figure 2. PA reconstruction using donor descending thoracic aorta for entire main PA and left PA reconstruction (top), completed reconstruction (bottom)

Figure 3. Case 1 Line Diagram

Figure 4. Case 1 Postoperaative CT Angiogram – 3D reconstruction. Yellow arrow indicates bovine pericardial tube as it passes posterior to the aorta. Red arrow indicates site of kink.

Figure 5. Case 2 Preoperative CT - axial (top), coronal (bottom)

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Figure 1. Case 1 Preoperative CT - axial (top), coronal (bottom)

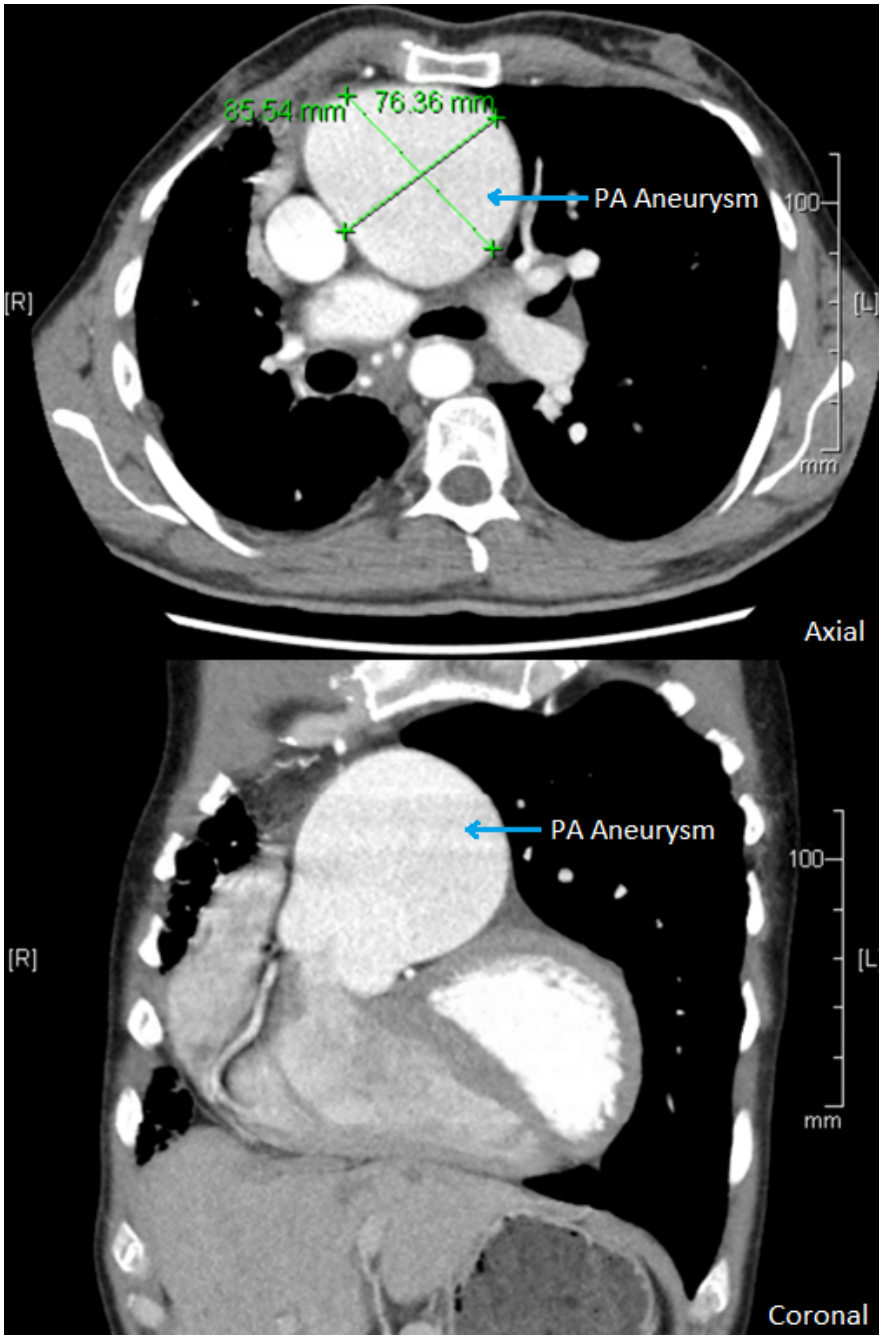


Figure 2 . PA reconstruction using donor descending thoracic aorta for entire main PA and left PA reconstruction (top), completed reconstruction (bottom)

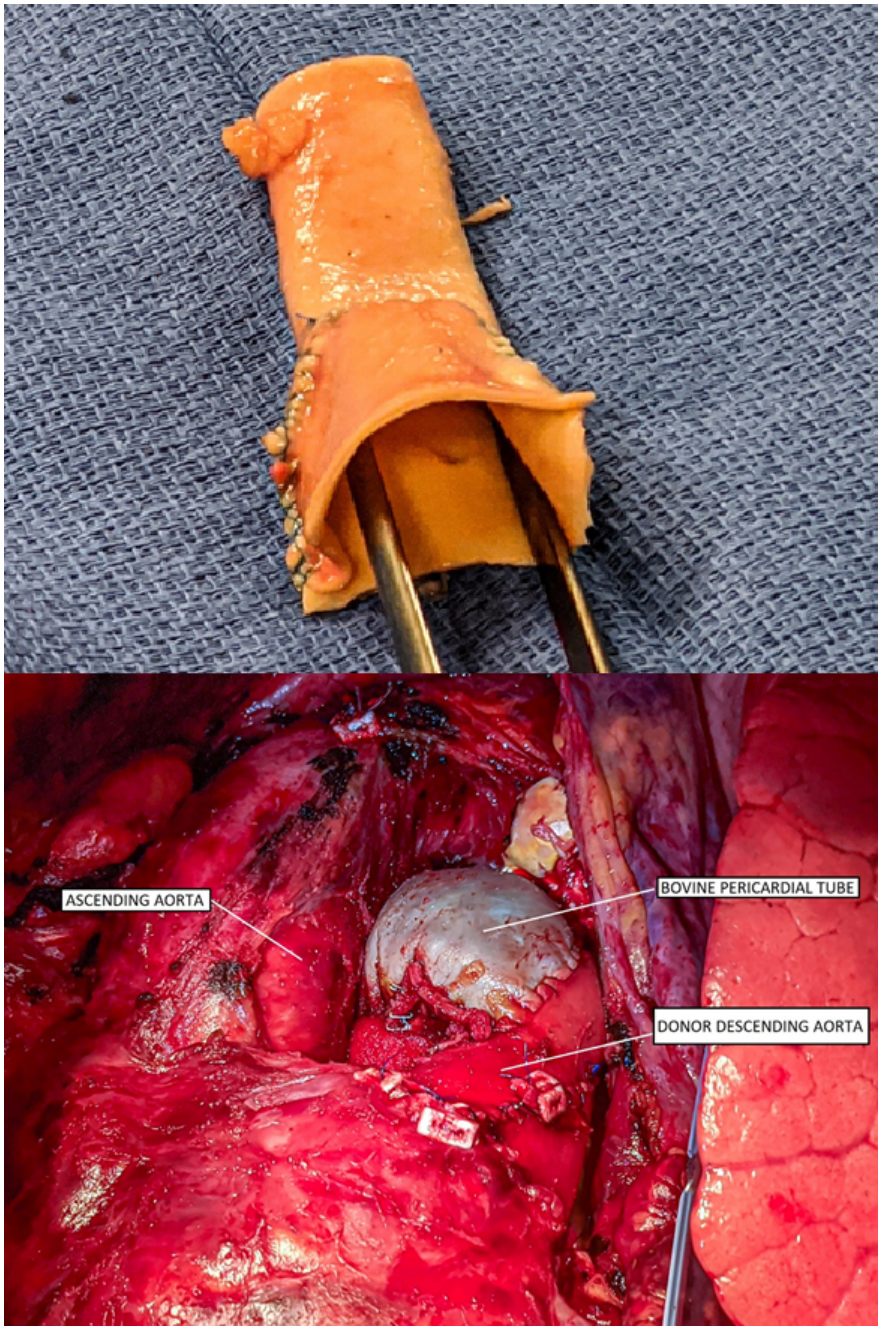
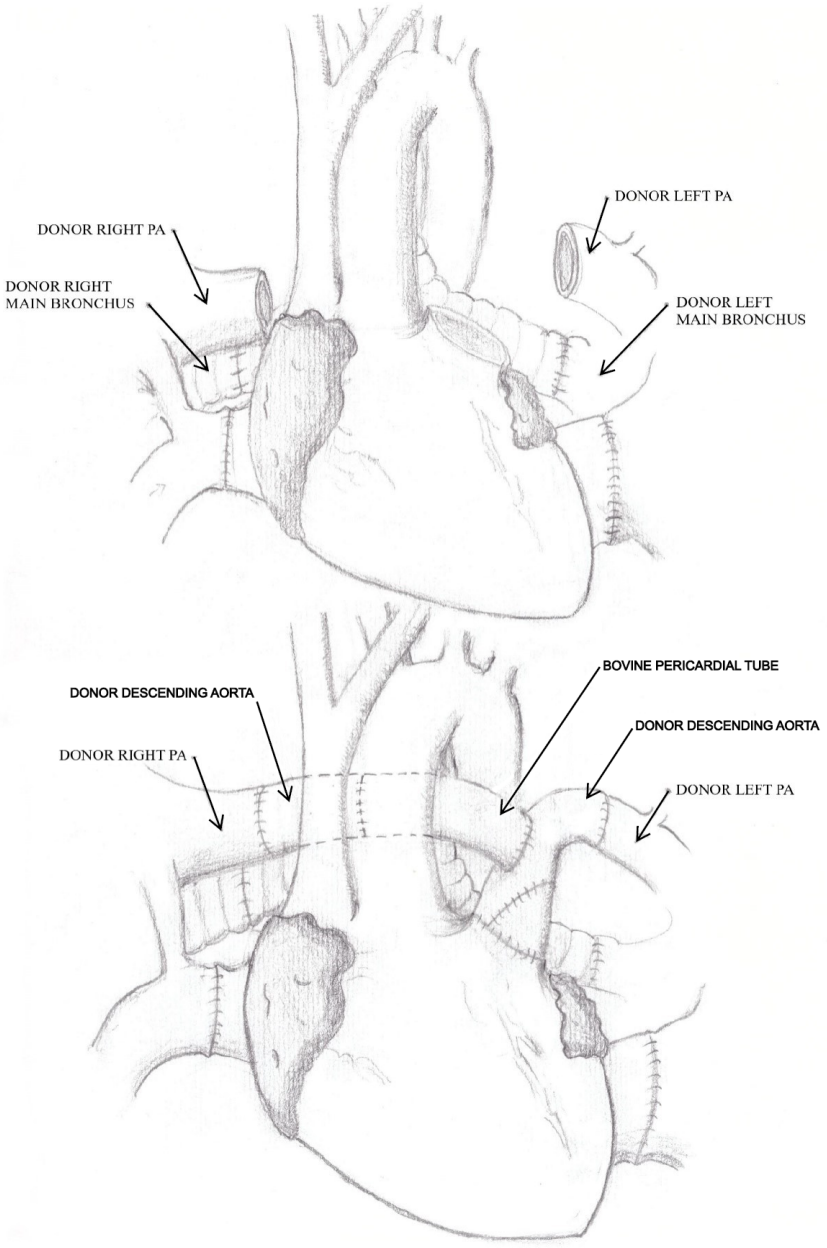


Figure 3. Case 1 Line Diagram



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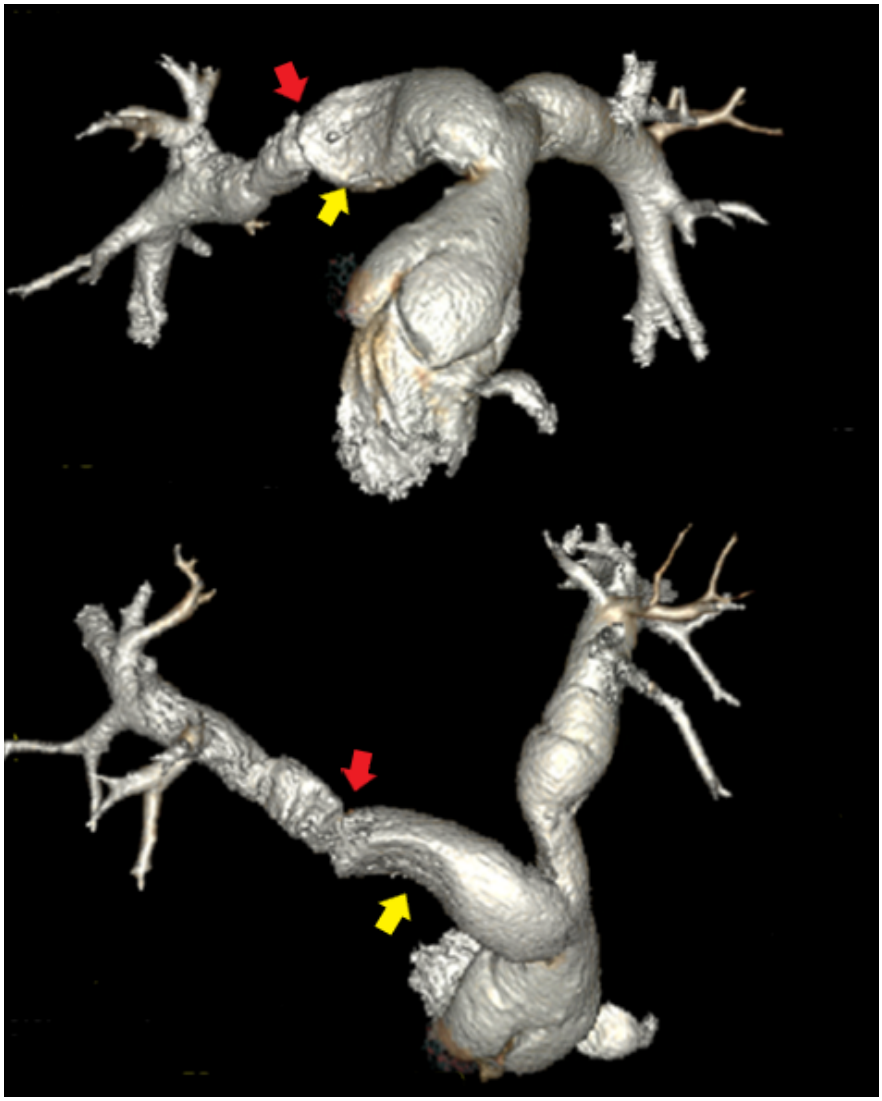

Figure 5. Case 2 Preoperative CT - axial (top), coronal (bottom)