Aortic complications following pediatric heart transplantation: A case series and review

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ABSTRACT

Aortic complications occur rarely after pediatric orthotopic heart transplantation, but are typically accompanied by catastrophic events. We describe the three cases of major aortic complications in our experience of 329 pediatric heart transplants. This case series and review highlight the important risk factors for aortic complications after heart transplantation.

Keywords: Aortic rupture, cardiac transplant, children, dissection

INTRODUCTION

Aortic complications following orthotopic heart transplantation (OHT) are rare in the adult population, ranging from 1% to 2%.[1] Rarer still are aortic complications in the pediatric OHT group.[2-5] Although infrequent these events are often catastrophic. We describe the three cases of major aortic complications in our experience of 329 pediatric heart transplants.

CASE REPORTS

Case 1

A nearly 4-year-old male with the diagnosis of heterotaxy with asplenia, unbalanced complete atrioventricular canal defect, total anomalous pulmonary venous return to the right atrium, and status post staged Fontan palliation was admitted to our institution secondary to worsening plastic bronchitis and congestive heart failure. After a 4-month hospital stay, the patient underwent OHT using bicaval anastomosis and patch pulmonary arterioplasty. He remained on antirejection medications, as well as amoxicillin for asplenia prophylaxis and clindamycin for the treatment of a superficial wound infection. Total parenteral nutrition was required before and after the OHT due to an inability to tolerate gastric tube feeds. On postoperative day 10, he complained of worsening nausea and belly pain. Drainage was noted from the sternotomy site, and a pericardial effusion was found on echocardiogram prompting fluid evacuation via a surgical subxiphoid incision. Cultures of the effusion grew *Candida tropicalis*, and appropriate antifungal medications were initiated. On postoperative day 13, the patient suffered a hypotensive cardiac arrest necessitating cardiopulmonary resuscitation. Due to active bleeding from the sternotomy site, he was taken emergently to the operating room. Upon opening the chest, the surgical field was inundated by pulsatile blood from a ruptured ascending aorta aneurysm. The bleeding was controlled manually, and the patient was placed on cardiopulmonary bypass. The ascending aorta was transected and replaced using a 20 mm Hemashield graft. Cultures obtained from the transected aorta grew *C. tropicalis*. Postoperatively, the patient remained hemodynamically unstable with persistent Candidal fungemia in spite of appropriate therapy. With continued multisystem organ failure, supportive care was ultimately withdrawn 2 months following the OHT.

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Case 2

A 23-year-old male with a history of hypoplastic left heart syndrome and staged Fontan palliation had undergone OHT at 13 years of age due to the progressive myocardial dysfunction. He had been noted previously to have dilation of the ascending aorta to 4.4 cm, and had complained of intermittent back and shoulder pain for several months. He suffered acute onset of chest pain and presented to an outside medical facility where a chest radiograph demonstrated a large, left-sided opacity. A computed tomography (CT) scan revealed a ruptured aortic arch pseudoaneurysm with a left-sided hemothorax [Figure 1]. He was rapidly transferred to our institution where he underwent urgent surgical repair. At surgery, a large aortic aneurysm was present on the lesser curvature of the transverse arch. The aneurysm had ruptured resulting in bleeding into the left pleural space. The aortic arch was completely resected and replaced with a 30 mm Hemashield graft to the descending aorta in an end-to-end fashion. The patient was discharged home without complications on postoperative day 9.

Case 3

A 14-year-old female with gene-positive Loeys-Dietz syndrome and significant phenotypic manifestations had previously undergone a valve-sparing aortic root reduction at 5 months of age, and subsequent aortic valvuloplasty at 12 years of age due to severe aortic insufficiency. Thereafter, she continued with moderate aortic stenosis and insufficiency resulting in progressive left ventricular dilation and dysfunction, ultimately necessitating OHT at 14 years of age. Prior to OHT, magnetic resonance angiography demonstrated an ascending aorta aneurysm measuring approximately 5.5 cm, with a second aneurysm in the isthmus region. In addition, significant tortuosity was present in the head and neck vessels. As a result of the aortic vasculopathy, the aortic arch was replaced with a branched Dacron aortic arch graft at the time of OHT. During the aortic arch surgery, the branch graft was sized to fit the tortuous neck vessels, dilated descending aorta, and the donor ascending aorta. In the immediate postoperative period, the patient was noted to have diminished pulses in the lower extremities. An urgent catheterization demonstrated an acute angulation of the distal aortic graft to descending aorta anastomosis believed to be the etiology of the pulse findings. As a result, the patient was taken back to the operating room where plication of the graft was performed. The patient convalesced and was discharged 3 weeks post-OHT after repeat catheterization demonstrated no evidence of aortic obstruction. Five days after discharge, the patient presented with the upper extremity weakness, ptosis, and asymmetric pupils. A CT angiogram demonstrated dissection of the descending aorta from the level of the aortic isthmus to the renal arteries with scattered intramural hematomas along the thoracic aorta [Figure 2]. The risk of surgical intervention was deemed so great as to preclude attempted surgery, and she was placed on a β-blockade infusion. Following rupture of the dissection, the patient developed a left-sided hemothorax and slowly deteriorated. The patient was placed on comfort care at the request of the family and expired. An autopsy revealed the dissection of the aorta from the suture line of the arch graft to the right renal artery. There was a large hematoma within the false lumen compressing the true lumen. In addition, a large blood clot was formed around the thoracic aorta. A perforation was detected at the right renal artery ostium, and a large amount of free blood was visualized in the abdomen.

DISCUSSION

Of the 329 pediatric OHTs performed at our center, there have only been three cases of major aortic complications (0.9%). Similar to the adult OHT population, our institution's experience shows that although rare, aortic complications are often fatal.

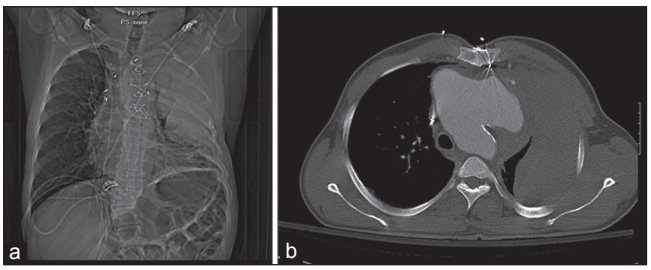

Figure 1: Computed tomography Case 2. (a) Anterior-posterior scout film demonstrating a large, left-sided opacity with tracheal deviation. (b) Axial projection demonstrating an aortic pseudoaneurysm with left-sided hemothorax

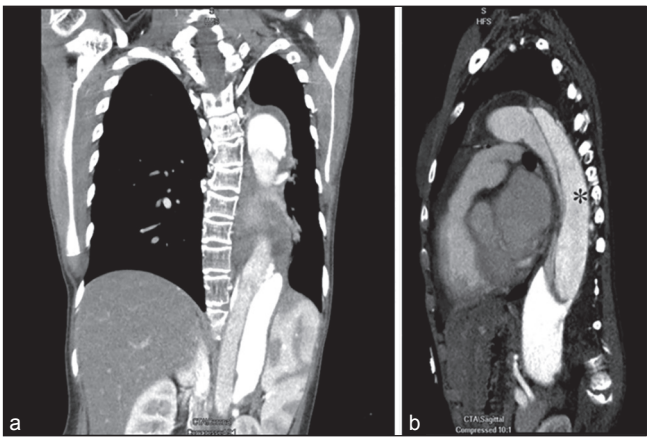

Figure 2: Computed tomographic angiography. Coronal (a) And sagittal (b) Reconstruction demonstrating a large aortic dissection extending from the aortic arch to the renal arteries. There is a large false lumen (*) compressing the true lumen of the aorta

The diagnosis of aortic dissection after OHT may be difficult to make due to cardiac denervation, which may result in dissection without pain. In addition, postoperatively, the mediastinal scarring may tamponade aortic rupture resulting in minimal hemorrhage.[6] Due to these difficulties, a healthy suspicion is required when encountering this patient population.

The etiology of aortic dissection after OHT may be acute, often related to the mismatch between the donor and recipient aorta. Infectious causes may also lead to dissection or rupture, as in our first case. The later phases of dissection are believed to be due to the degenerative processes augmented by immunosuppression drugs in combination with increased cardiac output.[6-8] For this reason, careful attention should be paid to aortic dimensions during the follow-up evaluations of patients after OHT.

In the pediatric cardiac transplant population, to our knowledge, there have only been five reported cases of major aortic complications.[2-5] Despite its rarity, our cases illustrate the possible additional risk factors to consider in pediatric OHT patients. These and other risk factors are listed in Table 1. Case 1 represents an infectious etiology for aortic rupture. Given the need for aggressive immunosuppression during the immediate posttransplant period, these patients are at high-risk for infectious complications. Studies have shown that OHT patients are at an increased risk of invasive fungal disease, with delayed sternal closure being an independent risk factor.[10] In critically ill patients, long-term antibiotics, central line usage, and total parenteral nutrition are also the risk factors for Candidal blood stream infections.[11] Those patients with heterotaxy, splenic dysfunction, and long-term antibiotic requirements, illustrated by Case 1, are likely at higher risk for invasive fungal infections.

The arterial wall is abnormal in the setting of congenital heart disease, as evidenced by significant medial necrosis.[12,13] The patients with a history of Norwood aortic arch reconstruction, such as Case 2, account for a significant proportion of pediatric OHTs. In patients with hypoplastic left heart syndrome, the neoaortic arch is known to dilate over the time. The etiology is believed to be related to compromised blood flow to the vasa vasorum.[13,14] Kanter *et al*. described the two cases of aortic complications in 234 pediatric OHT patients, both of which had undergone Norwood arch reconstructions. One of the complications was believed to be infectious, with a mycotic aneurysm from *Streptococcus viridans*. The second patient had the two separate pseudoaneurysms discovered 7 years apart at the times of re-transplantation. These pseudoaneurysms were felt to be secondary to intrinsic weakness in the reconstructed Norwood ascending aorta.[2]

Our case of aortic dissection (Case 3) demonstrates the risk of complications in patients with genetically abnormal vasculature. Kesler *et al*. surveyed centers with the experience in performing OHT in patients with Marfan syndrome. Those authors reported the dissection in 4 of 11 patients. This small shared experience on the risk of OHT in patients with Marfan syndrome appears to justify some centers' reluctance to perform OHT in populations with abnormal vasculature.[9] The patients with Loeys-Dietz syndrome have a higher risk of aortic dissection than those with Marfan syndrome, with histopathology demonstrating more diffuse medial degeneration of the aortic root tissue.[15,16] Subjecting these patients with an abnormal vascular tissue to long-term immunosuppression (with the side effects of hypertension and accelerated arteriosclerosis) may subject them to a much higher risk of major aortic complications over the long-term. This may be the reason that, to our knowledge, few patients with Loeys-Dietz syndrome have been reported to have received an OHT. As a result of this case, it has become our practice to perform tomographic aortic imaging prior to discharge after OHT in patients with known genetically mediated aortopathy.

Table 1: Etiologies of aortic complications in pediatric orthotopic heart transplant

Etiologies	Description
Acute phase	Mismatch between the donor and recipient aorta resulting in acute rupture[1,7]
Infectious	Mycotic pseudoaneurysm[2,3]
Traumatic	Secondary to rapid deceleration-type injury[4]
Genetic syndromes	Syndromes with predisposition to aortic aneurysms and dissection (i.e., Marfan and Loeys-Dietz)[9]
Previous aortic arch reconstruction	Secondary to intrinsic weakness in the reconstructed ascending aorta[2]
Late phase	True dissection secondary to high rates of arterial hypertension, diabetes mellitus, and accelerated arteriosclerosis[6-8]

CONCLUSION

The present three cases of aortic complications following pediatric OHT demonstrate the potential catastrophic nature of these events. We recommend careful assessment of the aortic dimensions in pediatric patients after OHT, including tomographic imaging when echocardiographic images are insufficient to demonstrate the anatomy clearly. A multi-institutional study may be required to determine the true prevalence of aortic pathology in these patients, and guide the long-term follow-up and therapy.

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Conflicts of interest

There are no conflicts of interest.

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