Technical challenges of lung transplantation in children after arterial switch operation

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Abstract

Pulmonary arterial hypertension after arterial switch operation for transposition of the great arteries is rare. Lung transplantation can be the last option in cases of failed medical therapy. We report 2 paediatric patients who underwent lung transplantation for this indication. Altered hilar anatomy, mediastinal adhesions and haemostatic control represent the main technical challenges. Volume-reduction surgery is sometimes necessary to address altered cardiopulmonary relationships while expanding the donor pool.

Keywords: Lung transplantation • Arterial switch • Pulmonary hypertension • Paediatric

INTRODUCTION

The arterial switch operation (ASO) is the preferred technique for surgical treatment of transposition of the great arteries [1]. Outcomes significantly improved over the last decades [2]. There is a low incidence (approximately 0.6–1%) of pulmonary artery hypertension (PAH) after ASO, which carries a poor prognosis [3]. Bilateral lung transplantation can be the last resort option [4].

CASE REPORT

Patient A

PAH was diagnosed 12 years after uneventful ASO. Because of increasing symptoms and failure to respond to medical treatment (peak VO2 of 13.7 ml/kg/min, normal range 36.5–52 ml/kg/min), the patient underwent bilateral lung transplantation at 14 years, and the weight of the patient was 35 kg and height 151 cm.

Using a clamshell incision, cardiopulmonary bypass (CPB) was commenced through central cannulation, and after extended dissection, both lungs were excised. Despite altered hilar anatomy and complex adhesions, the donor lungs were implanted in a standard fashion. This patient needed two re-explorations for bleeding. She was discharged after 54 days and is currently in New York heart association (NYHA) Class II 5 years post-transplant.

Patient B

This patient was diagnosed with PAH 4 years after ASO. Even under maximal medical therapy, the patient’s condition deteriorated to NYHA functional Class IV needing milrinone. Other surgical options (heart–lung transplantation or Potts shunt) were considered but rejected. The former due to acute deterioration without the necessary time to await size-matched heart and lungs, and the latter being technically more difficult with risk of massive bleeding without CPB backup. Finally, bilateral lung transplantation was performed. Weight of the patient was 20 kg and height 112 cm.

CPB was initiated through neck cannulation due to proximity of a severely dilated right ventricle to the back of the sternum, before the clamshell incision. Severe adhesions, especially perihi- lar, made extensive mobilization of the atrial cuff difficult. Bilateral donor lobectomy was performed to fit the smaller recipient. On the left side, the lower lobe was implanted, and on the right side, the upper and middle lobes were implanted. There was re-exploration for bleeding associated with unilateral white-out on a chest X-ray (CXR). The immediate postoperative course was further complicated by impaired oxygenation that led to cardiac arrest, needing veno-arterial extracorporeal membrane oxygenation and weaned after 9 days. The CXR continued to show the left-sided white out. Computed tomography (CT) demonstrated an upward kinking of the left main bronchial anastomosis, presumably due to the enlarged heart pushing the left lower lobe upwards. This was treated with endoluminal stenting (Fig. 1).
Figure 1: (A) A chest X-ray revealing complete atelectasis on Day 1. (B) Bronchoscopy showing the collapsed left main bronchus (arrow). The asterisk indicates the carina. (C) A chest X-ray post-stenting of the left main bronchus (arrow indicates the stent). (D) Coronal computed tomography reconstruction showing atelectasis recurrence despite the correct stent position.

Figure 2: Sagittal computed tomography reconstruction comparing the left hilar anatomy of a control patient (A, cystic fibrosis patient) with Patient (B, Lung transplant after ASO). LMB: left main bronchus; LPA: left pulmonary artery; LPV: left pulmonary vein.
Nevertheless, there was atelectasis recurrence despite repeated bronchoscopies with extensive lavages. In addition, head CT showed diffuse brain oedema and infarction, finally leading to a palliative care pathway. He expired 48 days after transplantation.

COMMENT

PAH after arterial switch operation is uncommon. Over the last 25 years, 25 PAH patients after ASO were identified through a recent multicentric study [3].

Normal hilar anatomy on the right comprises a main bronchus cranial to the pulmonary artery (PA) (epiarterial) and on the left side, a more caudal main bronchus in relation to the PA (hypoarterial). The manoeuvre reported by Lecompte et al. [5] altered the positional arrangements of the PA against bronchial anatomy. Figure 2 highlights the altered hilar anatomy and illustrates the more juxta-arterial position of the left main bronchus.

The heart is often enlarged. This should be taken into account when sizing the desirable volume of the donor lung, especially on the left. Lung-volume reduction can be used to accommodate the enlarged heart. It also provides an opportunity to enlarge the donor pool.

Multiple collaterals, as a result of underlying pathophysiology, can prolong the pulmonary explantation period and cause haemostasis problems.

This can lead to surgical re-exploration despite aggressive correction of coagulopathy as described in both of our patients.

The combination of PAH physiology and anatomical difficulties after previous extensive cardiac surgery represent the unique and challenging substrate in this particular group of patients, relying on transplantation as a possible last resort option, carrying high risk for severe complications.

Conflict of interest: none declared.

REFERENCES