Fontan completion after external stenting of the left bronchus and intrapulmonary artery septation for left pulmonary artery hypoplasia

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Abstract

Tracheobronchial obstruction and unilateral hypoplasia of the pulmonary artery are serious barriers to successful Fontan completion. We describe a 5-year old girl with left bronchial obstruction, hypoplasia of the left pulmonary artery and a single ventricle (double inlet left ventricle). She had undergone external stenting to treat left bronchial stenosis at 3 years of age, bidirectional cavopulmonary anastomosis and Damus–Kaye–Stansel anastomosis at 4 years of age, intrapulmonary artery septation 1 year later and, finally, completion of the Fontan operation with excellent postoperative haemodynamics.

Keywords: External bronchial stenting • Fontan procedure

INTRODUCTION

Tracheal or bronchial stenosis and unilateral hypoplasia of the pulmonary artery are critical issues in Fontan candidates that remain difficult to manage. Several procedures have been described to treat these conditions with variable results. The reported outcomes of external stenting for tracheobronchial obstructions have been good. Sakamoto et al. reported that intrapulmonary artery septation promoted symmetrical growth of the pulmonary artery in a patient with an unbalanced pulmonary arterial tree. We describe a patient with a single ventricle, left bronchial obstruction and hypoplasia of the left pulmonary artery. She underwent external stenting for the left bronchus, intrapulmonary artery septation for the left pulmonary artery and a final Fontan operation with excellent postoperative haemodynamics.

CASE REPORT

A female infant with severe cyanosis was referred to our hospital immediately after birth. She was diagnosed with a single ventricle (double inlet left ventricle), coarctation of the aorta and patent ductus arteriosus. She had undergone coarctation repair (subclavian flap aortoplasty) and pulmonary artery banding at the age of 9 days. Cardiac catheterization and computed tomography (CT) revealed left bronchial stenosis due to external compression caused by the descending aorta and high vascular resistance of the left pulmonary artery when she was 10 months old (Fig. 1A). Bronchoscopy revealed almost complete obstruction of the left bronchus and lung ventilation scintigraphy showed a right-to-left ratio of 0.096 (Fig. 2A and B). At the age of 1 year, she underwent aortoectomy and external stenting of the left bronchus to relieve the left bronchial stenosis. At that time, the bronchial wall was suspended within a 14-mm (diameter) ring-reinforced extended polytetrafluoroethylene (ePTFE) graft through a left thoracotomy. The graft was trimmed to a circumference of 12 mm and passed around the left main bronchus. The cartilaginous and membranous portions of the left bronchus were separately suspended within the ringed ePTFE grafts as described by Sakamoto et al. (Fig. 1D). Intraoperative bronchoscopy confirmed that the left bronchus was patent after external stenting. Since then, the left bronchial stenosis improved (Fig. 2C). She underwent bidirectional cavopulmonary anastomosis (BCPA) and Damus–Kaye–Stansel anastomosis at the age of 3 years. Although lung ventilation scintigraphy showed improved left ventilation (a right to left ratio of 1.0569), hypoplasia of the left pulmonary artery persisted (Figs 18 and 2D), and lung perfusion scintigraphy showed a right-to-left ratio of 1.0064. The diameter of the left pulmonary artery was 4.4 mm (41% of the normal value). To promote the growth of the left and central pulmonary artery for the next step in the Fontan completion, we created a systemic-to-pulmonary artery shunt (brachiocephalic artery to main pulmonary artery) with a 4-mm ePTFE graft and intrapulmonary artery septation with a 0.1-mm ePTFE membrane under cardiopulmonary bypass at the age of 4 years. This procedure increased the size of the left pulmonary artery on CT images. The diameter of the left pulmonary artery was 7.5 mm (67% of the normal value) and the pulmonary artery index was 229 (Fig. 1D). Cardiac catheterization showed superior vena caval and left pulmonary artery pressure of 10 and 12 mmHg, respectively. Finally, we created a total cavopulmonary connection in the extracardiac conduit fashion using an 18-mm ePTFE graft without fenestration when the patient was 5 years old. The postoperative course was uneventful and central venous pressure was 10 mmHg. Postoperative lung perfusion scintigraphy showed improved left lung perfusion (a right to left ratio of 1:0.404).
Figure 1: (A) CT showing the obstruction of the left bronchus (red arrows) compressed by the descending aorta, (B) CT showing external bronchial stent (red arrow) and hypoplastic left pulmonary artery, (C) CT showing systemic-pulmonary artery shunt (red arrow) and symmetrical size of bilateral pulmonary arteries and (D) illustration of external bronchial stent. The cartilaginous and membranous portions of the left bronchus were separately suspended within the ringed ePTFE grafts.

Figure 2: (A) Bronchoscopy showing obstruction of the left bronchus, (B) lung ventilation scintigraphy showing hypoventilation of the left lung, (C) bronchoscopy showing patency of the left bronchus and (D) lung ventilation scintigraphy showing the improvement of the left lung ventilation.
DISCUSSION

Tracheobronchial stenosis due to congenital broncho/tracheomalacia or secondary to external vascular compression has been reported. Left bronchial stenosis or occlusion frequently arises in patients with anomaly of the aortic arch [1, 2]. Surgical intervention for the left bronchial stenosis is needed under these circumstances. Furthermore, airway obstruction is critical in candidates for the Fontan procedure because a unique feature of this procedure is lung-dependent circulation.

Several manoeuvres have been established to address airway obstruction caused by external compression in children. Aortopexy and external stenting of the airway [1, 3] are popular approaches to treat these complicated lesions in children. We combined these procedures for our patient before BCPA and post-operative CT revealed excellent results. This procedure is thought to have growth potential because the external stent consists of two separate grafts, compared with the technique described by Hagl et al. [2].

Identical symmetrical growth of the bilateral pulmonary artery trees is thought to be necessary for Fontan completion. Although the mid-term outcome of a one-lung Fontan operation has been reported, the long-term outcome remains unclear [4]. Several procedures have been applied to address bilateral pulmonary arteries of different sizes. Intrapulmonary artery septation consists of BCPA to the side of the pulmonary artery with better growth, a systemic-pulmonary artery shunt and a septation patch inside the central pulmonary artery according to Sakamoto et al. [5]. This procedure promoted the growth of the left pulmonary artery in our patient. However, careful follow-up is considered necessary because the artificially created non-confluent nature of the pulmonary artery might cause asymmetrical vascular resistance in the pulmonary vasculature.

CONCLUSIONS

External stenting of the bronchus was safe and useful for this patient with bronchial stenosis complicated with single ventricular physiology. Intrapulmonary artery septation was also effective for treating unilateral hypoplasia of the pulmonary artery.

Conflict of interest: none declared.

REFERENCES


eComment. On-pump fontan versus off-pump fontan in patients with critical pulmonary status

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We read with great interest the manuscript by Shiraishi et al. [1], in which the authors successfully finalized surgical treatment in a patient with single ventricle physiology and critical pulmonary status. The manuscript lacks detailed information about the surgical procedure. Fontan procedure is the end-stage palliative option for univentricular hearts in the current era. Pulmonary properties are vitally important in this particular patient group. Operations may be performed with or without cardiopulmonary bypass and cross-clamping. It has been shown that extracardiac Fontan without cardiopulmonary bypass and cross-clamping have various advantages on ventricular and pulmonary functions. These include decreased incidence of arrhythmia attenuated usage of inotropic agents and blood/blood products, lesser activation of inflammatory cascades, decreased pleural effusion and earlier extubation [2-4]. Although adhesions after several cardiac procedures in the mediastinum of the patient presented by the authors may be expected and complicate further cardiac procedures, we believe off-pump Fontan completion might have been a good option for such patients with deteriorated pulmonary functions.

Conflict of interest: none declared.

References