

Case report

# Sutureless rescue of a severely hypoplastic pulmonary artery

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## Abstract

The management of severely hypoplastic pulmonary arteries in the adolescent or adult with congenital heart disease is challenging. A patient with pulmonary atresia and ventricular septal defect (PA-VSD) presented with severe cyanosis due to a kinked and thrombosed right ventricle to pulmonary artery conduit. Retrograde wedge angiography showed a patent right and a diminutive left pulmonary artery deep in the lung hilus. Treatment options were limited and therefore a different strategy had to be considered to rescue the lung segments. This case report describes the novel use of a covered stent to successfully obtain a sutureless connection between a vascular graft and a diminutive pulmonary artery during a hybrid procedure.

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**Keywords:** Stent; Shunt; Vascular graft; Pulmonary artery

## 1. Introduction

The management of severely hypoplastic pulmonary arteries in the adult with congenital heart disease is problematic. This is more than ever the case in pulmonary atresia and ventricular septal defect (PA-VSD) where every lung segment becomes important [1,2]. These patients often will have had several previous procedures, making surgery even more difficult. In this case report, we describe the use of a covered stent to obtain a sutureless connection between a vascular graft and diminutive pulmonary artery during a hybrid procedure.

## 2. Case report

A newborn female infant was diagnosed with PA-VSD and multiple aortopulmonary collateral vessels in association with a microdeletion of chromosome 22. At the age 3 years, a right ventricle to pulmonary artery conduit was inserted, which appeared to be kinked and subsequently thrombosed during early follow-up.

She presented at age 13 with severe cyanosis (oxygen saturation 65%). At this stage, her weight was 24 kg, height

138 cm with a body surface area of 0.99 m<sup>2</sup>. Retrograde wedge angiography showed a right pulmonary artery (PA) of 5 mm and a left PA of 2 mm in diameter. The right PA had an elongated hypoplastic and stenosed segment connecting to the right ventricular outflow tract. The left PA was patent from a region deep in the hilus and had branches to a considerable part of the lungs. We were faced with a dilemma – continuity had to be established, but the old conduit was calcified and thrombosed up to the level of hilus with a diminutive left PA. It would be extremely difficult to connect a graft to the hypoplastic thin-walled vessel without subsequent kinking; long-term patency could not be guaranteed. In addition, re-establishment of flow would, by default, lead to pulmonary oedema since flow had been minimal for years. Risks were discussed with the family and informed consent obtained. Since our strategy involved off-label use of devices, the local ethics committee was informed.

A branch to the right upper lobe and multiple aortico-pulmonary arteries were unifocalised on the right and connected to the distal end of an 8-mm stretch Gore-Tex<sup>®</sup> vascular graft (W.L. Gore & Associates, Inc., Flagstaff, AZ, USA). The calcified conduit was completely resected and the continuity with the right ventricle established by means of the 8-mm graft. A hybrid approach seemed the best option on the left side, considering the significant disparity in size between the desired graft and the left PA. The first step during surgery was to isolate and expose the proximal non-obiterated left PA by following a thin fibrotic string from

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the small main PA to the lung hilus. The vessel was subsequently cannulated and a 0.014 in. coronary guidewire was inserted. A 4 mm × 19 mm Jostent® Graftmaster coronary stent graft (Abbott Laboratories, Abbot Park, IL, USA) was positioned in the left PA with 60% of the stent inside the vessel and 40% outside. The balloon was inflated to 8 atm for expansion to nominal. This external area of the stent was covered with tissue glue and a 5-mm stretch Gore-Tex® vascular graft was slid over the stent. The combined graft–40% external stent unit was then dilated with a 6-mm Maverick® angioplasty balloon (Boston Scientific, Natick, MA, USA) to ensure tight approximation. One safety suture was placed on the graft–left PA contact area (Fig. 1). Intra-operative angiography was performed and the graft connected onto the 8-mm graft (Fig. 2).

The patient required extracorporeal membrane oxygenation (ECMO) postoperatively for 3 days. During the 11 months following the operation, the stent had to be redilated and cutting balloon dilatation was necessary to treat distal stenoses. Although her current saturation is 85%, further percutaneous procedures are anticipated to optimise flow to the left PA.

### 3. Discussion

With an increasing number of patients with complex congenital heart disease surviving beyond adolescence, clinicians are now faced with difficulties in management not previously encountered. Alternative techniques and hybrid procedures have to be considered and new strategies developed [3–7].

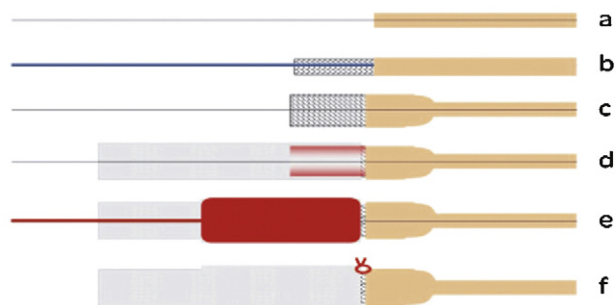
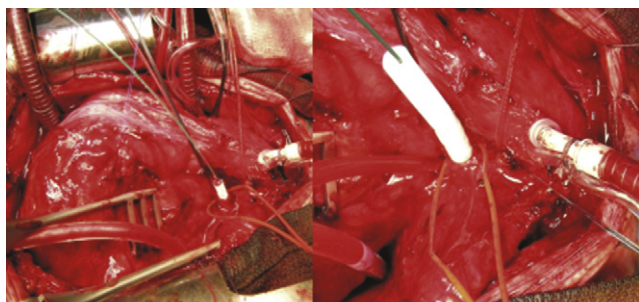


Fig. 1. Intra-operative photograph. Stent being inflated in distal left pulmonary artery (left). 5 mm Stretch Gore-Tex® vascular graft placed over covered stent after dilation – note guidewire still in place (middle). Sequential placement of stent and graft (right): (a) cannulation and 0.014 in. guidewire, (b) stent in hypoplastic pulmonary artery, (c) stent expanded to 4 mm, (d) 5 mm graft placed over stent after tissue glue, (e) stent and graft inflated with 6 mm balloon, (f) safety suture. See text for more details.

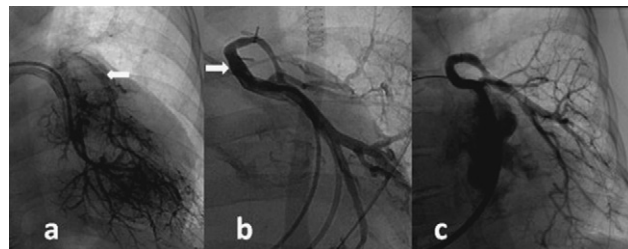


Fig. 2. Angiography. (a) Retrograde pulmonary wedge angiogram through 6F catheter (=2 mm) demonstrating diminutive left pulmonary artery (arrow); (b) Intra-operative angiogram showing graft (arrow) with stent (and distal left pulmonary artery extending to the left); (c) angiogram after 8 months shows improved pulmonary artery development after expansion of stent and rehabilitation of pulmonary artery.

In the patient described, several problems were anticipated during surgery. The anastomosis of a 5-mm graft to a thin-walled 2-mm native left PA deep in the lung hilus would technically be extremely difficult. Prolonged patency could hardly be guaranteed due to the risk of kinking. In addition, access for subsequent percutaneous interventions to rehabilitate the left PA flow was essential. The connection would require amplification as the distal pulmonary vessels would grow. The novel use of a covered stent implanted intra-operatively facilitated connecting the graft to a diminutive PA, thereby creating a sutureless distal connection (safety suture only on the outside). Kinking of the graft was avoided and by using a stretch vascular graft, the potential for further expansion of graft diameter and access to the distal LPA was made possible. Subsequent balloon angioplasty, using standard as well as cutting balloons, was possible during follow-up to further rehabilitate the left PA. Drawbacks of the technique include the theoretical risk of in-stent peel formation and limited future expansion capacity of the stent (off-label use, up to 6 mm).

On a technical note, it should be emphasised that even though the stent is implanted under direct vision, maintaining guidewire position is equally important to ensure controlled and accurate placement of the stent.

Not every patient is suitable for this technique. The procedure should ideally be performed in a hybrid catheterisation laboratory, since fluoroscopy and angiography are frequently necessary during the procedure. The availability of ECMO, as demonstrated in this case, is another prerequisite when re-establishing pulmonary perfusion after years of low-pressure and low-flow conditions.

In conclusion, a novel technique is described creating a successful sutureless anastomosis between a diminutive PA and a larger graft during a hybrid procedure. Correct stent positioning enables future transcatheter interventions to adjust PA flow according to growth.

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