Percutaneous management of complex acquired aortic coarctation in an adult with tetralogy of Fallot and pulmonary atresia

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Abstract

We present the case of a female adult with complex cyanotic congenital heart disease who had long-standing thoracic aortic obstruction due to scarring from earlier surgical procedures. She was symptomatic but felt to be too high risk for surgical intervention. With careful planning, she was able to undergo successful stenting of her aorta with subsequent clinical improvement. This case highlights some of the complexities of caring for adults with congenital heart disease and the importance of a thorough understanding of their anatomy and physiology and prior interventions before undertaking interventions.

Keywords: Adults, aortic repair, congenital heart disease, endovascular, pediatric intervention

INTRODUCTION

As the population of adults with complex congenital heart disease ages, more patients will present with residual postoperative defects that may require further intervention, often in the catheterization laboratory. It is critical to understand the pre- and post-operative anatomy as well as the previous surgical and catheter interventions to optimally treat these patients. We present an illustrative case.

CASE REPORT

The patient is a 28-year-old woman with tetralogy of Fallot, pulmonary artery atresia, and major aortopulmonary collateral arteries (MAPCAs) and a right aortic arch. She underwent attempted pulmonary artery unifocalization as a young child but did not tolerate this physiology and required urgent reoperation with unifocalization takedown and reanastomosis of a large aortopulmonary collateral to the mid thoracic descending aorta in the early postoperative period. When she was 9-year-old, she underwent surgical placement of a modified Blalock–Taussig shunt from the left subclavian artery to a hypoplastic pulmonary artery confluence for worsening cyanosis. Since that time, she has remained cyanotic and over the past 10 years developed progressive systemic hypertension in the upper extremities. On a recent clinic evaluation, she was noted to have severe hypoxemia (oxygen saturation 62% on room air), upper extremity hypertension (systolic blood pressure [SBP] 150 mmHg) with a 40 mmHg gradient to the lower extremities, and significantly decreased left ventricular systolic function (ejection fraction [EF] 30%). A cardiac computed

tomography (CT) scan revealed a 6 cm long calcified, narrowed segment in the descending thoracic aorta in the region of previous aortic surgical intervention (attempted collateral unifocalization and subsequent takedown) with the caliber decreased to 9 mm (proximal descending aortic diameter 18 mm). Importantly, a large MAPCA arose off the middle of this segment to supply the right lung [Figure 1a and b]. A surgical shunt was stenotic and supplied a markedly hypoplastic right branch pulmonary artery (<3 mm diameter) and there was no central left pulmonary artery, making complete repair unattainable at this point. She had poor activity tolerance and was in New York Heart Association (NYHA) Class III–IV heart failure. While her systemic hypertension likely provided some improvement in pulmonary artery flow, the distal stenoses clearly limited this effect. Further, it was felt that her systemic hypertension, combined with severe chronic cyanosis, was contrinuting to decrease left ventricular systolic function, and if the systemic and pulmonary artery obstructions could be relieved, she would have clinical improvement. Given her overall condition, she was not felt to be an appropriate surgical candidate, so percutaneous intervention was planned.

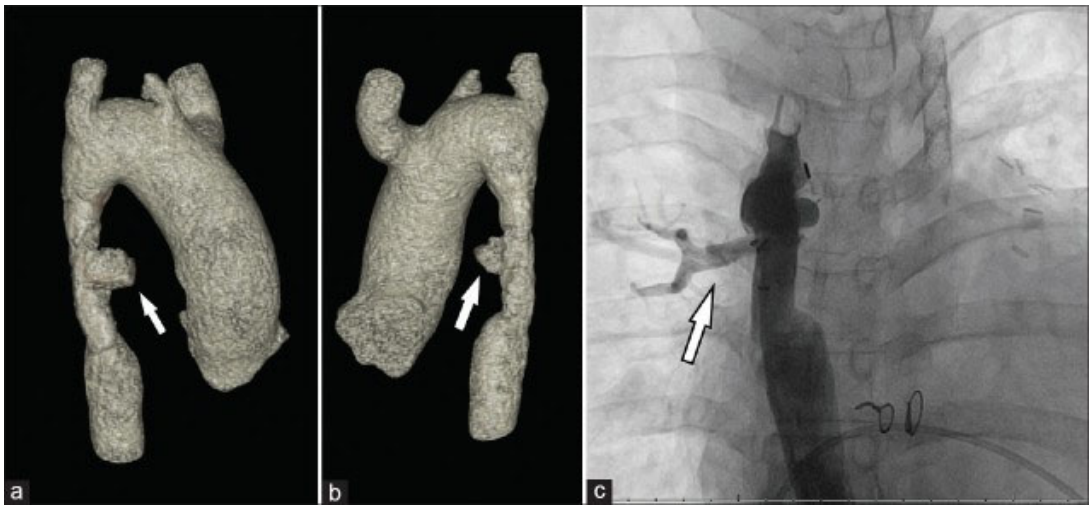

Figure 1

Three-dimensional reconstruction from cardiac computed tomography angiogram of the aorta from right anterior oblique (a) and posterior (b) views. There is long segment narrowing of the descending thoracic aorta with a large aortopulmonary collateral arising from the midportion of this segment and supplying a segment of the right lung (arrow). Angiography at catheterization (c) confirms the anatomy, with the aortopulmonary collateral supplying a segment of the right lung (arrow)

Under general anesthesia, hemodynamics revealed a 35 mmHg peak gradient from the transverse aortic arch to the abdominal aorta. Aortic angiography showed the anatomy as before and confirmed the MAPCA supplying the right lung arising off the midportion of the narrowed segment [Figure 1c]. Given the long narrowing and calcified aorta, stent therapy was felt to be the optimal intervention over angioplasty alone. However, because of the concern for obstructing any flow through the MAPCA and worsening her cyanosis, covered stents were not felt to be appropriate. Three IntraStent® Max™ LD stents (eV3, Inc., Plymouth, MN, USA) were deployed on 14 mm balloons in telescoped fashion. These were then postdilated with a 15 mm Z-Med II balloon (B. Braun Interventional Systems Inc., Bethlehem, PA, USA) and a 16 mm Atlas balloon (Bard Peripheral Vascular, Tempe, AZ, USA) with a marked improvement in vessel caliber and complete elimination of the gradient [Figure 2a]. The surgical shunt and hypoplastic right pulmonary artery were both stented with improved vessel caliber. The procedure was tolerated well and she recovered without incident.

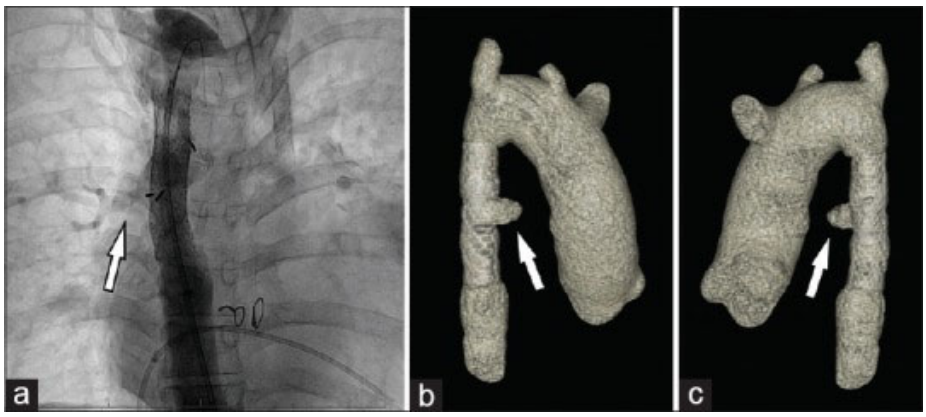

Figure 2

Immediately after stent implantation, an angiogram of the transverse aortic arch (a) demonstrates a marked improvement in thoracic aortic caliber and continued patency of the aortopulmonary collateral (arrow); note that the collateral does not fill as densely with contrast due to brisker antegrade aortic flow after intervention. Three-dimensional reconstruction from cardiac computed tomography angiogram of the aorta 1 year after intervention from right anterior oblique (b) and posterior (c) views shows persistent patency of the stented aortic segment and the aortopulmonary collateral supplying a segment of the right lung (arrow)

On 2-year follow-up, her oxygen saturation improved to 80%, she had only mild systemic hypertension (SBP 138 mmHg) with a 10 mmHg gradient to the lower extremities, left ventricular EF had improved to 60%, and her NYHA decreased to II–III. A cardiac CT showed continued patency of the aortic stents with no aneurysms and no obstruction of flow to the MAPCA [Figure 2b and c].

DISCUSSION

An understanding of the complex postoperative anatomy of adult survivors of congenital heart disease is critical to avoid compromising systemic and pulmonary blood flow during interventions. While primary therapy with covered stents would have minimized the risk for severe aortic disruption,[1,2] given the extensive calcification present, it would have compromised pulmonary blood supply in a patient with significant cyanosis.

With careful planning and preparation, complex aortic obstruction can before performed successfully in adults with complex congenital heart disease. An understanding of the sources of systemic and pulmonary flow is critical to ensure successful treatment.

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Conflicts of interest

There are no conflicts of interest.

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