

SHORT REPORT

Three Stage Hybrid Approach for Congenital Aortic Coarctation and Bicuspid Aortic Valve With Severe Aortic Stenosis in an Adult Patient

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Introduction: Untreated congenital aortic coarctation and valvular abnormalities are rare in an adult patient and require complex management.

Report: A 68 year old woman with congenital aortic coarctation and bicuspid aortic valve with severe aortic stenosis is described. She was treated by a two week, staged, hybrid approach: (i) aortic valvuloplasty, (ii) endovascular aortic repair and (iii) surgical aortic valve replacement with tricuspid ring annuloplasty and left atrial appendage ligation. Two and six month follow up blood pressure measurements demonstrated improved control. Six month imaging showed a patent aortic stent graft.

Discussion: A three stage, hybrid approach is a viable method for treating concomitant aortic coarctation and valvular heart disease in a symptomatic adult patient.

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INTRODUCTION

Congenital coarctation of the aorta (CoA) is a discrete constriction of the aortic lumen, typically occurring at the insertion site of the ductus arteriosus distal to the left subclavian artery. The pathophysiology of CoA is unclear, but it is reported to occur in 5%–8% of patients with congenital heart disease, including 70%–75% of patients with bicuspid aortic valve (BAV), while isolated CoA occurs in <0.05% of live births.¹ CoA is most often identified in infancy with reduced or absent femoral pulses. However, it may remain undiagnosed until adulthood when patients can present with symptoms from systemic hypertension, left sided heart failure, and/or hypoperfusion of the lower extremities.¹ Adult patients with concomitant intracardiac defects are rare and present complex challenges for management without standard guidelines.² An adult patient with congenital CoA and BAV with severe aortic stenosis and tricuspid valve disease is presented, who was managed with a hybrid, three stage approach within a three week time frame.

CASE REPORT

The patient, a 68 year old woman, was referred to vascular surgery for evaluation of congenital CoA. She reported

gradually worsening dyspnoea on exertion for the past two years with more acute deterioration in the past two months, now becoming short of breath after walking six feet. She also had dizziness and bilateral lower extremity oedema, with two hospitalisations over the past year for fluid volume overload. She had known about her congenital BAV and coarctation condition since her twenties, but reported that she has remained untreated until now due to insurance issues and the intervention risks described to her by another institution.

She had a past history significant for rate controlled atrial fibrillation (on warfarin), heart failure (New York Heart Association class 3), obesity (body mass index 30), left breast cancer in remission after radical mastectomy with lymph node dissection and chemotherapy complicated by left upper extremity lymphoedema, insulin dependent type 2 diabetes mellitus, hypertension, chronic kidney disease stage 3, and depression. She had a four pack year history of smoking (quit 1978) and worked as a cashier. She had a family history of heart disease in her father and several siblings, with no known genetic disease.

Physical examination was significant for an irregularly irregular heart rhythm and a 3/6 systolic murmur in right upper sternal border. Femoral and distal pulses were weakly palpable. There was left upper and bilateral lower extremity oedema, with a brachial blood pressure of 156/77 (lower extremity blood pressure was not obtained). Transthoracic echocardiogram showed mild to moderate left ventricular hypertrophy with an ejection fraction of 58%, severe tricuspid regurgitation, spontaneous echo contrast in left atrial appendage, and a mean transaortic gradient of

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43.7 mmHg, consistent with severe aortic stenosis. Computed tomography angiography (CTA) demonstrated severe coarctation of the descending thoracic aorta located 2.3 cm distal to the left subclavian artery, measuring 6.5 mm in diameter at the narrowest point (Fig. 1). Extensive collaterals were present (Fig. 2): both subclavian arteries measured ~ 20 mm at the origin, and prominent internal thoracic arteries to epigastric and intercostal arteries were visualised, including inferior rib notching on X-ray (Fig. 3). Pre-operative left heart catheterisation revealed no evidence of coronary artery disease.

For treatment a staged, hybrid approach was selected: in the first stage, the patient underwent percutaneous balloon aortic valvuloplasty in order to reduce the risks of general anaesthesia, with a peak to peak transaortic gradient decrease from 50 mmHg to 30 mmHg and a mean trans-aortic gradient of 35 mmHg. Overnight, she remained stable with a heart rate of 73–82 bpm and systolic blood pressure of 128–144 mmHg. In the second stage the following day, she underwent balloon angioplasty and thoracic endovascular aortic repair (TEVAR) of CoA via the left common femoral artery (7.9×9.0 mm diameter). The CoA was confirmed on digital subtraction angiography (Figs. 4 and 5), and a 24 mm \times 105 mm Cook Zenith Alpha device packaged in 16 Fr sheath was inserted. It was brought to the level of the coarctation with difficulty and deployed easily with intentional partial covering of the left subclavian artery. There was slight difficulty removing the delivery system due

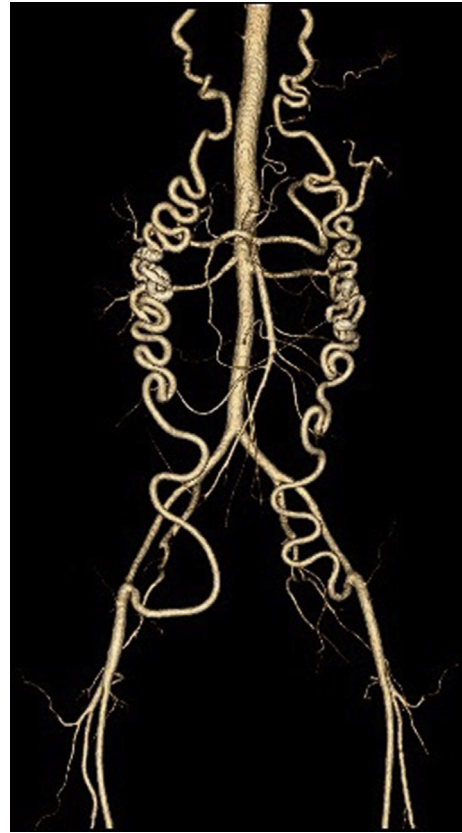


Figure 2. Computed tomography three dimensional reconstruction showing extensive collaterals through the internal thoracic arteries to epigastric and intercostal arteries.

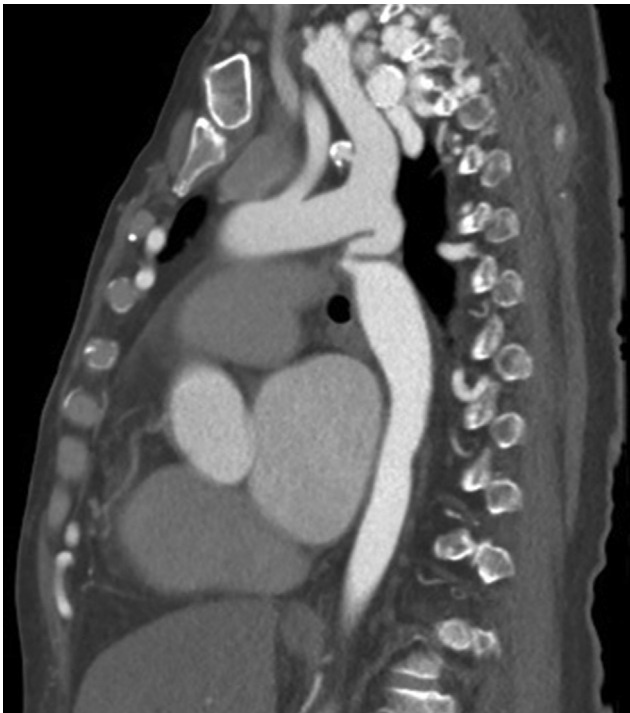


Figure 1. Computed tomography angiography demonstrating severe coarctation of the descending thoracic aorta located 2.3 cm distal to the left subclavian artery, measuring 6.5 mm in diameter at the narrowest point.

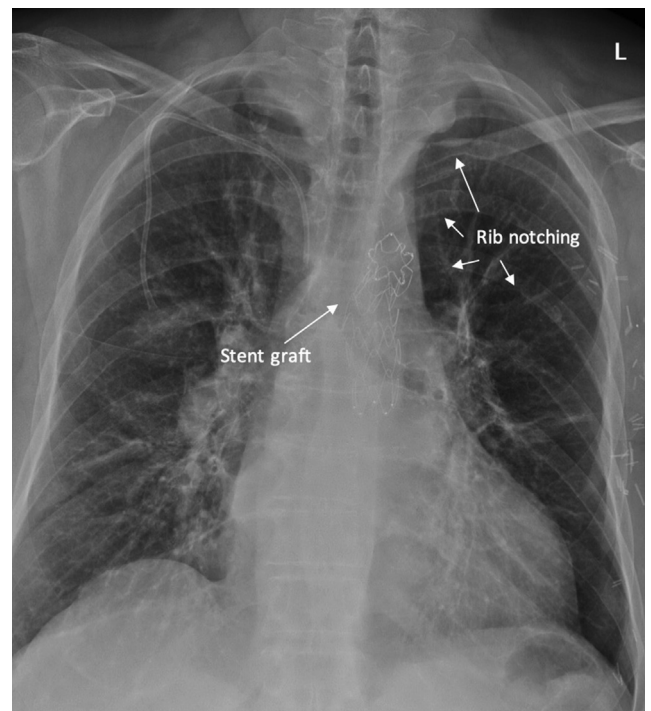


Figure 3. Chest X-ray demonstrating inferior rib notching consistent with collateral blood flow through the intercostal arteries from long standing aortic coarctation. This image was taken after aortic stent graft placement prior to aortic valve replacement.

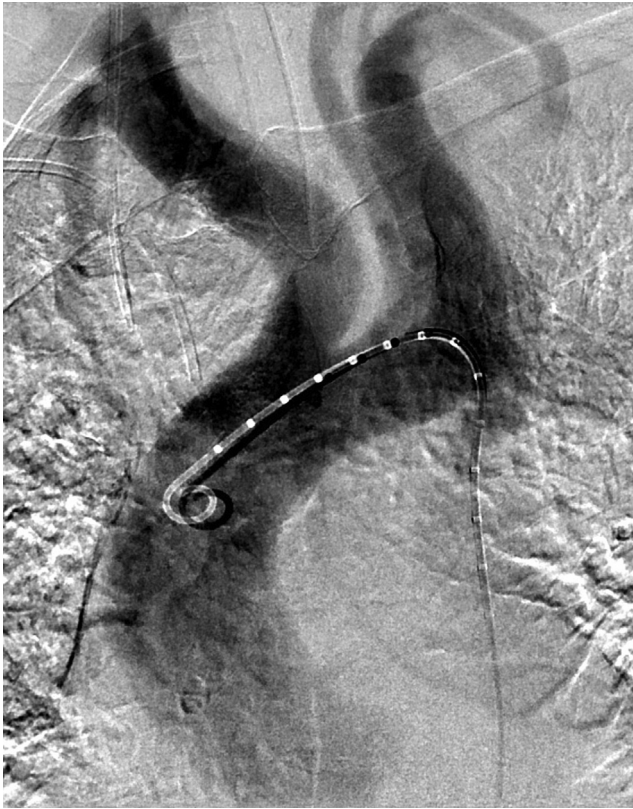


Figure 4. Digital subtraction angiography of the descending aorta, demonstrating filling defect (circle) attributed to severe aortic coarctation prior to stent graft placement.



Figure 5. Proximal aortogram demonstrating limited blood flow distal to coarctation (circle) with preferential flow to aortic arch vessels, measuring approximately 17.5 mm (right brachiocephalic), 11.0 mm (left internal carotid), and 20.0 mm (left subclavian).



Figure 6. Computed tomography three dimensional reconstruction of the aortic stent graft which was positioned with intentional partial coverage of the left subclavian artery.

to the coarctation, but this was achieved successfully without complication.

Serial balloon angioplasty was performed, starting with 10 mm up to 16 mm in 2 mm increments (Fig. 6). There was near elimination of the pressure gradient across the CoA as determined by radial and femoral arterial line monitoring. Her post-operative course was uncomplicated, and she was discharged the following day. She reported improved dyspnoea on exertion one week later. Two weeks post-TEVAR she underwent the third stage surgical aortic valve replacement (SAVR) with a 29 mm Trifecta prosthetic valve and tricuspid valve ring annuloplasty. Her brachial blood pressure at two and six month follow up was 126/70 and 134/67, respectively, and two month transthoracic echocardiogram demonstrated a mean aortic gradient of 2.4 mmHg. Creatinine remained stable post-operatively. She has reported renewed dyspnoea on exertion, attributed to chronic combined diastolic and systolic heart failure, and pulmonary hypertension. Her six month CTA showed a patent stent graft without evidence of compromised aortic flow.

DISCUSSION

Patients with CoA have been shown to have a reduced life expectancy, largely from cardiovascular sequelae, and the mortality of untreated CoA is up to 75% by 46 years of age.^{3,4} Therefore, early treatment is critical. The guidelines for CoA intervention are based on systemic hypertension, upper extremity/lower extremity blood pressure gradient, echocardiography Doppler gradient, and anatomical (imaging) evidence of CoA.⁵ Treatment options include open surgical or endovascular approaches, with the latter being the treatment of choice in adults. Stent graft placement is preferred to balloon angioplasty alone and is associated with similar short and mid term haemodynamic improvements and fewer acute complications than surgery.^{3,5,6} However, adult patients with concurrent untreated valvular lesions and CoA require complex management with no specific management guidelines. This case report highlights a hybrid management strategy for CoA and BAV with severe aortic stenosis and tricuspid regurgitation in a 68 year old woman. Six month follow up showed improvement in blood pressures with a patent stent graft and mean aortic gradients ≤ 3 mmHg.

CONFLICTS OF INTEREST

None.

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