

CASE REPORT

A soldier's return to duty after minimally invasive correction of complex congenital cardiovascular disease

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Abstract

We report a case of incidental detection of severe aortic coarctation, severe secundum atrial septal defect, and bicuspid aortic valve in an active-duty military service member. A single complex minimally invasive procedure successfully corrected his coarctation and atrial septal defect allowing this patient to continue military service.

KEYWORDS

aortic coarctation, atrial septal defect, bicuspid aortic valve

1 | INTRODUCTION

The incidence of congenital heart disease (CHD) is roughly 40,000 cases in the United States every year.¹ Aortic coarctation (CoA) occurs in 4 of every 10,000 births, making up roughly 5%–8% of all CHD.^{2,3} Of those with a bicuspid aortic valve (BAV), only 7% have a concomitant CoA, but 75% of patients with a CoA also have BAV.³ Atrial septal defects (ASD) occur in roughly 1 in every 1860 births in the United States.² Genetic abnormalities are common in patients with CHD, and genetic testing for concomitant syndromes may be reasonable in patients with adult CHD. While there appears to be a genetic link between CoA and BAV, there is no unifying etiology for the development of these conditions with a concomitant ASD.⁴

CoA presentation is largely dependent on the severity ranging from asymptomatic in most cases to headaches, epistaxis, exertional intolerance, dizziness, and lower extremity claudication.⁵ These symptoms are largely driven by increased afterload proximal to the narrowing and decrease blood flow distally leading to discrepant upper extremity hypertension, elevated upper to lower extremity pressure gradient, and weak or absent femoral pulses.⁵

Additionally, a prominent, nondisplaced apical impulse, loud A2, or continuous or systolic murmur radiating to the scapula due to significant collaterals may be appreciated on examination.⁵

Most ASDs identified during adulthood are asymptomatic and found incidentally. A minority of these patients will develop a gradual decline in exercise capacity and occasionally overt dyspnea on exertion particularly if pulmonary arterial hypertension is present.⁶ One of the ways ASDs are characterized is the degree of left-to-right shunting. Shunts that are hemodynamically significant enough to lead to physiologic sequelae should usually be closed. The significance of shunts is measured with a Qp:Qs, which is the ratio of pulmonary blood flow to systemic blood flow. If the Qp:Qs is greater than 1.5, then the hemodynamic impact of the shunt will be significant. Intracardiac shunts and the subsequent increase in intracardiac circulatory volume can lead to chamber dilation particularly of the atria. In some patients, this can contribute to atrial arrhythmias.⁶ On examination, patients with significant ASD will classically have a fixed split S2 due to increased pulmonary circulatory volume and the subsequent delayed closure of the pulmonic valve.⁶

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The presence of CoA, BAV with any complications, or ASD is disqualifying for any potential United States military recruit.⁷ For retaining current service members, the United States Army allows exceptions for CHD that can be repaired with resolution of long-term risk and complications.⁷

2 | CASE PRESENTATION

A 20-year-old fit active-duty male with a history of growth delay as a child and bilateral upper extremity hypertension. Despite these abnormalities, this service member completed all military entrance processing, medical evaluation, and predeployment screening without detection of any CHD. The patient did not disclose his history of growth delay until after his CHD was discovered. During an overseas deployment training exercise, he was rescued from the driver's seat of an M1A2 Abrams tank after suffering carbon monoxide poisoning from an exhaust system malfunction. He was intubated in the field and transferred to higher level of care where there was concern for myocardial infarction. After stabilization and extubation, he was medically evacuated to a military treatment facility in the United States where his troponin level was normal, and his ECG was normal. However, a computed tomography scan (CT) of his chest revealed narrowing of the proximal descending thoracic aorta, and a transthoracic echocardiogram (TTE) was notable for a bicuspid aortic valve. Thoracic magnetic resonance imaging, shown in [Figure 1](#), corroborated these findings and demonstrated right atrial enlargement concerning for atrial septal defect (ASD). Left and right heart catheterization showed severe CoA with a peak-to-peak gradient of 30 mmHg and extensive collateral artery formation. The catheterization also demonstrated a mildly elevated mean pulmonary artery pressure and a pulmonary-to-systemic blood flow ratio of 1.5, reflecting a severe left-to-right shunt. CT coronary angiography measured the ASD at 7 mm.

After multidisciplinary heart team discussion, the patient underwent successful percutaneous transcatheter aortic coarctation stenting and ASD closure during a single procedure.⁸ Immediate postprocedure coarctation stent gradient was 5 mmHg. Follow-up TTE demonstrated no residual interatrial shunt, and CT aortic angiography demonstrated patent endovascular stent without complication, shown in [Figure 2](#). The patient's upper extremity hypertension improved, and he was able to return to non-straining exercise without symptoms. 4 months following the procedure, the patient completed a Bruce protocol treadmill stress test running for 12:36 min and reaching 14.5 METS with normal ECG and hemodynamic response. Given his CHD, it was recommended that he refrain from

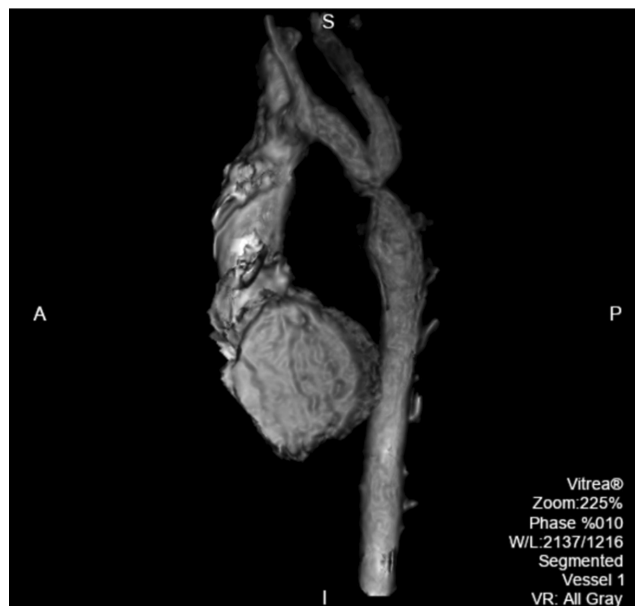


FIGURE 1 Aortic MRI demonstrating aortic coarctation with focal narrowing to 10 mm with severely increased collateral blood supply to the lower descending aorta, bicuspid aortic valve, and dilated right atrium concerning for atrial septal defect.

high-intensity competitive athletics with forceful body contact and exercises involving straining. After appropriate recovery, the patient was able to return to duty and met cardiovascular military retention criteria.

3 | DISCUSSION

We present a case of complex CHD that was corrected with a single minimally invasive percutaneous procedure that allowed this service member to be retained for duty in the United States Army. Although transcatheter correction of ASD and of CoA is each individually the standard of care for these conditions at centers for CHD, simultaneous transcatheter correction of multiple congenital issues is an important treatment approach for referring cardiologist to be aware of.

In addition to multidisciplinary team discussion, it is important to consider the patient's wishes when making management decisions for CHD. Prior to making any treatment decisions, the patient was counseled thoroughly on the implications that his condition and its management would have on his health and career. In cases where multiple treatment options are reasonable and timing of intervention is flexible, a patient should be well informed, and their autonomy must be preserved regardless of the social implications of their decision.

Our patient met current guidelines for the correction of his coarctation given his systemic hypertension, elevated peak-to-peak gradient, and severe

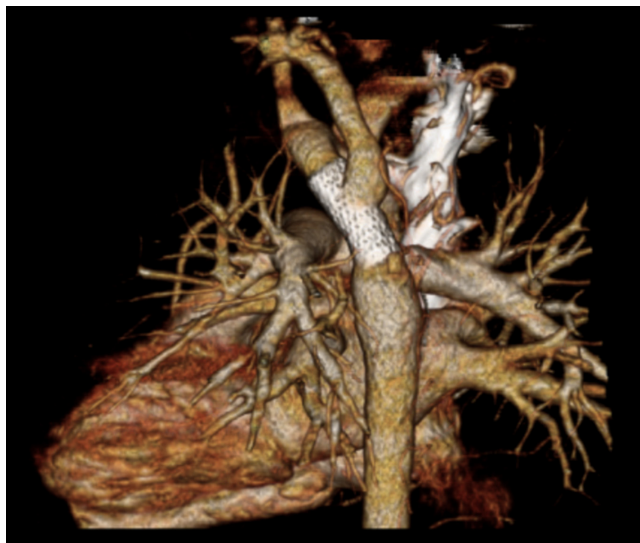


FIGURE 2 Cardiac CT angiogram 1 month following procedure demonstrating patent coarctation stent with intact flow to the subclavian artery and no evidence of aneurysm or other complication.

collateralization.⁸ The focal narrowing of the CoA was just distal to the subclavian artery making stenting technically challenging. Often, covered stents are used for CoA, but this case was not amenable due to concern for occlusion of the subclavian artery. In our patient, a 36-mm intrastent mega LD was used beginning proximal and extending distal to the subclavian artery correcting the focal narrowing. This temporarily reduced blood flow to the subclavian artery until a stent fenestration could be ballooned creating an intrastent ostium for the subclavian artery. This is depicted in the postprocedural follow-up CT aortic angiogram in [Figure 2](#) with patent endovascular stent. Postoperatively, this patient's upper extremity to lower extremity blood pressure discrepancy normalized, but he developed systemic hypertension, which is a common occurrence following CoA stenting that responded well to losartan.

Regarding his secundum ASD, there was evidence of left-to-right shunt on multiple image modalities. This patient had normal pulmonary vascular resistance and a pulmonary artery systolic pressure less than half of the system pressure. This is important because ASD closure is contraindicated in adults with PA systolic pressure greater than two-thirds systemic, pulmonary vascular resistance greater than two-thirds systemic, or a net right-to-left shunt.⁸ Qp:Qs measurement showed that there was a significantly higher pulmonary blood flow than systemic blood flow. This Qp:Qs ≥ 1.5 indicated that the shunt was sufficiently large to cause physiological sequelae. Additionally, there was already chamber dilation in the form of right atrial enlargement due to

increased volume load. When hemodynamically significant ASDs are identified, it is important to assess the patient's functional capacity and whether the ASD is symptomatic as there is more clear benefit for patients who are functionally impaired by their ASD.⁸ In this patient, a 10 mm Amplatzer Septal Occluder was placed with usual technique without complication. Serial follow-up echocardiograms demonstrated a well-seated interatrial device without residual shunt.

Bicuspid aortic valves are the most common congenital heart disease, and fusion of the left and right coronary cusps is the most common phenotype.^{3,9} At the time of evaluation, this patient had not developed valvular dysfunction. The development of aortic stenosis often occurs earlier than in tri-leaf patients and is suggested to be related to leaflet orientation.⁹ Aortic regurgitation is also a common finding in BAV patients and is independent of valve stenosis.⁹ At baseline, patients with BAV generally have larger aortic size, which predisposes the development of early root and ascending aortic dilation.⁹ Given these findings in conjunction with the patient's other congenital disease, it is paramount that annual lifelong cardiology evaluation is completed. The presence of a bicuspid aortic valve is an important consideration when determining the timing and method for treatment of these associated conditions. Additionally, it is recommended that all first-degree relatives be screened for BAV and aortopathy with echocardiography.⁸

This case highlights the patient-centered shared decision-making afforded to congenital heart disease patients by the contemporary multidisciplinary heart team. The single transcatheter procedure performed for the correction of this patient's aortic coarctation and atrial septal defect provided complete correction of all indicated congenital lesions while remarkably permitting his continued service in the United States Armed Forces.

AUTHOR CONTRIBUTIONS

Adam Kisling: Conceptualization; data curation; writing – review and editing. **Robert Gallagher:** Supervision; writing – review and editing.

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None.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors have no competing interests to declare that are relevant to the content of this article.

DATA AVAILABILITY STATEMENT

All data supporting this study's findings are available from the corresponding author upon reasonable request.

INFORMED CONSENT

Written informed consent in accordance with Walter Reed National Military Medical Center was obtained from the patient.

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