

Profound Aortopathy: A Rare Case of Massive Ascending and Descending Aortic Aneurysms, Type B Aortic Dissection, and Severe Aortic Valve Regurgitation in an Adult Patient With Uncorrected Congenital Aortic Coarctation

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INTRODUCTION

Aortic coarctation is one of the more common congenital cardiovascular abnormalities.¹ However, uncorrected aortic coarctation is rarely encountered in adults.² Furthermore, when present, aortic coarctation



Figure 1 Surface-rendered CT angiogram revealing massive ascending and descending TAAAs and the aortic coarctation just proximal to the descending TAA.

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Figure 2 ECG-gated thoracic aorta CT angiogram, sagittal view of the aortic root, ascending aorta, and descending TAAAs; also shown are the dissection flaps.



Figure 3 MRA, sagittal view revealing ascending and descending TAAAs as well as dissection flaps within descending TAA.

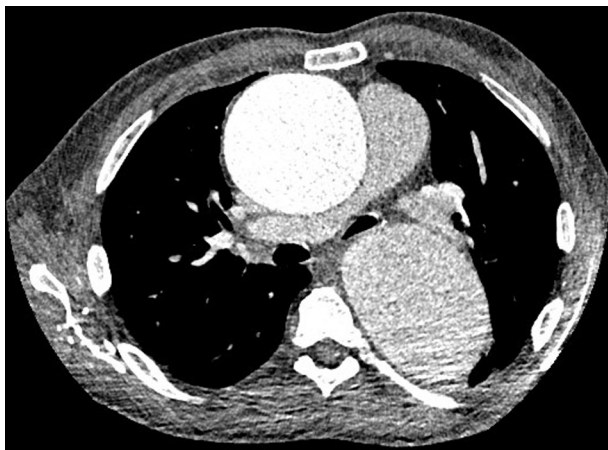


Figure 4 ECG-gated thoracic aorta CT angiogram, axial view of the ascending and descending TAA. Also shown is compression of the right pulmonary artery by the ascending TAA.

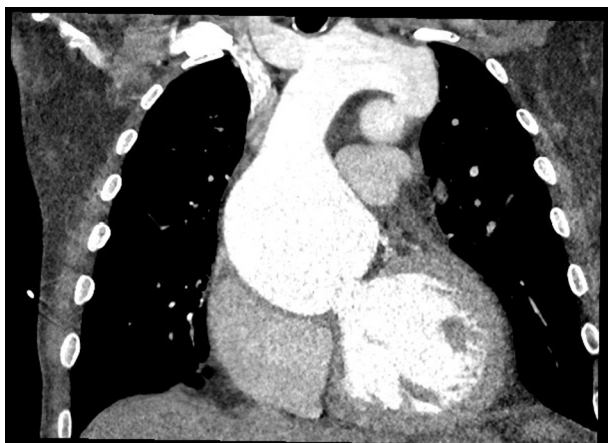


Figure 5 ECG-gated thoracic aorta CT angiogram, coronal view illustrating aortic coarctation just proximal to the descending TAA.



Figure 6 ECG-gated thoracic aorta CT angiogram, axial view illustrating descending TAA compressing the left pulmonary artery.



Figure 7 ECG-gated thoracic aorta CT angiogram, sagittal view illustrating descending TAA compressing the left pulmonary artery.

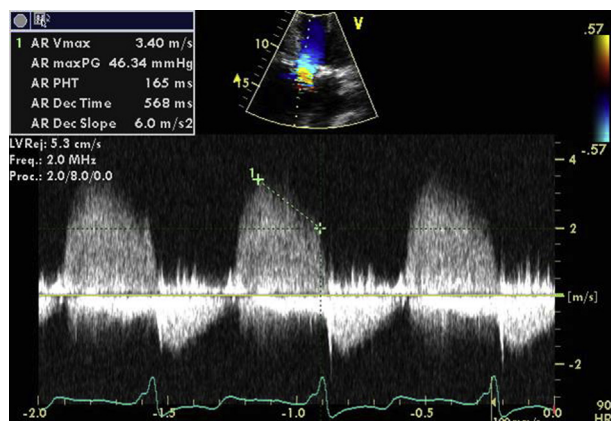


Figure 8 Continuous-wave Doppler revealing aortic valve regurgitant jet deceleration and very short pressure half-time consistent with severe aortic valve regurgitation. AR, Aortic regurgitation; Dec, deceleration; maxPG, maximum pressure gradient; PHT, pressure half time; Vmax, maximum velocity.

is most commonly associated with ascending aortic aneurysms.² Here we present a case of a rare combination of massive ascending and descending thoracic aortic aneurysms (TAAs), type B aortic dissection, and severe aortic valve regurgitation further complicated by severe left ventricular (LV) systolic dysfunction in a patient with an uncorrected aortic coarctation. To our knowledge, this is the first known report with this combination and severity of aortopathy.

CASE PRESENTATION

A 58-year-old male firefighter with a history of a ventricular septal defect that was repaired at age 6 presented with bilateral lower

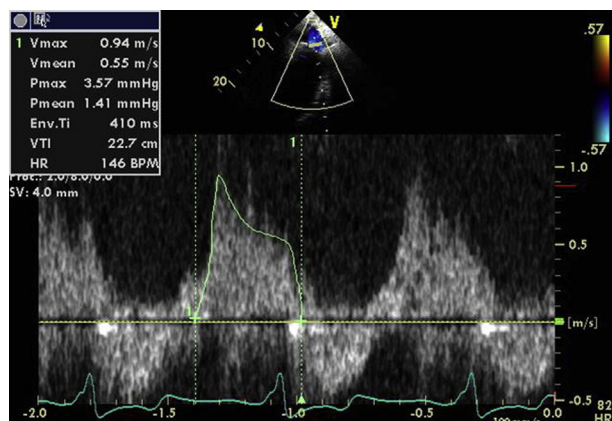


Figure 9 Pulse-wave Doppler at suprasternal notch view showing holodiastolic flow reversal consistent with severe aortic valve regurgitation.

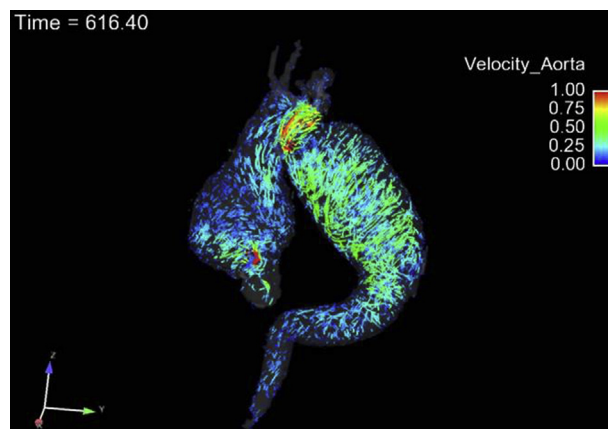


Figure 10 Cardiac MRI/MRA 4D flow analysis prior to surgery.

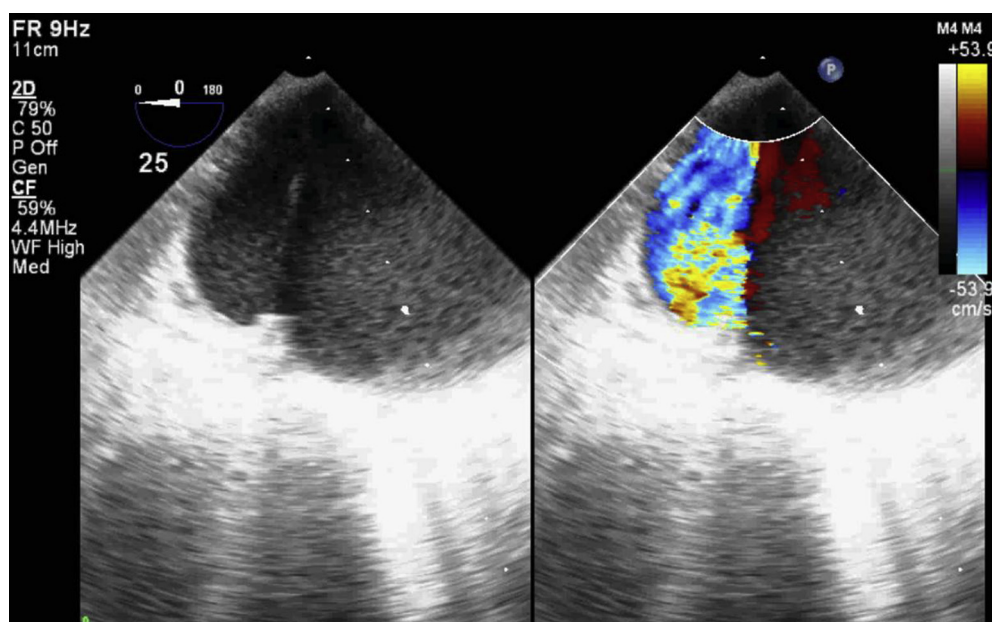


Figure 11 TEE revealing type B aortic dissection flap at 25 cm distal to the incisors.

extremity edema, orthopnea, and cough 1 month prior to hospital admission. He had no family history of congenital or acquired cardiovascular disease. Physical exam revealed blood pressure of 129/62 mm Hg, clear lungs, II/IV decrescendo holodiastolic murmur at the right upper sternal border, III/VI systolic murmur at the right upper sternal border, bounding radial and carotid artery pulses, DeMusset's sign (head bobbing with each heartbeat), and Muller's sign (rhythmic pulsation of the uvula). Chest x-ray showed a widened mediastinum and mild to moderate cardiomegaly. Electrocardiogram (ECG)-gated thoracic computed tomography (CT) angiogram and magnetic resonance angiography (MRA) showed massive ascending and descending TAAs (Figures 1-4, Video 1). The ascending TAA orthogonally measured $6.9 \times 6.3 \times 6.1$ cm at the sinuses of Valsalva, 8.6×8.4 cm at the sinotubular junction, 8.7×8.7 cm at the mid ascending aorta, and 4.0×3.9 cm at the proximal aortic arch. The proximal descending TAA measured 8.3×7.5 cm. There also was an aortic coarctation just proximal to the descending TAA

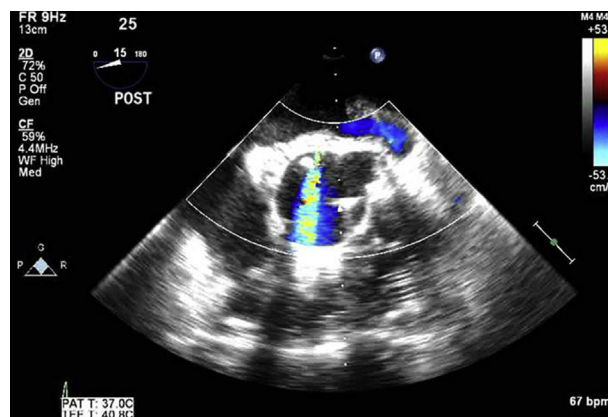


Figure 12 TEE, short-axis view of the aortic valve with color flow after surgery.

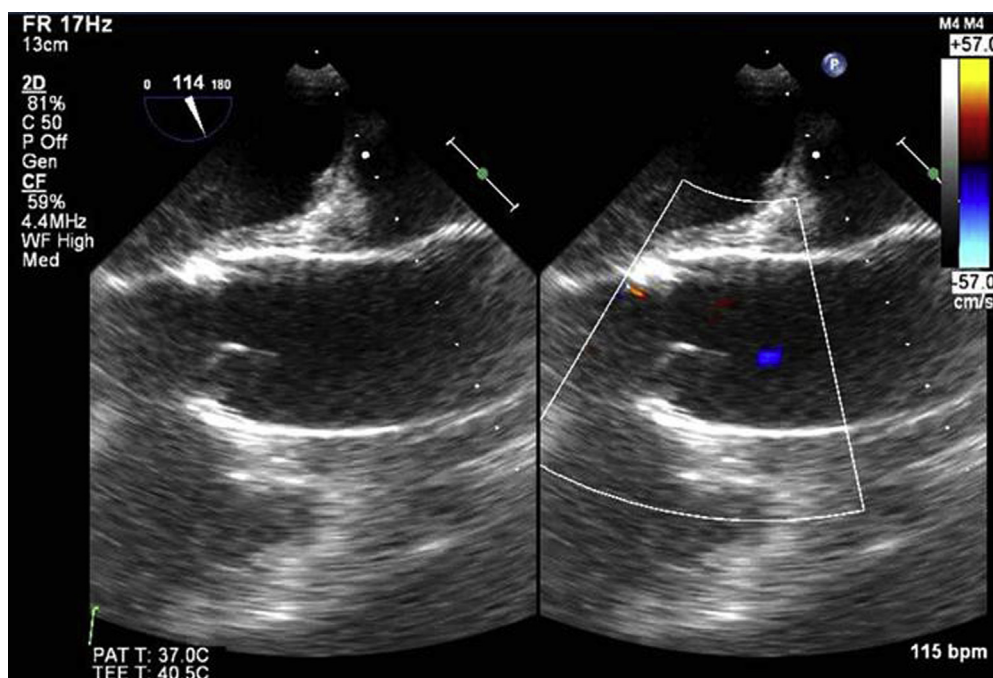


Figure 13 TEE, long-axis view of the aortic valve and aortic graft with color flow after surgery.

(Figures 1 and 5). Despite the presence of this aortic coarctation, the patient did not manifest systemic hypertension, likely due to the severe LV systolic dysfunction prevailing at the time of clinical presentation. On CT, there were no collaterals visualized extending to the intercostals. Interestingly, the ascending and descending TAA caused a mass effect on the proximal right and proximal left pulmonary arteries, respectively (Figures 4, 6, and 7) with thin linear lines in the distal transverse arch and mid descending aorta representing short dissection flaps (Figures 2 and 3). The transthoracic echocardiogram (TTE) revealed a dilated LV (LV end-systolic diameter 5.6 cm, LV end-diastolic diameter 7.0 cm) and severe LV systolic dysfunction (ejection fraction of 23%; Videos 2 and 3). Severe aortic valve regurgitation was present with pressure half time of 165 msec and holodiastolic flow reversal (Figures 8 and 9).

Cardiac MRA nicely delineated the expanse of the aneurysms (Video 4). Additionally, four-dimensional (4D) flow analysis showed profound vorticeal flow at the initiation of the ascending TAA and distal to the aortic coarctation as well regurgitant flow extending into great vessels coming off the aorta (Video 5, Figure 10). Coronary angiogram revealed a left dominant system with nonobstructive coronary artery disease. Left heart catheterization showed an LV end-diastolic pressure of 27 mm Hg with rapid rise in diastolic pressure and widened pulse pressure consistent with severe aortic valve regurgitation.

The patient underwent a staged surgical approach. Regarding his first operation, intraoperatively the aortic root aneurysm measured nearly 9 cm and the aortic valve was trileaflet. The transesophageal echocardiogram (TEE) showed the type B aortic dissection at 25 cm distal to the incisors (Figure 11). The proximal dissection flap also appeared to be the aortic coarctation shelf. The patient then underwent a standard Bentall operation with aortic valve replacement (29-mm Carpentier-Edwards Perimount bovine pericardial aortic valve prosthesis), aortic root/ascending aorta replacement with graft (36-mm Gelweave Dacron graft), and reimplantation of the coronary ostia. The postoperative TEE revealed qualitatively improved LV

function, a well-seated aortic valve and aortic root/ascending aorta graft replacement, and no aortic insufficiency (Figures 12 and 13). The postoperative TTE 2 days later showed reduction in LV end-systolic diameter and LV end-diastolic diameter to 5.1 cm and 6.2 cm, respectively, LV ejection fraction of 30%, no aortic regurgitation, and well-seated prostheses. Postoperative MRA/cardiac magnetic resonance imaging (MRI) revealed a well-positioned aortic graft and improved vorticeal flow in the ascending aorta (Videos 6 and 7, Figure 14). The aortic valve was trileaflet, fibrotic, and myxomatous on pathologic analysis.

Approximately 8 weeks later, our patient underwent surgical correction of his aortic coarctation, descending TAA, and type B aortic dissection. Intraoperatively, his descending TAA was found to be massively dilated (approximately 8 cm) and distorted all of the associated anatomy (Figure 15). There were very few collaterals. The coarctation was juxtaductal to the ductus arteriosus. The patient had a 28-mm Gelweave Dacron interposition graft placed that extended from the left subclavian to the diaphragmatic hiatus. Postoperative 4D MRA showed a well-positioned graft and normalized vorticeal flow within the descending aorta (Video 8, Figure 16). Pathology of the aorta revealed medial fibrosis and atherosclerotic changes. We performed a comprehensive sequence analysis of 16 genes linked with TAA and dissection, Marfan syndrome, and related connective tissue disorders (e.g., Loeys-Dietz syndrome, Ehlers-Danlos syndrome). The specific genes tested included ACTA2, CBS, COL3A1, COL5A1, COL5A2, FBN1, FBN2, FLNA, MED12, MYH11, SKI, SLC2A10, SMAD3, TGFBR1, and TGFBR2. Our patient was negative for any pathogenic variant of the genes tested. It is possible that his associated aortic pathologies are driven by an oligogenic inheritance pattern (i.e., controlled by a few genes) that we cannot yet probe biochemically. Clinically, he is doing well and is asymptomatic. His postoperative TEE revealed an LV ejection fraction of 50%. He continues on appropriate medical management for systolic heart failure, including neurohormonal blockade.

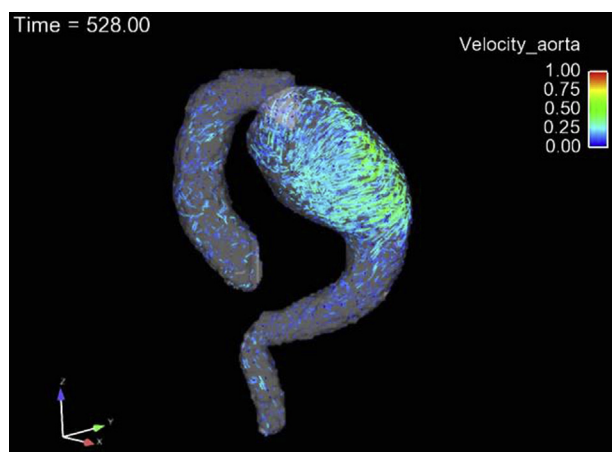


Figure 14 Cardiac MRI/MRA 4D flow analysis after the first surgery.

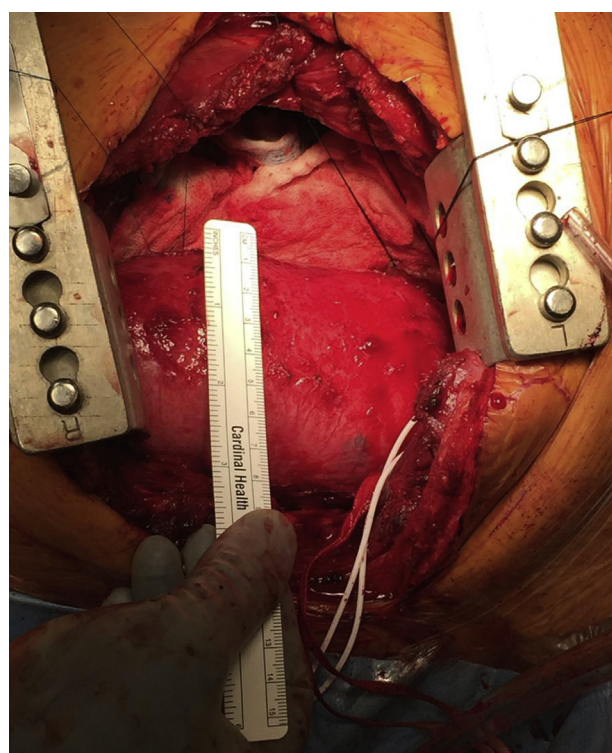


Figure 15 Intraoperative photograph of massive descending thoracic aneurysm at the time of surgery.

DISCUSSION

Here we present a case of a rare combination of massive ascending and descending TAAs, type B aortic dissection, and severe aortic valve regurgitation further complicated by severe LV systolic dysfunction in a patient with an uncorrected aortic coarctation. We used a multimodality imaging approach including echocardiography (transthoracic and transesophageal), CT, cardiac MRI/MRA, and 4D flow cardiovascular MRI to delineate the severity and complexity of our patient's associated aortopathies and appropriately plan surgical intervention. Importantly, we used 4D flow cardiovascular MRI to comprehensively evaluate and illustrate the complex array of flow

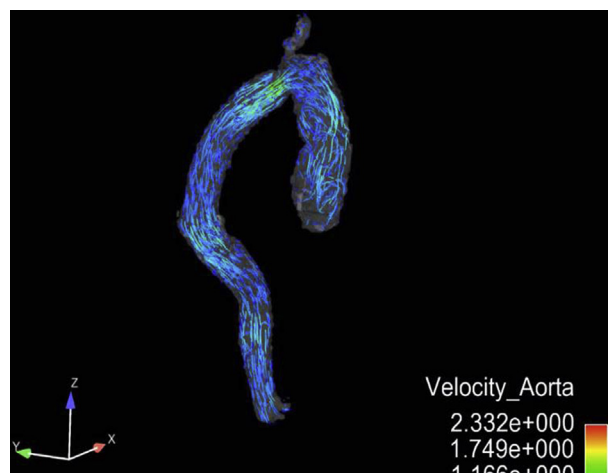


Figure 16 Cardiac MRI/MRA 4D flow analysis after the second surgery.

dynamics that occurred in our patient's large aneurysms and how the flow dynamics changed after surgical repair. Our case illustrates that 4D flow cardiovascular MRI has tremendous clinical application and utility in assessing blood flow through the heart and great vessels in various cardiovascular physiologic and pathophysiologic states. While there are several known genetic causes of such potentially impressive aortopathy, our patient appears to have developed his aortic pathology sporadically. To our knowledge, this is the first known report with this combination and severity of aortopathy.

CONCLUSION

Uncorrected aortic coarctation, while one of the more common congenital cardiovascular abnormalities, is infrequently observed in adults. Moreover, when it is present, coarctation of the aorta is seen more often with a bicuspid aortic valve in adult patients. However, we observed a very unique combination of massive ascending and descending aortic aneurysms, type B aortic dissection, and severe aortic valve regurgitation in a middle-aged male patient with a tricuspid aortic valve. Until now, such a unique combination of aortopathy had not been described in the literature.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.06.004>.

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