Ventricular Dysfunction in a 40-Year-Old With Coronary Compression From Aortic Aneurysm Following Waterston Shunt and Complete Repair of Tetralogy of Fallot

Carolyn M. Wilhelm, MD, MS, Jenne Hickey, FNP-BC, Patrick McConnell, MD, Kan N. Hor, MD, and May Ling Mah, MD, MS, *Columbus, Ohio*

INTRODUCTION

Tetralogy of Fallot (TOF) is the most prevalent form of cyanotic congenital heart disease in the adult population and requires lifelong follow-up with a specialist in adult congenital heart disease to closely monitor for known long-term complications.[1,2] Common complications seen in TOF include aortic root dilation and sequelae related to right ventricular volume overload, although they can differ depending on the surgical history.[3,4] Recommendations for surveillance include annual imaging with either echocardiography or cardiac magnetic resonance imaging to evaluate anatomy and hemodynamics.[1,5] We present a case of an unusual complication related to surgical palliation in an adult with TOF requiring multimodality imaging for diagnosis.

CASE PRESENTATION

A 40-year-old male with TOF status post-Waterston palliation followed by complete repair at 6 years with ventricular septal defect patch, pulmonary valvectomy, and Waterston ligation was followed until age 28 with typical aortic root dilation. He was lost to follow-up until he presented with fatigue, exertional dyspnea, and chest pain. His physical exam at that time was significant for a 2/6 systolic ejection murmur, 2/4 low-frequency diastolic murmur, and 2/4 high-frequency diastolic murmur at the left sternum. He underwent transthoracic echocardiogram, which demonstrated aortic aneurysms cranial to both right and left coronary origins (Figure 1A), depressed left ventricular function (ejection fraction of 40%–45%), and mild to moderate aortic regurgitation (Videos 1 and 2). He had anticipated right ventricular enlargement and severe pulmonary insufficiency (Video 3). An exercise stress perfusion study was negative for inducible ischemia. He underwent subsequent cardiac computed tomography demonstrating aneurysm compression of the right (green arrow) and left (red arrow) coronaries (Figure 1B and 1C; Videos 4 and 5).

DISCUSSION

Our patient was an adult congenital patient lost to follow-up. Echocardiogram demonstrated saccular aneurysmal formation near both coronaries and abnormal left ventricular function concerning for coronary compression. Moderate left heart dysfunction is not typical or attributable to TOF. Given the aneurysm locations on echocardiogram, computed tomography scan was indicated to evaluate for coronary compression and culminated in urgent operative repair. While right ventricular dysfunction due to residual pulmonary insufficiency is routine, this case represents an unusual etiology of early left ventricular dysfunction. While aortic root dilation in TOF is common due to the conotruncal defect and cystic medial necrosis, this patient developed aneurysms at both his prior Waterston anastomosis and the aortic cannulation site. With new onset of angina, left ventricular dysfunction, and appearance of aortic aneurysm, he underwent prompt imaging to confirm coronary compression and referral for urgent surgical repair. Intraoperative findings were consistent with prior imaging (Figure 1D). The patient underwent successful ascending aortic replacement and inclusion-technique valve-sparing aortic root replacement as well as pulmonary valve replacement. At 6-month follow-up, left ventricular function had normalized (Videos 6 and 7).

CONCLUSION

Adult congenital patients with repaired TOF require life long follow-up with an adult congenital heart disease specialist to properly monitor for long-term complications from their disease and subsequent surgical repair. Multimodality imaging should be considered in patients with unexpected findings on echocardiography.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.10.001.

From the Nationwide Children's Hospital, Columbus, Ohio.

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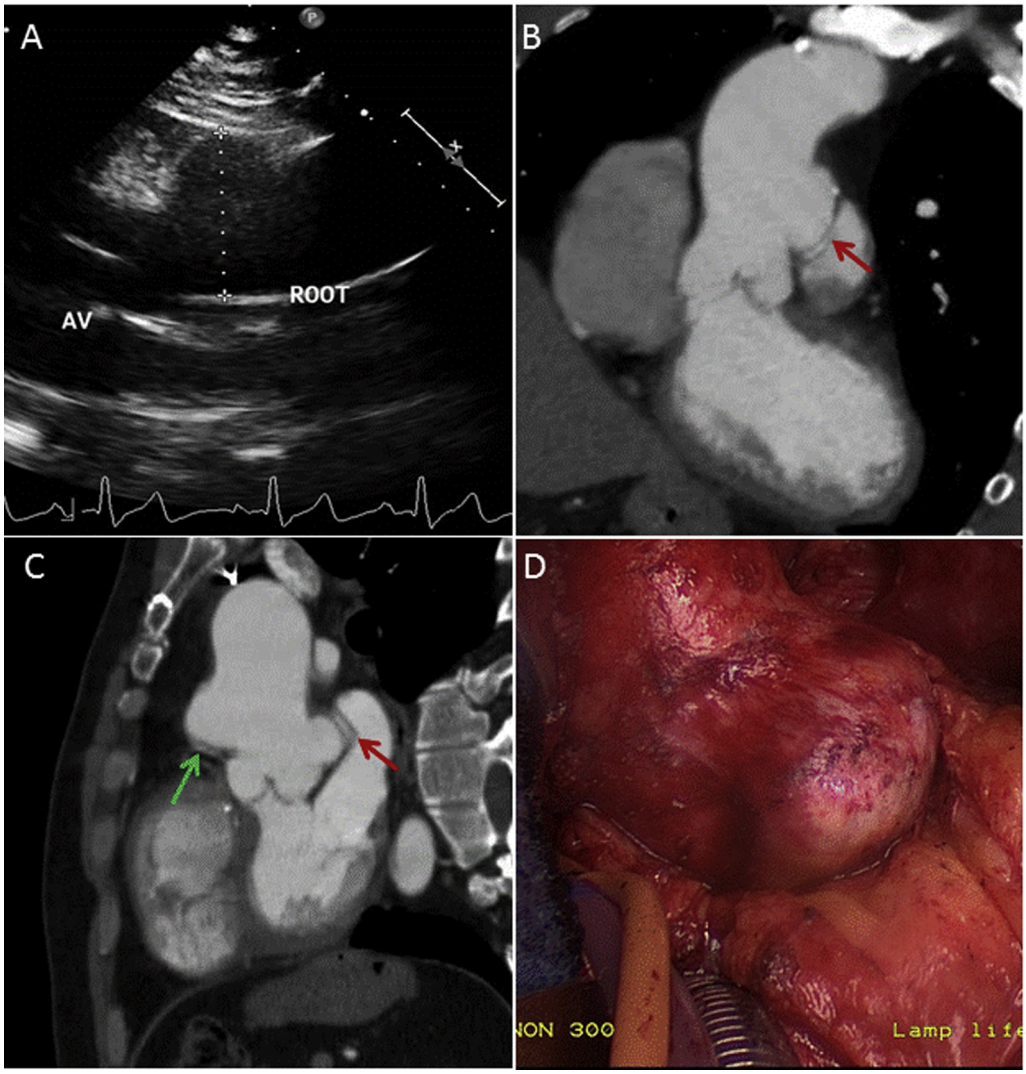

Figure 1 **(A)** Transthoracic echocardiogram parasternal long-axis view with left and right aortic aneurysms superior to the aortic root. **(B)** Cardiac computed tomography coronal oblique image demonstrating left aortic aneurysm with compression of the left coronary artery (*red arrow*). **(C)** Cardiac computed tomography sagittal oblique image demonstrating two aortic aneurysms with compression of the right (*green arrow*) and left (*red arrow*) coronaries. **(D)** Intraoperative image of the aortic root and ascending aorta prior to surgical repair. *AV*, Aortic valve.

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