JACC: CASE REPORTS VOL. 4, NO. 10, 2022

CASE REPORT

INTERMEDIATE

CLINICAL CASE

Tetralogy of Fallot and Aortic Dissection

Implications in Management

Sumeet S. Vaikunth, MD, MED,[a] Joshua L. Chan, MD,[b] Jennifer P. Woo, MD,[a] Michael R. Bykhovsky, MD,[a] George K. Lui, MD,[a,c] Michael Ma, MD,[b] Anitra W. Romfh, MD,[a,c] John Lamberti, MD,[b] Domenico Mastrodicasa, MD,[d] Dominik Fleischmann, MD,[d] Michael P. Fischbein, MD, PHD[b]

ABSTRACT

We present the case of a 61-year-old man with tetralogy of Fallot postrepair and mechanical aortic valve replacement with an aortic root/ascending/arch aneurysm with chronic type A aortic dissection. He underwent uncomplicated aortic root and total arch replacement. Continued surveillance for aortic aneurysm is necessary in the tetralogy of Fallot population. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:581-586) © 2022 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENT ILLNESS

A 61-year-old man with a tetralogy of Fallot (TOF) complete repair (age 5 years) followed by a mechanical aortic valve replacement (age 38 years) presented to an outside clinic with progressive exertional dyspnea. He was afebrile with a heart rate of 80 beats/min, blood pressure of 129/79 mm Hg, and a respiratory rate of 16 breaths/min with oxygen saturation of 97% on room air; his body mass index was 26 kg/m^2. Physical examination showed a median sternotomy scar and normal S1 and prosthetic S2 sounds with an early peaking grade 2 of 6 systolic murmur best heard at the left upper sternal border. He had 2+ palpable radial, femoral, and pedal pulses. He did not have a rub, gallop, peripheral edema, or elevated jugular venous pulsations.

LEARNING OBJECTIVES

- To recognize that patients with tetralogy of Fallot and aortic dilation can develop aortic dissection.
- To manage acquired cardiovascular risk factors in the tetralogy of Fallot population.
- To promote collaboration between adult and pediatric cardiologists and cardiothoracic surgeons.

PAST MEDICAL HISTORY

The patient's medical history included hypertension, hyperlipidemia, hemorrhagic stroke, central retinal artery occlusion, benign prostatic hypertrophy, and osteopenia. His social history was negative for tobacco use. His home medications included atorvastatin, losartan, verapamil, tamsulosin, and warfarin.

From the [a]Department of Medicine, Division of Cardiovascular Medicine, Stanford University School of Medicine, Palo Alto, California, USA; [b]Department of Cardiothoracic Surgery, Stanford University School of Medicine, Palo Alto, California, USA; [c]Department of Pediatrics, Division of Pediatric Cardiology, Stanford University School of Medicine, Palo Alto, California, USA; and the [d]Department of Radiology, Stanford University School of Medicine, Palo Alto, California, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received August 11, 2021; revised manuscript received February 10, 2022, accepted February 27, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.02.021

ABBREVIATIONS AND ACRONYMS

CT = computed tomography

TOF = tetralogy of Fallot

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of exertional dyspnea in an adult patient with history of TOF surgical repair is broad. It includes pulmonary or tricuspid regurgitation, right heart failure, left ventricular heart failure with reduced or preserved ejection fraction, endocarditis, arrhythmia, and/or coronary artery disease.

INVESTIGATIONS

Transthoracic echocardiogram showed normal left ventricular systolic function with moderate concentric hypertrophy, normal right ventricular function, a mechanical aortic valve with a mean gradient of 22 mm Hg, and a small restrictive residual ventricular septal defect patch leak with left-to-right shunt. There was no right ventricular outflow tract obstruction and trace pulmonary and tricuspid regurgitation. Also shown was a dilated ascending aorta measuring 6.1 cm. A computed tomography (CT) scan of the chest revealed an aortic root, ascending aorta, and arch aneurysm. The ascending aorta measured 6.9 cm in largest dimension compared with 5.3 cm on a scan in 2016. The patient was therefore referred for surgical evaluation. No other studies were performed. A repeat CT scan of the chest, abdomen, and pelvis done 10 days later for surgical planning revealed a Stanford type A aortic dissection that extended from the aortic root and ended in the arch (Figure 1). A single coronary artery arose from the left sinus with a retroaortic course of the right coronary artery (Figure 2).

MANAGEMENT

General anesthesia was induced and neuromonitoring used. A third time redo sternotomy was performed. Cardiopulmonary bypass was initiated, and the patient cooled systemically to 24 °C. A large primary intimal tear was identified within the aortic root (Figure 3). The mechanical valve was removed, and an aortic root replacement performed (bioroot: 29-mm Edwards Lifesciences INSPIRIS RESILIA aortic valve sewn inside a 32-mm Dacron graft). The single coronary artery was reimplanted (Figure 4). A total arch replacement (zone 2) was performed by using unilateral selective antegrade perfusion for neuroprotection via an axillary artery perfusion graft with stable head oxygen saturations with near-infrared oximetry (Figure 5). A branched prosthesis with separate reimplantation of the innominate and left carotid arteries allowed us to excise the aneurysmal and dissected aorta entirely. The branch technique provides several advantages over the island technique, including that: 1) the anastomoses are performed where the arteries are free of dissection and

FIGURE 1 Preoperative Computed Tomography Scan Showing Stanford Type A Aortic Dissection Extending From the Aortic Root to Arch

FIGURE 2 Preoperative Computed Tomography Scan Showing Single Coronary Artery From the Left Sinus With a Retroaortic Right Coronary Artery Course

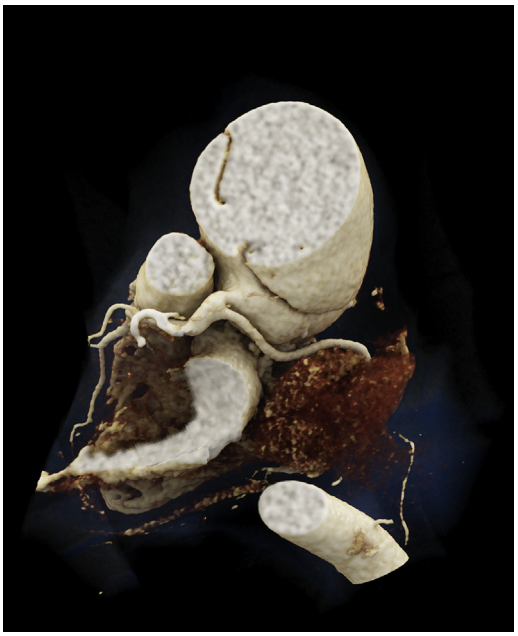
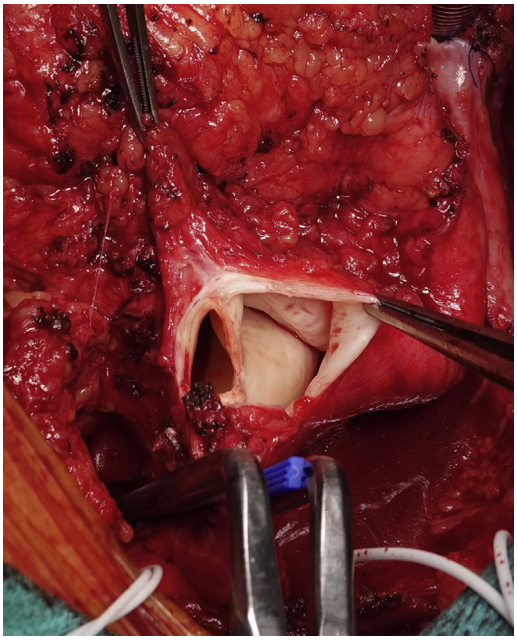
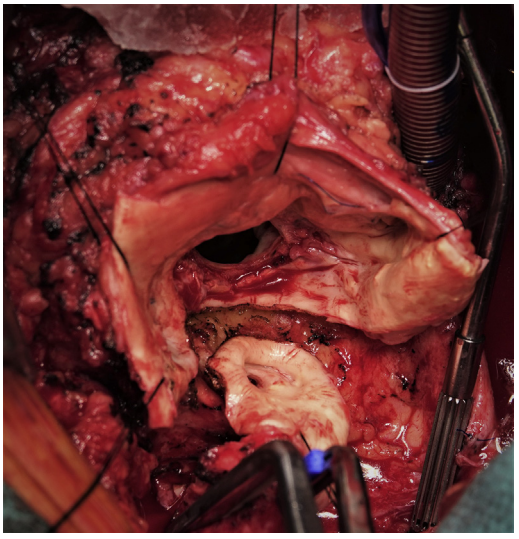


FIGURE 3 Intraoperative Photograph Demonstrating Chronic Type A Dissection With True and False Lumens



Photograph demonstrating chronic type A dissection with true **(left)** and false **(right)** lumens.

FIGURE 4 Intraoperative Photograph Demonstrating the Aortic Root With Previous Mechanical Aortic Valve and Single Coronary Artery Ostium



atherosclerosis; 2) the pathologic arch tissue can be completely excised; and 3) bleeding from the arch vessel anastomoses can be more easily controlled.

The patient was extubated on postoperative day 1, had an uneventful postoperative recovery, and was discharged home on postoperative day 8. A repeat CT scan showed the newly replaced aorta without complications (**Figures 6 and 7**).

DISCUSSION

Although TOF is considered a disease of the right side of the heart and right ventricular outflow tract, aortic dilatation is present in at least 20% of patients with repaired TOF.[1] Aortic dilation can result from the increased aortic flow due to right-to-left shunting at the ventricular septal defect before repair, but it is not known what causes further dilation after TOF repair. However, a retrospective autopsy study showed higher grade histologic changes (including the presence of medionecrosis, fibrosis, cystic medial necrosis, and elastic fragmentation) in the ascending aortas of a cohort of TOF patients compared with normal control subjects.[2] Similarly, a subsequent prospective study of aortic dilation in TOF found that 75% of patients had histologic abnormalities of the aortic wall, including elastic tissue fragmentation, increased ground substance, medionecrosis, muscle disarray, and fibrosis.[3] A recent analysis of hospitalized TOF patients from the National Inpatient Sample found 11 cases of aortic dissection with mortality of 45%.[4] In addition, 6 case reports of aortic dissection in TOF (**Table 1**) have been reported in the literature.[5-10]

Unlike aortopathies associated with bicuspid aortic valve, in which the 2020 American College of Cardiology valvular heart disease guidelines establish a diameter of >5.5 cm as a Class I indication for aortic root replacement (or a diameter of 5-5.5 cm as a Class IIb indication if surgery is being performed at a Comprehensive Valve Center), there are no statements in the 2018 American College of Cardiology/American Heart Association Adult Congenital Heart Disease guidelines on the management of the dilated aortic root in TOF.[11] Although a dilated aortic root is not uncommon in the adult with repaired TOF, conservative management and observation in the setting of aortic root dilation with conotruncal defects such as TOF are typically recommended.[12]

Age >60 years, male sex, and hypertension are associated with aortic dissection in the TOF population.[4] Perhaps not surprising, our patient exhibited all 3 risk factors. His initial repair was performed at a later age, which has been associated with a dilated aortic root. As the TOF patients continue to age, they are susceptible to acquired atherosclerotic cardiovascular risk factors.[13] These acquired comorbidities

FIGURE 5 Postoperative Image of Redo-Aortic Root and Total Arch Replacement

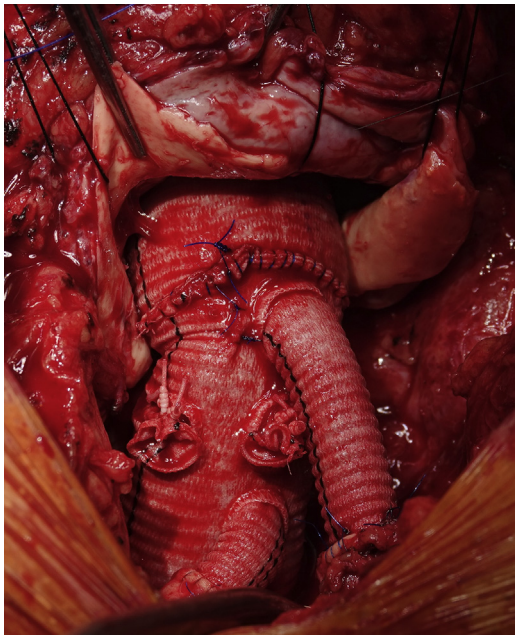
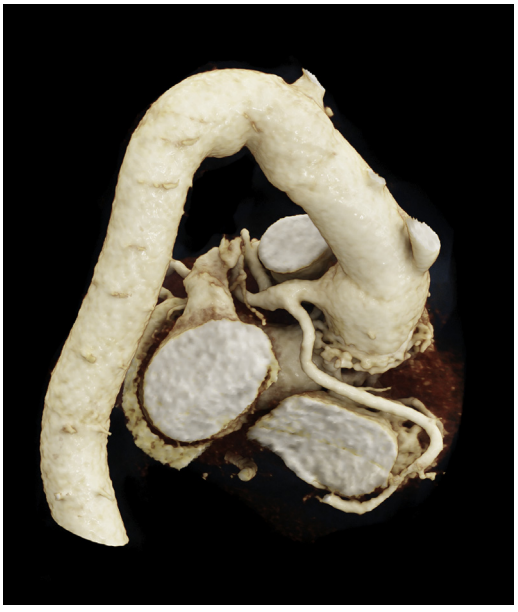
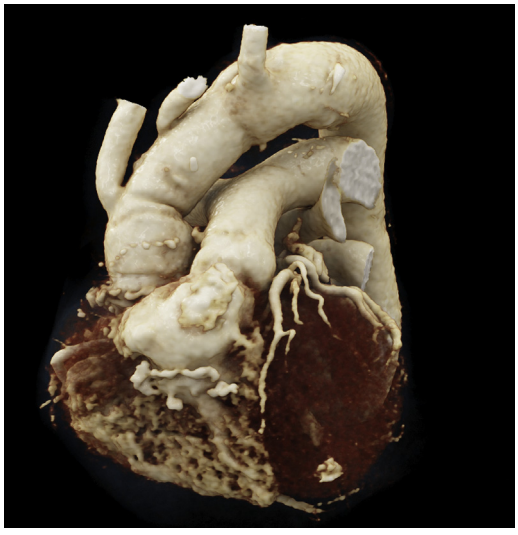


FIGURE 7 Postoperative Computed Tomography Scan Showing Posterior View of Total Arch Replacement and Reimplanted Coronary Artery



may serve as a "second hit" to the increased aortic flow before repair and intrinsic aortic medial aortopathy and lead to further dilatation of the aorta. Thus, in the presence of adult acquired comorbidities such as hypertension and atherosclerosis, one may consider aortic root replacement at a diameter of ≥5.5 cm and at 5.0 to 5.5 cm in the setting of reoperation for aortic or pulmonary valve replacement in TOF.

FIGURE 6 Postoperative Computed Tomography Scan Showing View of Great Arteries After Total Arch Replacement



This case will make us consider more frequent advanced imaging surveillance and earlier referral for prophylactic aortic root replacement in the setting of a patient with a conotruncal lesion and significant cardiovascular comorbidities (eg, hypertension). Importantly, immediate surgical repair is recommended in the setting of acute type A dissection. Although rupture and cardiac tamponade may be reduced in patients with previous sternotomies due to mediastinal adhesions, acute aortic regurgitation and coronary artery dissection or occlusion account for the high mortality rate in untreated acute type A dissection. The International Registry of Acute Aortic Dissection reports that medical therapy alone has a higher mortality rate compared with surgery (in-hospital and intermediate-term) even in patients with prior cardiac surgery, supporting an operative strategy for acute type A dissection.[14] Lastly, current guidelines for surgical repair in chronic type A dissection (>14 days) are based on degenerative aneurysm recommendations (5.5 cm). A recent study by Kim et al[15] reported increased adverse aortic events in 82 patients with chronic type A dissections over 19 years, noting a worse prognosis in dissected

TABLE 1 Literature Review of Prior Case Reports of Aortic Dissection in Tetralogy of Fallot

First Author, Year	Age, Presentation	Acute or Chronic Location and Size Histology	Treatment
Kim et al,[5] 2005	30-year-old man with chest pain	Acute Ascending aorta 7 cm Fibrous, thrombotic exudates. No cystic medial degeneration	Bentall procedure
Rathi et al,[6] 2005	36-year-old man with chest pain	Acute Ascending aorta 9.3 × 8.3 cm	Not available
Konstantinov et al,[7] 2010	18-year-old man with 22q11 deletion syndrome with chest pain	Acute Ascending aorta 6 × 7 cm	Valve sparing aortic root replacement was performed with a 30-mm Valsalva graft
Wijesekera et al,[8] 2014	60-year-old man with chest pain and dyspnea	Acute Ascending aorta 5.5 cm (4.9 cm ascending aorta 2 y earlier)	Bentall procedure
Jariwala et al,[9] 2017	30-year-old man unrepaired with dyspnea and back pain	Subacute NA	22-mm interposition graft in the ascending aorta was performed
Chow et al,[10] 2020	35-year-old man with chronic chest discomfort over past months	Acute Ascending aorta 8.8 × 7.6 cm (4.8 × 5.1 cm ascending aorta 3 y prior [stopped losartan]) Myxoid generation and focal calcification, mild cystic medial degeneration	Bentall

vs nondissected ascending aneurysms. We also believe that the natural history of aneurysmal chronic type A dissection is more precarious than nondissected ascending aneurysm and therefore recommend surgery at a lower aortic dimension (5 cm).

FOLLOW-UP

At the 12-month follow-up visit, the patient's blood pressure is well controlled on losartan and metoprolol.

CONCLUSIONS

We present the case of a 61-year-old man with TOF status postrepair with comorbidities of hypertension and hyperlipidemia who presented with aortic dissection. This is the seventh case reported of aortic dissection in this population. Serial surveillance of aortic aneurysms in TOF is necessary, and it would be reasonable to consider replacement of the aorta in TOF patients with an ascending aorta or aortic sinus with a diameter ≥5.5 cm.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Michael P. Fischbein, Department of Cardiothoracic Surgery, Stanford University School of Medicine, Falk CVRB, 300 Pasteur Drive, Falk CVRB ULN MC5407, Stanford, California 94305, USA. E-mail: mfischbe@stanford.edu.

REFERENCES

1. Mongeon F-P, Gurvitz MZ, Broberg CS, et al. Aortic root dilatation in adults with surgically repaired tetralogy of Fallot: a multicenter cross-sectional study. *Circulation*. 2013;127:172–179.

2. Tan JL, Davlouros PA, McCarthy KP, Gatzoulis MA, Ho SY. Intrinsic histological abnormalities of aortic root and ascending aorta in tetralogy of Fallot: evidence of causative mechanism for aortic dilatation and aortopathy. *Circulation*. 2005;112:961–968.

3. Chowdhury UK, Mishra AK, Ray R, Kalaivani M, Reddy SM, Venugopal P. Histopathologic changes in ascending aorta and risk factors related to histopathologic conditions and aortic dilatation in patients with tetralogy of Fallot. *J Thorac Cardiovasc Surg*. 2008;135:69–77. e11.

4. Egbe AC, Crestanello J, Miranda WR, Connolly HM. Thoracic aortic dissection in tetralogy of Fallot: a review of the National Inpatient Sample Database. *J Am Heart Assoc*. 2019;8:e011943.

5. Kim WH, Seo JW, Kim SJ, Song J, Lee J, Na CY. Aortic dissection late after repair of tetralogy of Fallot. *Int J Cardiol*. 2005;101:515–516.

6. Rathi VK, Doyle M, Williams RB, Yamrozik J, Shannon RP, Biederman RWW. Massive aortic aneurysm and dissection in repaired tetralogy of Fallot; diagnosis by cardiovascular magnetic resonance imaging. *Int J Cardiol*. 2005;101:169–170.

7. Konstantinov IE, Fricke TA, d'Udekem Y, Robertson T. Aortic dissection and rupture in

adolescents after tetralogy of Fallot repair. *J Thorac Cardiovasc Surg*. 2010;140:e71–e73.

8. Wijesekera VA, Kiess MC, Grewal J, et al. Aortic dissection in a patient with a dilated aortic root following tetralogy of Fallot repair. *Int J Cardiol*. 2014;174:833–834.

9. Jariwala P, Kale SS, Sepur L, Padma Kumar EA. Tetralogy of Fallot, left ventricular clot, aortic dissection: rare association. *Asian Cardiovasc Thorac Ann*. 2017;25:534–536.

10. Chow P-C, Rocha BA, Au TWK, Yung T-C. Aortic dissection in a Chinese patient 31 years after surgical repair of tetralogy of Fallot. *J Cardiol Cases*. 2020;22:174–176.

11. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2021;77(4):e25–e197.

12. Stulak JM, Dearani JA, Burkhart HM, Sundt TM, Connolly HM, Schaff HV. Does the dilated ascending aorta in an adult with congenital heart disease require intervention? *J Thorac Cardiovasc Surg*. 2010;140:S52–S57.

13. Lui GK, Fernandes S, McElhinney DB. Management of cardiovascular risk factors in adults with congenital heart disease. *J Am Heart Assoc*. 2014;3:e001076.

14. Teman NR, Peterson MD, Russo MJ, et al. Outcomes of patients presenting with acute type A aortic dissection in the setting of prior cardiac surgery: an analysis from the International Registry of Acute Aortic Dissection. *Circulation*. 2013;128:S180–S185.

15. Kim WK, Park SJ, Kim HJ, Kim HJ, Choo SJ, Kim JB. The fate of unrepaired chronic type A aortic dissection. *J Thorac Cardiovasc Surg*. 2019;158:996–1004.e3.

KEY WORDS **aortic dissection, hypertension, tetralogy of Fallot**