An unusual cause of heart failure in postoperative tetralogy of Fallot

Gurbhej Singh, Arun Gopalakrishnan, Sivasubramonian Sivasankaran
Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India

ABSTRACT

The long-term results of transatrial transpulmonary tetralogy of Fallot (TOF) repair have been excellent. Progressive pulmonary regurgitation and consequent right ventricular (RV) dilatation are the most common long-term sequel of definitive repair in childhood. Overt systemic venous congestion after TOF repair is limited to the rare setting where RV dysfunction sets in due to deferred surgery or progressive arrhythmia. Here, we report a unique case of right heart failure from an unexpected etiology, 28 years after TOF repair. Cardiac catheterization confirmed findings of elevated right heart pressures. Magnetic resonance imaging showed obliteration of the RV apex with late gadolinium enhancement of the right ventricular apical endomyocardium.

Keywords: Congenital heart disease, echocardiography, endomyocardial fibrosis, heart failure, Tetralogy of Fallot

INTRODUCTION

The long-term outcomes of transatrial transpulmonary tetralogy of Fallot (TOF) repair have been excellent. While TOF repair is best undertaken at 6–12 months of life, late presentation and repair is still prevalent in the developing world.[1] This constitutes a mixed bag – patients often have issues related to chronic hypoxemia, higher rates of pleural and pericardial effusion but with comparable rates of sepsis, early mortality, and overall shorter hospital stay.[2,3] Progressive pulmonary regurgitation and consequent right ventricular (RV) dilatation are the most common long-term sequel of definitive repair in childhood. Overt systemic venous congestion after TOF repair is limited to the rare setting where RV dysfunction sets in due to deferred surgery or progressive arrhythmia. Here, we report a unique case where a patient developed right heart failure from an unexpected etiology, 28 years after TOF repair in adolescence.

Video avaliable on: www.annalspc.com

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.apc_181_21

CASE REPORT

A 14-year-old boy born to nonconsanguineous parents, hailing from Kanyakumari, along the southern coast of India was diagnosed with TOF and referred to our institute. He underwent a cardiac catheterization and was found to have elevated right-sided filling pressures and systemic RV pressures. Right atrial mean pressure was 15 mmHg and RV end diastolic pressure was 26 mmHg. The RV apex was reported to be muscle bound in the preoperative angiogram. He underwent transatrial intracardiac repair with transannular patch leaving behind a patent foramen ovale. Intraoperatively, the right ventricle was noted to be severely hypertrophied with severe infundibular obstruction. The right atrium was noted to be normal in size. The postoperative period was complicated by right-sided pleural effusion which was managed with intercostal drain for 6 days. The patient also underwent surgical re-exploration on the 2nd postoperative day in view of bleeding. Re-exploration ruled out surgical causes of bleeding. The patient also had

How to cite this article: Singh G, Gopalakrishnan A, Sivasankaran S. An unusual cause of heart failure in postoperative tetralogy of Fallot. Ann Pediatr Card 2022;15:308-10.

Address for correspondence: Dr. Arun Gopalakrishnan, Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India.
E-mail: arungopalakrishnan99@gmail.com

Submitted: 14-Sep-2021 **Revised:** 01-Apr-2022 **Accepted:** 04-Apr-2022 **Published:** 16-Nov-2022

right heart failure with elevated jugular venous pressure, pulsus paradoxus, and hepatomegaly in the postoperative period, which improved with diuresis and salt restricted diet. A tiny residual perimembranous ventricular septal defect (VSD) was also noted and was kept on conservative management. He remained asymptomatic until he was lost to follow-up, 5 years after surgery.

He presented again at 40 years of life with functional class II dyspnea on exertion for 6 months and recent onset pedal edema. He was noted to be in atrial fibrillation with controlled ventricular rate. The mean jugular venous pressure was elevated with prominent v waves and sharp y descent. Echocardiography revealed moderate tricuspid and pulmonary regurgitation and a dilated right ventricle. A restrictive perimembranous VSD was noted shunting left-to-right during systole and right-to-left during diastole [Figure 1a and b]. Closer evaluation showed apical obliteration of the RV apex and reduced trabeculations. Cardiac magnetic resonance imaging showed RV apex obliteration [Figure 1c and d] with late gadolinium enhancement of RV apical endomyocardium. RV volumes were 145 ml/m^2 in end-diastole and 85 ml/m^2 in end systole.

After stabilization, cardiac catheterization was done to study the hemodynamics [Table 1]. Right atrial mean was elevated. There was no gradient from RV inflow to RV outflow and pulmonary artery pressures were normal [Figure 2]. Low cardiac output state was confirmed (cardiac index 1.68 L/min/m^2). Simultaneous pressure recording from the RV and left ventricle suggested concordant changes with respiration. The absence of ventricular interdependence ruled out significant pericardial constriction. There was a 14% step up in oxygen saturation from the mixed venous to pulmonary artery level with a shunt fraction of 1.3:1. RV angiogram showed RV apical obliteration and moderate tricuspid regurgitation [Video 1]. Left ventricular angiogram showed normal left ventricular contour with good ventricular function and a small subaortic VSD shunting left to right.

He was instituted on diuretics for stabilization. He has been listed for early pulmonary valve replacement with bidirectional Glenn shunt.

DISCUSSION

Endomyocardial fibrosis (EMF) is a dwindling disease worldwide, now limited to coastal India and parts of Africa and South America.[4] It is characterized by fibrotic replacement of ventricular myocardium leading to impaired relaxation, thus causing elevated diastolic pressures and valvular regurgitation. While medical management is restricted to the treatment of heart failure, surgical options include endocardial decortication and valve repair or replacement. Further, the role of bidirectional Glenn has been reported in advanced course of the disease.[5]

Ours is a unique case where this tropical disease process complicated the long-term management of TOF. The index patient was documented to have elevated right-sided filling pressures at the age of 14 years. However, in the absence of ventricular dilatation or effacement, EMF

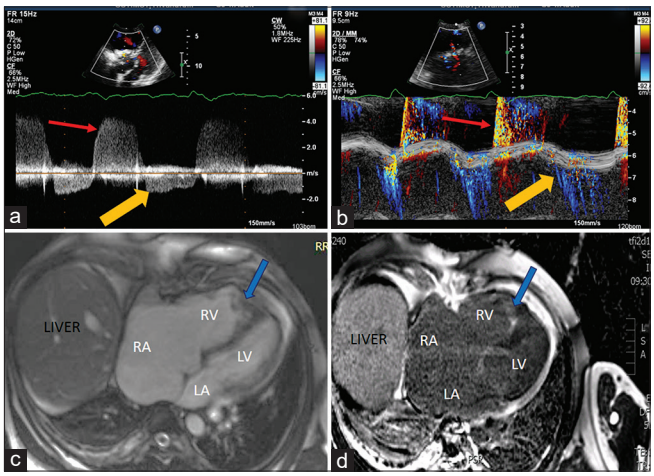

Figure 1: (a) Continuous wave Doppler across ventricular septal defect showing left-to-right shunt (red arrow) during systole and right-to-left shunt during diastole (yellow arrow). (b) Color M-Mode shows the left-to-right shunt (red arrow) during systole and right-to-left shunt during diastole (yellow arrow). (c) Magnetic resonance image showing RV apical obliteration (Blue arrow). (d) Magnetic resonance imaging showing late gadolinium enhancement of RV apex endomyocardium (Blue arrow). RV: right ventricular

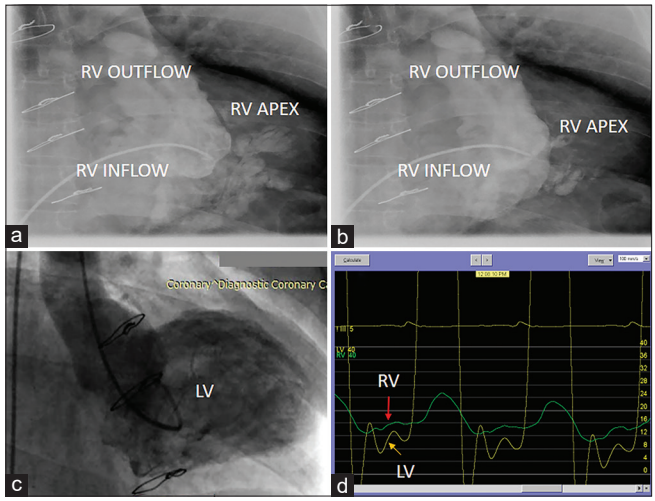

Figure 2: (a) RV angiogram (RAO 30°) during diastole shows apical obliteration and diverticuli resembling a caterpillar. (b) RV angiogram during systole shows disappearances of diverticuli. (c) LV angiogram in RAO view shows no involvement of left ventricle and site of VSD jet (red arrow). (d) Simultaneous left ventricular and right ventricular pressure traces showing RV ed 17 mmHg, LV ed 10 mmHg. LV ed – Left ventricular end-diastolic pressure, RV ed – Right ventricle end diastolic pressure. RV: Right ventricular, RAO: right anterior oblique, LV: Left ventricular, VSD: Ventricular septal defect

Table 1: Cardiac catheterization data of the patient at 40 years of life

Site	Pressure	Oxygen saturation (FiO_2 0.21)
Superior caval vein	V28 m16	50%
Inferior caval vein	V29 m16	67%
Mixed venous		55%
Right atrium	V28 m16	
Right ventricle (inflow)	29 dd 12 Ed 18	
Right ventricle (outflow)	28 dd 12 Ed 17	
Main pulmonary artery, systole/diastole (mean)	22/12 (16)	
Right pulmonary artery, systole/diastole (mean)	22/12 (16)	69%
Left pulmonary artery, systole/diastole (mean)	22/12 (16)	69%
Pulmonary capillary wedge	V11 m9	
Left ventricle	128 Ed 10	
Ascending aorta	128/84 (104)	95%

dd: Dip diastolic pressure, Ed: End-diastolic pressure, FiO_2: Fraction of inspired oxygen

was not suspected and elevated filling pressures were attributed to the restrictive pattern seen in delayed presentation of TOF.[2] In hindsight, the elevated right heart pressures noted before surgical repair were due to concomitant presence of EMF. Unfortunately, the previous angiographic records were not available for review. It is extremely unusual to have marked elevation of right heart pressures alone in unoperated TOF even at 14 years of life. Lower preoperative saturation, higher hematocrit, RV dysfunction, aortopulmonary collaterals, and high preoperative RV outflow tract gradients are the factors associated with prolonged postoperative course in delayed repair of TOF.[3] The coastal districts of southern Tamil Nadu and Kerala constituted areas of higher incidence of EMF in the days when the disease was common.[6] While a diagnosis of EMF in its inflammatory stages would have helped us initiate anti-inflammatory therapy, it seems unlikely that it would have altered the surgical plan. Since the patient tolerated the postoperative period although with transient effusions, it is clear that RV volumes were adequate for biventricular repair. Obliteration of the apex and body of the right ventricle despite moderate pulmonary and tricuspid regurgitation after TOF repair was suggestive of an alternate pathology, EMF in this case. Concomitant restrictive hemodynamics of the right ventricle explain the onset of right heart failure and atrial fibrillation in the patient while the RV dilatation is only bordering the conventional volume-based indications for pulmonary valve replacement. Advanced EMF contributes to low cardiac output in postoperative TOF in addition to the issues related to pulmonary regurgitation and tricuspid regurgitation.

The diastolic right-to-left shunt across the residual VSD was due to the elevation of RV end-diastolic pressures.

CONCLUSIONS

In tropical countries, EMF can present with heart failure in operated congenital heart disease including TOF. EMF can be an underlying cause for restrictive physiology of the right ventricle in delayed presentation of TOF. RV dilatation from long-standing free pulmonary regurgitation might be subtle in postoperative TOF with concomitant RV EMF, leading to insidious onset of right heart failure and atrial arrhythmia. Advanced RV EMF accentuates right heart failure and low cardiac output in postoperative TOF.

Consent

Informed written consent was taken from the patient for publication.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Van Den Bosch E, Bogers AJ, Roos-Hesselink JW, van Dijk AP, van Wijngaarden MH, Boersma E, *et al.* Long-term follow-up after transatrial-transpulmonary repair of tetralogy of Fallot: Influence of timing on outcome. Eur J Cardiothorac Surg 2020;57:635-43.
2. Benbrik N, Romefort B, Le Gloan L, Warin K, Hauet Q, Guerin P, *et al.* Late repair of tetralogy of Fallot during childhood in patients from developing countries. Eur J Cardiothorac Surg 2015;47:e113-7.
3. Talwar S, Meena A, Choudhary SK, Saxena A, Kothari SS, Juneja R, *et al.* Repair of tetralogy of Fallot in or beyond the fourth decade of life. Congenit Heart Dis 2014;9:424-32.
4. Vijayaraghavan G, Sivasankaran S. Tropical endomyocardial fibrosis in India: A vanishing disease! Indian J Med Res 2012;136:729.
5. Mishra A, Krishna Manohar SR, Sankar Kumar R, Valiathan MS. Bidirectional Glenn shunt for right ventricular endomyocardial fibrosis. Asian Cardiovasc Thorac Ann 2002;10:351-3.
6. Sivasankaran S. Restrictive cardiomyopathy in India: The story of a vanishing mystery. Heart 2009;95:9-14.