Acquired Fontan paradox in isolated right ventricular cardiomyopathy

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ABSTRACT

A 44-year-old woman presented with features of congestive heart failure. Echocardiography revealed severe right ventricular dysfunction along with passive minimally pulsatile pulmonary blood flow suggesting very high systemic venous pressures. This was confirmed with cardiac catheterization in which the pressures of superior vena cava and inferior vena cava (19 mmHg) were higher than the pulmonary artery pressures (17 mmHg). Elevation of systemic venous pressures above the pulmonary venous pressures, Fontan paradox, to drive the forward flow, is a specific feature of artificially created cavopulmonary shunts. Late stage of isolated right ventricular cardiomyopathy resulted in the spontaneous evolution of Fontan circulation with a nonfunctional right ventricle in this patient.

Keywords: Acquired Fontan, isolated right ventricular cardiomyopathy, minimally pulsatile pulmonary blood flow, passive conduit

INTRODUCTION

Surgical creation of a Fontan circuit in a complex congenital malformation with a single functioning ventricle is a common palliative surgical procedure. However, establishment of a spontaneous Fontan physiology in adults with severe right ventricular dysfunction is a rare entity and has been reported in only a few cases till date.[1-3]

Isolated right ventricular (RV) cardiomyopathy in itself is also quite uncommon and is a diagnosis of exclusion.[4-6] There is a definite male preponderance. These cases present as heart failure/syncope and features of ventricular tachycardia or left bundle branch block on electrocardiography. Many cases of suspected isolated RV cardiomyopathy turn out to be arrhythmogenic right ventricular cardiomyopathy (ARVC) with fibrofatty infiltration. Other differential diagnosis being Uhl's anomaly, isolated RV noncompaction cardiomyopathy, familial RV cardiomyopathy, and ARVC.[4]

We report a case of spontaneous Fontan physiology in a 44-year-old woman with RV cardiomyopathy.

CASE HISTORY

A 44-year-old woman, mother of two children, presented with gradually progressive ascites, pedal edema, and generalized fatigue for over the past 1 year. Jugular venous pressure was intracranial, with mild cardiomegaly and a loud RV third heart sound. On echocardiography [Figure 1a], congested inferior vena cava with no respiratory variation indicated high right atrial pressure >20 mmHg. RV systolic function was poor [see Videos, Supplemental Digital Content 1-3, which demonstrate poor RV systolic function] with tricuspid annular planar excursions less than 10 mm [Figure 1b]. Doppler study of the right-sided chambers showed rapid

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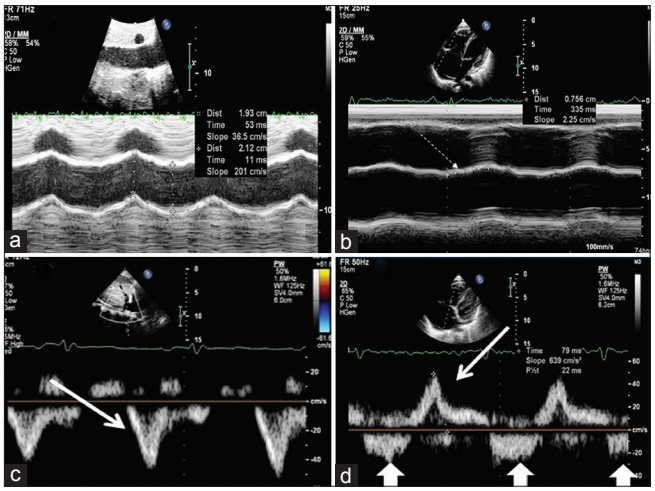

Figure 1: 2D echocardiographic evaluation. (a) M-mode of inferior vena cava with no inspiratory collapse. (b) Tricuspid annular planar excursion of 7.5 mm. (c) Pulse Doppler of hepatic vein with maximum forward flow in early diastole (arrow). (d) Pulse Doppler across the tricuspid valve with maximum filling in early diastole (thin arrow) and low-pressure tricuspid regurgitation (thick arrows)

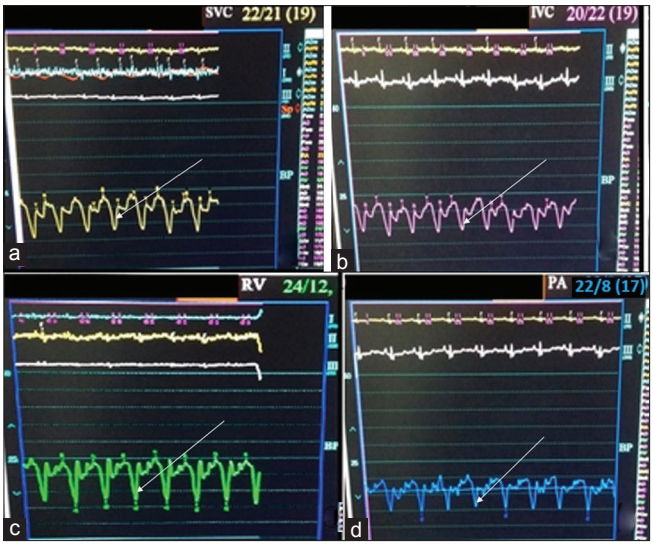

Figure 2: Hemodynamic tracings of the cardiac chambers. Mean pressures of SVC and IVC were higher than PA mean pressures explaining the preatrial systolic forward flow into PA (noted in Figure 3). Note the ventricularization of pressures of all the right-sided chambers denoting the rapid early diastolic filling of the right ventricle accounting for transient early-diastolic pulmonary regurgitation. IVC: Inferior vena cava, PA: Pulmonary artery, RV: Right ventricle, SVC: superior vena cava

early diastolic filling [thin arrows; Figure 1c and d)]. The high venous and RV end-diastolic pressures [Figure 2] kept the pulmonary valve open, with preatrial systolic forward flow into pulmonary artery [broad arrows, Figure 3, see Video, Supplemental Digital Content 4, showing continuous flow in pulmonary artery with pre atrial systolic flow and early diastolic pulmonary regurgitation]. Owing to the high venous pressures, the impact of atrial systole was minimal in hepatic veins and across the tricuspid valve, but generated a presystolic forward flow into pulmonary artery.

Cardiac catheterization confirmed higher superior vena cava and inferior vena cava (19 mmHg) pressures compared with the pulmonary artery pressures (17 mmHg) [Figure 2]. The pulmonary artery waveforms were similar to the venous pressure tracings. Left heart functions were normal [see Video, Supplemental Digital Content 5, which demonstrated normal LV volumes and function].

Magnetic resonance imaging showed dilated and dysfunctional right ventricle with late gadolinium enhancement of the basal inflow and body of right ventricle with an organized thrombus at the RV apex and normal left ventricle. A diagnosis of RV cardiomyopathy was made.

The elevated systemic venous pressures above the pulmonary venous pressures drove the cardiac output in this case of RV cardiomyopathy with severe RV dysfunction, thus creating an acquired Fontan circulation with RV acting as a passive conduit. The disease could have been long standing and asymptomatic in this patient. Age-related increase in left ventricular filling pressures reflected in systemic venous pressures to manifest as heart failure. Successful asymptomatic vaginal delivery of two kids essentially represents the hemodynamic adjustments that are possible in case of isolated RV cardiomyopathy.

DISCUSSION

Fontan paradox refers[7,8] to elevation of systemic venous pressures above the pulmonary venous pressures to drive the cardiac output in the absence of a functioning right ventricle, after total cavopulmonary connection surgery, but can rarely occur in severe RV dysfunction.[1-3] Although surgical creation of Fontan circuit is a common practice, to the best of our knowledge, only a few cases of spontaneously evolved Fontan circulation have been described in literature[1-3] in which noncontractile right ventricle behaved as a passive conduit. The RV end-diastolic and systemic venous pressure being higher than the pulmonary pressures caused the premature opening of the pulmonary valve and facilitated the pulmonary blood flow during diastasis. In our case, RV cardiomyopathy was the cause of severe RV dysfunction. This was labeled as isolated RV cardiomyopathy because there was no evidence of infiltration of right ventricle and left ventricle was normal.

Preatrial systolic flow in the pulmonary artery reflects a transient hemodynamic situation wherein systemic venous pressure exceeds the pulmonary arterial

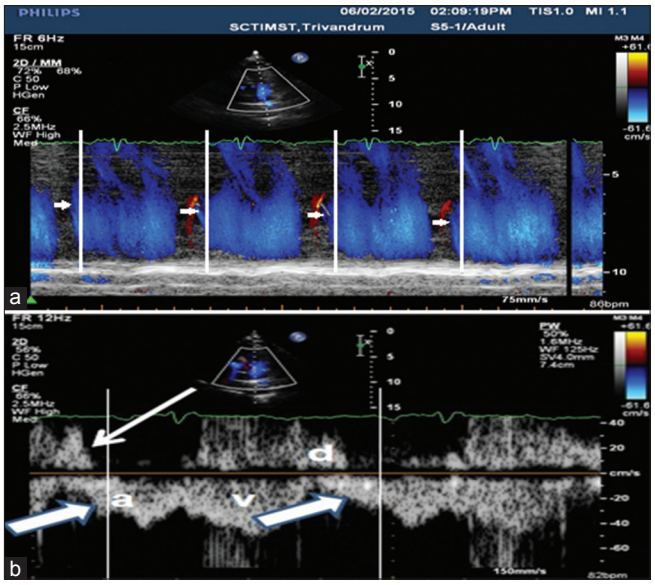

Figure 3: Color M-mode of the pulmonary artery showing preatrial systolic flow (note the flow in pulmonary artery prior to the P wave on electrocardiography marked with horizontal arrows). (a) Pulmonary artery pulse wave Doppler showing flow prior to (b) atrial systole marked with broad arrows

pressure during diastasis. This causes the flow in the pulmonary artery similar to a Fontan circulation during diastasis [Figure 3].

The patient is currently well on oral anticoagulants and decongestive measures.

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Conflicts of interest

There are no conflicts of interest.

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