Pandiastolic antegrade flow in patient with severe right ventricle to pulmonary artery conduit stenosis: An interesting phenomenon

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ABSTRACT

In a post operative tetralogy of fallot (TOF) physiology patient, abnormal right ventricular (RV) function remains the greatest matter of concern. Due to restrictive RV diastolic dysfunction, there is detectable antegrade diastolic flow in the pulmonary artery during atrial systole. We report a case of 21 year old male patient with total correction done in infancy using right ventricle to pulmonary artery conduit. He was relatively asymptomatic with a unique pattern of antegrade diastolic flow in both early and late diastolic phases (pan diastolic). This physiology was supportive and made him relatively asymptomatic. We discuss the physiology and clinical implication of the same.

Keywords: Diastolic dysfunction, pandiastolic antegrade flow, postoperative tetrology of Fallot

INTRODUCTION

Abnormal right ventricular (RV) physiology is the most important problem encountered following total correction of tetralogy of fallot (TOF) during late follow-up. Due to the restrictive RV diastolic dysfunction, there is detectable antegrade diastolic flow in the pulmonary artery (PA) during atrial systole. This case report highlights a unique pattern of antegrade diastolic flow in both early and late diastolic phases (pan diastolic) in a patient with severe RV outflow tract (RVOT) obstruction late after conduit repair. We discuss the physiology and clinical implication of the same.

CASE REPORT

A 21-year-old male was diagnosed to have double outlet left ventricle with normally related great arteries, subaortic ventricular septal defect (VSD), and severe pulmonary stenosis for which he underwent VSD closure and placement of RV to PA conduit (16 mm Dacron) at the age of 6 months. He was subsequently lost to follow-up and was asymptomatic until 1-year ago when he started to have mild dyspnea on exertion. On evaluation, he had stable vitals and ejection systolic murmur of grade 2/6 with late peaking at the upper left sternal border.

Echocardiography showed dilated right atrium (8.5 cm × 7 cm) and right ventricle (8 cm × 7 cm). He also had severe RV dysfunction along with tricuspid valve inflow restrictive pattern. Pulsed Doppler of the tricuspid valve during diastole showed higher velocities during the early filling phase (e>a) [Figure 1a] and on tissue Doppler velocities assessment e'> a' [Figure 1b] inferior vena cava (IVC) size was 2.59 cm on inspiration and IVC variation with respiration was 30%. There was severe stenosis of RV to PA conduit with peak gradient of 80 mmHg. The antegrade diastolic flow was detected [Figure 1c] which started after a gap, which correlated with isovolumic relaxation time phase. There was moderate tricuspid regurgitation with a maximum pressure gradient 72 mmHg. The anatomical features were defined by

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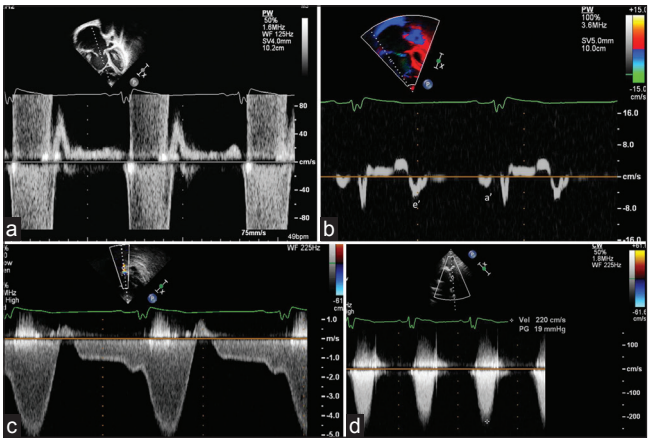

Figure 1: (a) Tricuspid inflow pattern was taken in end expiration during quite breathing showing elevated e wave velocity and deceleration time of 180 ms. (b) Tissue Doppler velocity taken across the tricuspid valve. (c) Two-dimensional echocardiography with continuous wave Doppler applied across the right ventricular outflow tract showing the characteristic pandiastolic antegrade flow across the right ventricular outflow tract. (d) Two-dimensional echocardiography with continuous wave Doppler applied across the right ventricular outflow tract showing the systolic antegrade flow into the right ventricular outflow tract with complete disappearance of the pandiastolic antegrade flow across the right ventricular outflow tract

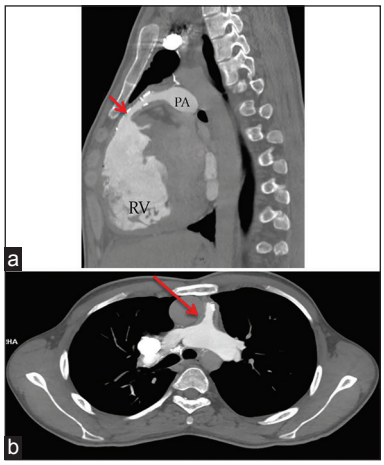

Figure 2: (a) Computerized tomography scan with sagittal cut showing the severely stenosed right ventricular to pulmonary artery conduit (marked by arrow) with dilated right ventricle. (b) Computerized tomography scan with axial cut showing the severely stenosed right ventricular to pulmonary artery conduit (arrow) with adequate sized distal branch pulmonary arteries

computerized tomography scan [Figure 2a and b].

He underwent RVOT reconstruction with bovine pericardium and pulmonary valve replacement with a 21 mm bioprosthetic perimount valve (Edwards Life Sciences, Irvine, California, USA) after excision of the previously placed conduit. Postoperative course was uneventful and he was successfully extubated on the 2nd postoperative day.

Postoperative echocardiography showed well opened RVOT, peak gradient of 20 mmHg, laminar flow in branch pulmonary arteries, mild PR, RV systolic dysfunction in the form of RV fractional area change of 28%, and tricuspid annular plane systolic excursion (TAPSE) of 1.16 cm. RV diastolic dysfunction was present (Tricuspid valve inflow e > a and TDI a > e) suggestive of pseudonormalization. There was no diastolic antegrade flow seen across RVOT [Figure 1d] as was seen preoperatively, left ventricular ejection fraction was normal.

DISCUSSION

Isolated RV restriction is common after TOF repair.[1] Although it reflects abnormal hemodynamics, the "a" wave contributes to forward pulmonary arterial flow and shortens the duration of pulmonary regurgitation. Consequently, there is less cardiomegaly and improved exercise performance in this group of patients. It was probably because of this reason that our patient was relatively asymptomatic inspite of having a severe stenosis of the RV to PA conduit and severe RV systolic dysfunction. The diastolic dysfunction with severe restrictive pattern favored the physiology. Studies have demonstrated laminar antegrade pulmonary arteries flow in late diastole as a marker of reduce right ventricles diastolic compliance.[2] The restrictive pattern after TOF repair is not rare, but what was unusual was the pandiastolic antegrade flow in our patient. This flow pattern demonstrates that mid-diastolic RV pressure exceeded main PA pressure, which we believe was essentially due to the development of restrictive physiology of the myocardium. Pulmonary regurgitation is now recognized as one of the most important factor in the late follow-up of such patients and it is related to RV dilatation and increases the incidence of arrhythmia and possible sudden death.[3-6] Hence, there is the emerging importance of RV diastolic events for long-term outcome of TOF repair. This phenomenon of antegrade diastolic flow on Doppler that starts before atrial systole that is, early diastole has not been described in previously. After the repeat surgery diastolic antegrade flow completely disappeared. One possible explanation of the disappearance of the flow pattern after surgery could be because of the placement of pulmonary valve and hence increase in mid-diastolic pressures in main PA, also the disappearance of small pulmonary regurgitation present initially in restrictive right ventricle, decreased the RV diastolic pressures. The pulmonary regurgitation was not an isolated process that was responsible for the diastolic dysfunction preoperatively, rather the compensatory hypertrophy to severe obstruction too contributed to diastolic dysfunction. It was the result of these factors that a protective effect of restrictive physiology was operational in this case making the patient asymptomatic and resulting pandiastolic antegrade flow. This kind of early diastolic antegrade flow across PA needs validation

by means of further studies and case series.

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Conflicts of interest

There are no conflicts of interest.

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