Lack of "ventricular interdependence" in constrictive pericarditis and atrial septal defect

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ABSTRACT

Ventricular interdependence, i.e., reciprocal variations in the left and right ventricle pressures with respiration, is a hallmark of the hemodynamic diagnosis of constrictive pericarditis (CP). Similarly, respiratory variations in the mitral and tricuspid valve Doppler inflow velocities on echocardiogram are very helpful in the diagnosis of CP. We document the absence of such variations in a patient with CP and associated atrial septal defect. It is important to be mindful of this intuitively obvious fact; otherwise, the diagnosis of CP might be missed.

Keywords: Echocardiography, hemodynamics, pericardium

INTRODUCTION

Constrictive pericarditis (CP) is an important treatable cause of heart failure. In real-world scenarios, the diagnosis is often missed and the patient may be treated as having liver disease, heart failure with preserved ejection fraction, or other disorders. A comprehensive clinical evaluation and careful echocardiography are very helpful in the correct diagnosis of CP.[1,2] Presence of ventricular interdependence is a very important hemodynamic finding in patients with CP that distinguishes it from restrictive cardiomyopathy.[3] The variation in mitral valve (MV) and tricuspid valve (TV) inflow velocity (peak E-wave velocities) with respiration are very helpful in the diagnosis of pericardial restrain in CP, and also in tamponade. As reported by Hatle *et al.*,[1] and Oh *et al.*,[2] such variations are reliable in distinguishing CP from restrictive cardiomyopathy. We report a child with CP and associated atrial septal defect (ASD) in whom respiratory variations in the Doppler signals across the mitral valve (MV) and TV inflow were absent. On cardiac catheterization, there was no evidence of ventricular interdependence. The diagnosis of CP was confirmed at the operation. This intuitively obvious lack of ventricular dependence in patients of CP with ASD has not been well documented in the literature.

CASE REPORT

A 14-year-old boy presented with breathlessness and abdominal distention of 3 months. He was treated for tubercular pleural and pericardial effusions 2 years earlier. No pericardial drainage was done. Clinical examination revealed a raised jugular venous pressure with prominent V waves and a pan systolic murmur. The echocardiogram was remarkable for a large secundum ASD, normal ventricular functions, and moderate mitral regurgitation. However, no respiratory variations in the MV or TV Doppler inflow signals were seen. The TV

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Doppler is shown in Figure 1. The septal bounce was present on 2-dimensional (2D) imaging, but there was no septal shift visualized on 2-D echo [Supplementary Video 1]. The tissue Doppler myocardial velocities on the septal and lateral ventricular walls were normal. The lack of respiratory variation also persisted after a few days of diuretic use. The cardiac cath data were consistent with CP with a raised right atrial pressure (22 mmHg) and raised ventricular end-diastolic pressures (24–30 mm) in the right and left ventricles. The pulmonary artery pressures were mildly elevated (45 mmHg systolic). The calculated pulmonary/systemic flow ratio was 2.3:1 [Table 1]. There was no change in ventricular systolic or diastolic pressures with respiration [Figure 2]. The computed tomography scan showed a thickened pericardium. The patient was operated under cardiopulmonary bypass. Pericardiectomy, ASD closure, and MV repair were done. He showed marked improvement and was symptom-free on 6 months follow-up.

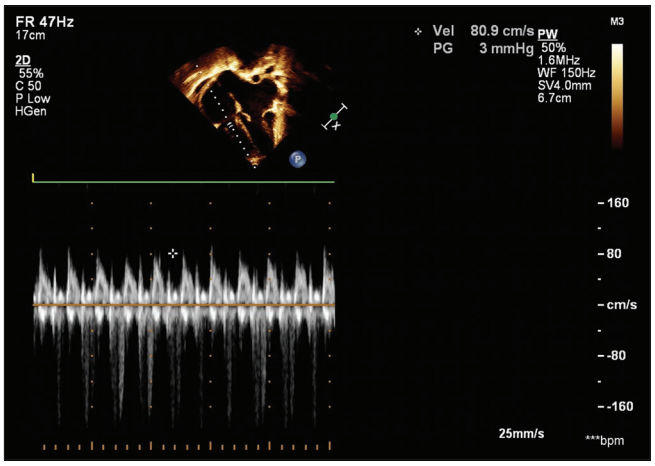

Figure 1: Lack of respiratory variation in tricuspid valve Doppler

DISCUSSION

The fortuitous occurrence of CP in a patient with ASD is rare but may provide insights into some pathophysiologic concepts. The clinical diagnosis is often missed in such patients.[4-6] The purpose of this brief communication is to highlight the fact that the ventricular interdependence on cardiac catheterization and Doppler variations in the MV and TV inflow velocities on echocardiography may be absent in a patient with CP when there is an associated ASD. The presence of septal bounce on echo [Supplementary Video 1] and clinical congestive heart failure in the patient was suspicious of some additional problems with ASD. There were no respiratory variations in mitral or tricuspid inflow velocities. The septal "bounce" is often the initial starting point for the suspicion of CP, and was present, but it is not specific to CP.[7]

The quantum of changes with inspiration and expiration suggesting pericardial restrain might vary. About 30%–50% variations in peak E-wave velocities were reported in the initial observational series.[1,2] A large series from Mayo Clinic found as much as 14% variation might be

Table 1: Hemodynamic data of the patient

	Pressures (mmHg)	Oxygen saturation %
Superior vena cava		55
Right atrium	a-22, v-26 (22)	
Right ventricle	45/26	
Pulmonary artery	40/18 (27)	86
Left ventricle	110/25	
Aorta	102/90	98
Qp/Qs	2.5	
Cardiac index	2.8 (L/min/m^2)	
PVRI/SVRI	0.27/25	

PVRI: Pulmonary vascular resistance index, SVRI: Systemic vascular resistance index

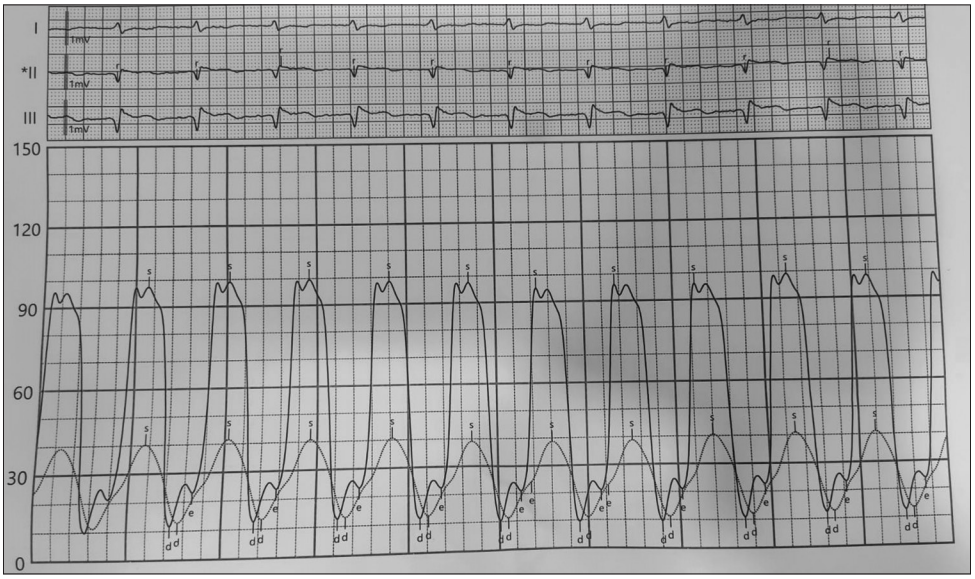

Figure 2: Simultaneous right and left ventricular pressures showing a lack of ventricular interdependence

the useful cutoff for the diagnosis of CP,[7] although generally, 25% variation in the MV peak E velocities and 40% variation in the TV peak E velocities are considered acceptable.[8]

Ventricular interdependence is seen in cardiac tamponade, CP, acute asthma, right ventricular volume overload, and positive pressure ventilation in the setting of pulmonary artery hypertension.[9] It is a hallmark of the hemodynamic diagnosis of CP and its differentiation from restrictive cardiomyopathy.[3] Ventricular interdependence might be absent in patients with ASD and CP is intuitively obvious, but if the clinical diagnosis is not suspected, such an absence might lead to misdiagnosis.

The presence of ASD in a patient with CP may be entirely missed on oximetry run as equalization of diastolic pressures in the intracardiac chambers due to CP would reduce the shunt. On the other hand, Doppler interrogation alone or reliance on ventricular interdependence alone in such a patient might miss CP, as shown in this case. The reason for the absence of variation seems clear, as the preload of the ventricles would remain reasonably fixed despite respiration owing to reciprocal changes in the systemic and pulmonary venous returns and the shunt from the ASD being balanced by the two atria. These factors account for the absence of variation in the ventricular pressures as well.

The absence of respiratory variations has also been shown when the preload is markedly increased, and the variation might be brought out after diuresis.[10] Associated restrictive cardiomyopathy (as in CP with radiation)[8] or cardiac arrhythmia might also be responsible for such an observation.

CONCLUSIONS

The ventricular interdependence on cardiac catheterization and respiratory variation in the MV and TV Doppler inflow velocities on echocardiography might be absent in patients with CP and associated ASD. The diagnosis of CP should not be excluded in a patient with ASD because of the lack of ventricular interdependence.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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