Abstract
A case of isolated right ventricular hypertrophic obstructive cardiomyopathy without left ventricular involvement is reported. This is a rare cause of isolated right ventricular strain and needs to be differentiated from ventricular septal defect and pulmonary valve stenosis.

Isolated Right Ventricular Hypertrophic Obstructive Cardiomyopathy
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Introduction
Hypertrophic cardiomyopathy usually involves left ventricle. Concomitant involvement of right ventricle occurs in some cases. Isolated involvement of right ventricle without involvement of left ventricle is rare.1,2

Case Report
A 24 year female presented with complaint of breathlessness on effort. She was told to have “Congenital heart disease” in early childhood. On examination there was no cyanosis or clubbing. Jugular venous pulse was normal. A grade 3/6 mild systolic murmur was audible in left third and second intercostal space. There was no ejection click and second sound was normally split with normal intensity of both components. Electrocardiograph revealed right axis deviation and right ventricular hypertrophy (Fig. 1).

Echocardiographic evaluation revealed gross hypertrophy of free wall of right ventricle with mild increase in thickness of inter ventricular septum (Fig. 2), Colour Doppler imaging revealed turbulence across right ventricular outflow tract with a peak gradient of 33mmHg (Fig.3). Pulmonary valve was normal and there was no additional gradient across it. Main and branch pulmonary arteries were normal and pulse Doppler flow of pulmonary artery was normal. There was no gradient

Fig. 1 : Electrocardiogram showing right axis deviation and right ventricular hypertrophy
Fig. 2 : M-Mode echocardiograph showing gross thickening of RV free wall (AW) & normal thickness of inter ventricular septum (IVS).
Fig. 4 : Showing absence of gradient across LV outflow tract (LVOT).
Fig. 5 : Showing normal mitral flow.
across left ventricular outflow tract (Fig. 4). Mitral valve flow (Fig. 5) and pulmonary vein flow were normal. Doppler tissue imaging of lateral tricuspid annulus showed Aa velocity more than Ea velocity.

Patient did not agree for further evaluation. She was advised beta-blockers which gave partial relief in symptoms.

Discussion

Isolated right ventricular hypertrophic obstructive cardiomyopathy presents as systolic murmur in left upper parasternal region with clinical and electrocardiographic evidence of right ventricular and right atrial strain. Absence of pulmonary valvular click and normal intensity and splitting of second sound differentiate it from pulmonary valve stenosis. Absence of left ventricular enlargement on clinical examination, inspiratory increase in the systolic murmur and absence of left ventricular enlargement on electrocardiograph differentiate it from ventricular septal defect. Echocardiography with colour flow imaging excludes other possibilities and documents hypertrophy of right ventricular outflow tract muscle with systolic gradient across the obstruction.

Like classical hypertrophic obstructive cardiomyopathy, patient may have partial relief of symptoms with beta-blockers. Non-responders may need surgical intervention.

References