

Mid-cavity Obstruction of Right Ventricle Without Any Other Congenital Anomaly

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A 7-day-old female infant born of full-term normal delivery was referred for evaluation of a systolic murmur in left lower parasternal region. Electrocardiogram (ECG) revealed P-pulmonale with right axis deviation and prominent RS configuration in lead V₁ (Fig. 1). Echocardiography revealed hypertrophy of right ventricle with a systolic turbulent jet between apex and main cavity of right ventricle (Fig. 2). Continuous wave Doppler evaluation of the jet revealed a systolic gradient of 65.4 mmHg (Fig. 3). Pulmonary valve was normal. Pulsed wave Doppler evaluation of pulmonary flow was normal with a peak velocity of 0.87 m/sec and peak gradient of 3 mmHg (Fig. 4). Tricuspid leaflets were normal with mild tricuspid regurgitation. Left ventricle, mitral valve flow, left ventricular outflow tract and aortic flow were normal. There was no other congenital anomaly.

Echocardiography findings of this case resemble left ventricular mid-cavity obstruction seen in some cases of hypertrophic cardiomyopathy. Isolated right ventricular hypertrophy with hypertrophied muscle bundles created gradient between apex and cavity of right ventricle. Such hypertrophied obstructive muscle bundles in the cavity of right ventricle are invariably associated with other congenital anomalies like ventricular septal defect, pulmonary valve stenosis, Ebstein malformation or tetralogy of Fallot. Our case could be a variant of hypertrophic cardiomyopathy. Parents did not agree for any other investigation, chromosome analysis or evaluation of other family members. Patient was first issue and parents denied any history of cardiac disease or sudden death in close relation.

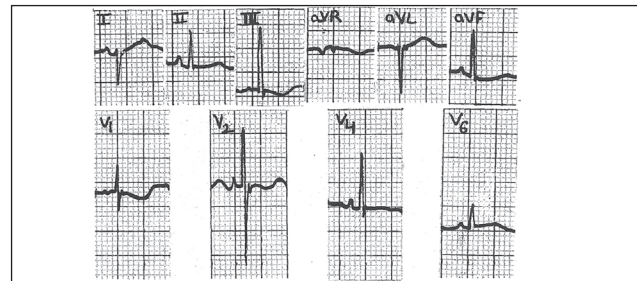


Figure 1. ECG showing right axis deviation, P-pulmonale and R/S configuration in lead V₁.

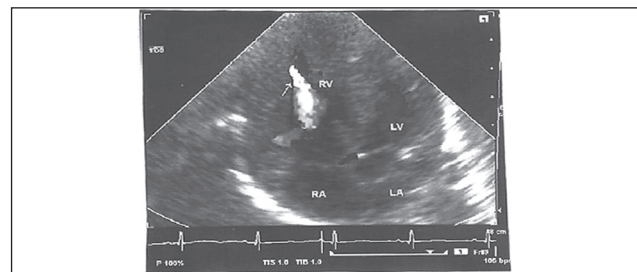


Figure 2. Color Doppler echocardiograph in apical four-chamber view showing turbulent jet coming from RV apex to RV cavity.

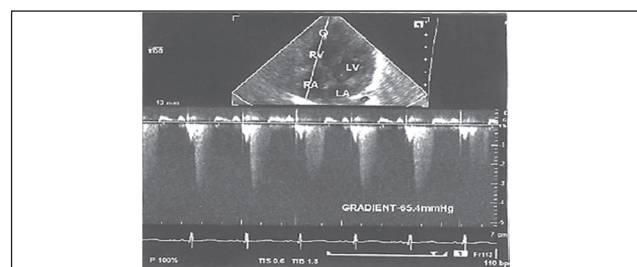


Figure 3. Continuous wave Doppler showing gradient across the turbulent flow.

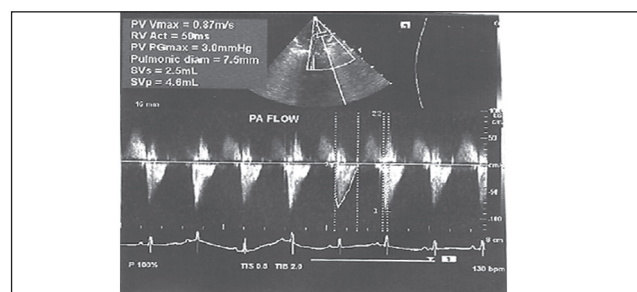


Figure 4. Pulsed Doppler evaluation of pulmonary artery flow.

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