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Case Report

Extremely large size VSD with pulmonary stenosis

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ABSTRACT

Defect in the ventricular septum with obstruction to right ventricular outflow tract encompass a wide anatomic, physiological & clinical spectrum. Large ventricular septal defects occur with pulmonary stenosis that varies from mild to severe to complete (pulmonary atresia). Very large VSD (size 6.4cm, in our case) with severe PS is a rare CHD & without surgical correction only 10% patients can survive beyond 20 year of age. With the help of noninvasive investigation (echocardiography) we can diagnose CHD very easily.

Keywords: CHD, VSD, PS, TOF

INTRODUCTION

Two-dimensional echocardiogram with color flow imaging & spectral doppler interrogation in VSD with PS generally demonstrate an infundibular or pulmonary stenosis gradient along with interventricular communication. Among the most prevalent cardiac malformations, defects of the ventricular septum occur commonly, both as isolated anomalies & in combination with other anomalies. VSD with PS is the most common form of cyanotic congenital heart disease after 1 year of age, with an incidence approaching 10% of all congenital heart diseases.

CASE REPORT

A 30 year old male patient came to medicine outpatient department, with history of shortness of breath since last 2 year & cyanosis. Clinical examination revealed moderately build, moderately nourished, grade II clubbing, pulse 90/minute, regular, hypervolemic, no radio femoral delay, all peripheral pulse palpable, blood pressure 120/70 mmHg in supine position, in right upper limb, JVP not raised, apical impulse palpable in 5 intercostal space, medial to mid clavicular line, forcible in character, on auscultation S1 & S2 heard, an ejection systolic murmur heard in left 3 & 4 intercostal space, increase on respiration. ECG shows (Figure 1) prominent "R" wave in V1 & V2 chest lead and poor progression of "R" wave from V4 to V6 chest lead. X-ray chest PA view shows normal cardiac silhouette, normal costophrenic angle.

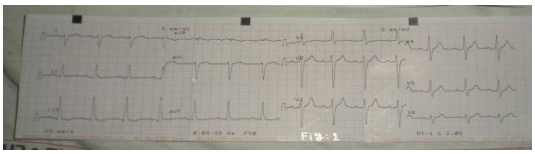

Figure 1: ECG shows, prominent "R" wave in V1 & V2 chest lead and poor progression of "R" wave from V4 to V6 lead.

TTE (Trans Thoracic Echocardiography) revealed - Situs solitus, Visceroatrial, Atrioventriculo concordance present. IVC drains in to RA & pulmonary veins draining into LA. Subcostal & Apical 4 chamber view show IAS intact. Two separate normal atria present, both atrioventricular valves anatomically appear normal & opening to ventricle. Apical 4 chamber view (Figure 2) and parasternal long axis views shows large size non-restrictive VSD (6.4 cm in size) present, basal & mid IVS completely absent, only small amount of apical IVS tissue is remaining (looking like rudimentary IVS). There is no rudimentary chamber. RV appears hypertrophied (Figure 2). Parasternal short axis view at aortic valve level show normal, trileaflet aortic valve (Figure 3) and

thickened pulmonary valve. Continuous wave doppler at pulmonary valve show severe pulmonary stenosis (Figure 4), peak flow velocity across PV is 5.50m/sec, peak pressure gradient (PPG) is 121 mmHg & Mean pressure gradient (MPG) is 71 mmHg.

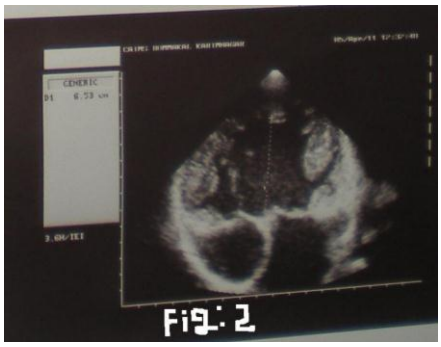

Figure 2: Apical 4 chamber view in systole shows, a very large size VSD, intact IAS, two separate AV valves and hypertrophied right ventricle.

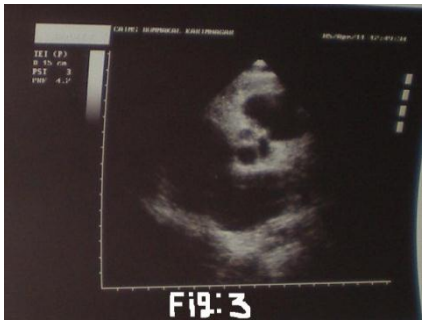

Figure 3: Parasternal short axis view at aortic valve level shows, normal trileaflet aortic valve.

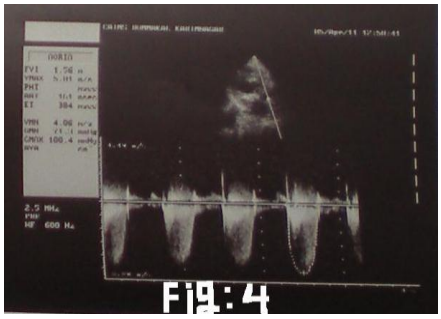

Figure 4: Continuous wave doppler at pulmonary valve show severe pulmonary stenosis.

DISCUSSION

The most common congenital heart disease (CHD) is VSD, but such an extreme large VSD is very rare CHD. One of the most common cyanotic adult CHD is TOF; very large VSD with PS is rare congenital heart disease. Without surgical correction only 10% of patients can survive beyond 20 years of age.[1-3] Some believe VSD closure is beneficial at any pulmonary artery pressure or resistance.[4-6] Fenestrated VSD patch closure (Fenestrations act like safety exits in case of acute raise in pulmonary artery pressure has been tried). Our 30 years old male was labeled inoperable and he is on medical management. Other mode of treatment is heart lung transplantation.

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