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 and clinical spectrum. Large ventricular septal defects occur with pulmonary stenosis that varies from mild to severe to complete (pulmonary atresia). Very large VSD (size 6.4 cm, in our case) with severe PS is a rare CHD and without surgical correction only 10% patient can survive beyond 20 year of age. With the help of noninvasive investigation (Echocardiography) we can diagnose CHD very easily.

Introduction
Two-dimensional echocardiogram with colour flow imaging and 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 and 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 out patient department, with history of shortness of breath since last 2 year and cyanosis. Clinical examination revealed moderately build, moderately nourished, grade II clubbing, Pulse 90/minute, regular, hypervolumic, no radio femoral delay, all peripheral pulse palpable, BP120/70 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 and S2 heard, an ejection systolic

Fig. 1: ECG shows, prominent “R” wave in V1 and V2 chest lead and poor progression of “R” wave from V4 to V6 lead

Fig. 2: Apical 4 chamber view in systole shows, a very large size VSD, intact IAS, two separate AV valves and hypertrophied Right ventricle
murmur heard in left 3 and 4 intercostal space, increase on respiration. ECG show (Figure 1) prominent “R” wave in V1 and 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.

- TTE (Trans Thoracic Echocardiography) revealed - Situs solitus, visceroatrial, atrioventriculo concordance present. IVC drains in to RA and pulmonary veins draining into LA. Subcostal and apical 4 chamber view show IAS intact. Two separate normal atria present, both atrioventricular valves anatomically appear normal and opening to ventricle. AP4C (Figure 2) and Parasternal long axis view shows large size non restrictive VSD (6.4 cm in size) present, basal and mid IVS completely absent, only small amount of apical IVS tissue is remaining (looking like rudimentary IVS). There is no rudimentary chamber. RV appear 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 5.50 m/sec, Peak pressure gradient (PPG) is 121 mmHg and Mean pressure gradient (MPG) is 71 mmHg.

Discussion

- Most common congenital heart diseases (CHD) is VSD, but such an extreme large VSD is very rare CHD

Treatment

Some believe VSD closure is beneficial at any pulmonary artery pressure or resistance.

Fenestrated VSD patch closure (Fenestrations act like safety exits in case of acute raise in pulmonary artery pressure has been tried). Our 30 year old male was labelled inoperable and he is on medical management. Other mode of treatment is heart lung transplantation.

References

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