Objective
Abnormal fistulous connection between right ventricular cavity and coronary artery is a well-known phenomenon in a subset of patients with pulmonary atresia and intact ventricular septum (PA-IVS). Such Ventriculo Coronary Arterial Connections (VCACs) has significant impact on surgical outcome of these patients. The morphogenesis of VCAC has been a matter of debate. This abstract aims to illustrate the missing link in the morphogenesis theory of ventriculo-coronary artery connections seen in a subset of pulmonary atresia - intact ventricular septum (PA/IVS) spectrum.

Methods
19-year-old primigravida referred for second opinion for abnormal cardiac views at 21 weeks and 2 days gestation to The Fetal Clinic, Pondicherry. Fetal evaluation revealed a normally grown fetus (EFW at 12th centile for GA) and no extracardiac anomalies. Detailed fetal echocardiography revealed normally arranged atria with normal systemic and pulmonary venous connections. Atroventricular connections were concordant with small tricuspid valve annulus (z score -1.68) and hypoplastic Right Ventricle (RV diastolic diameter z score -4.36) (figures 1 and 2). The RV morphology was bipartite with a poorly developed infundibulum that was difficult to image in B mode (figure 3). Forward flow with aliasing (figure 4) was demonstrable across a narrow pulmonary valve (pulmonary valve annulus z score -4.06, Vmax across pulmonary valve 107.64 cm/s, Peak Pressure Gradient 4.63 mmHg). The main pulmonary artery was of small caliber (2 mm, z score – 4.61) while the branch pulmonary arteries were of normal caliber (RPA z score -0.52; LPA z score -0.54). There was an abnormal fistulous connection between the right ventricle and the right coronary artery along the RV free wall, showing aliasing bidirectional pulsatile flow (figures 5 – 7). (Cardiac images could not be uploaded).

Results
The cardiac diagnosis in the fetus was: Pulmonary Stenosis, bipartite and hypoplastic Right Ventricle with abnormal Ventriculo-Coronary Arterial Connection.

Conclusion
Discussion: The morphogenesis of VCAC has been a matter of debate over decades. Initial researchers suggested VCAC to be secondary to intraventricular hypertension in the setting of primary pulmonary valvar atresia and intact septum. Subsequently, this notion has been challenged. VCAC may be the primary defect with consequent flow – dependent pulmonary atresia as suggested by Chaoui et al. The proposed morphogenetic pathway involves abnormal formation of right ventricle resulting in fistulous connection between the cavity lined by endocardium and the developing coronary arteries on the epicardium. The nascent placental circuit is a high resistance circulation and thereby imposes a high afterload to the developing right ventricle. The abnormal VCAC provides a low resistance pathway for RV decompression. This shunting decreases the forward flow across the normally formed pulmonary valves leading sequentially to pulmonary stenosis and the atresia. The missing link in this theory is the demonstration of VCAC in the presence of pulmonary stenosis and intact ventricular septum. Our case provides this missing link and thereby strengthens the theory of VCAC being the primary lesion.