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Congenital Mitral Atresia: A Rare Anomaly Diagnosed in Fetal Echocardiography: A Case Study.

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ABSTRACT

We describe a case of Mitral atresia associated with a ventricular septal defect (VSD) and hypo plastic left ventricle detected in fetal echocardiography performed in a 27+ weeks of gestation, a primi gravida woman. Deviating from the finding of hypoplastic left heart syndrome the present case had normal aortic root and ascending aorta. The mother of the affected fetus was apparently normal with no specific risk factors.
Keywords: Mitral atresia, Anomaly, Ventricular septal defect, Echocardiography

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BACKGROUND

Mitral atresia is a rare congenital heart anomaly in its isolated form, whereas this often present as a part of hypo plastic left heart syndrome[1] .Literature gives brief description of certain cases of mitral atresia and its associations including patent ductus arteriosus, transposition of great arteries, co-arctation of aorta, pulmonary stenosis, shunt defects, chamber and valvular anomalies, cor triatriatum, and also hypoplastic pulmonary arteries[2] .But all these case were identified in neonates/ infants. There are limited data on diagnosis of mitral atresia in antenatal scan- fetal echocardiography. The present case was diagnosed in fetus with gestational age of 27+ weeks. The fetal echo showed situs solitus, levocardia, atretic mitral valve, with relatively small sized left ventricle filling via 4mm small muscular ventricular septal defect and normal sized aorta.

Case Presentation

27 year old pregnant woman was referred for routine fetal echocardiographic scan at 27+ weeks of gestation which revealed Mitral valve atresia associated with VSD and hypoplastic LV, However 2D and Doppler echo showed normal ante grade flow across aortic valve with well-developed aortic root. The parents were counselled about the case and associated risks, hence advised for the delivery in a center where well established neonatal cardiac surgery care is present, however the parents were not willing for the same. The woman underwent vacuum assisted vaginal delivery at 39 weeks since there was fetal distress in second stage of labour. The neonate was relatively stable, started with phototherapy on day 3 in view of high TCB. TB, DB and TSH on day 5 were within normal range. Fetal echo findings were confirmed on neonatal echo screening, and showed mitral atresia with 3mm small ASD, 4mm apical muscular VSD, additional small sub-aortic VSD, well developed but relatively small sized left ventricle and 2mm PDA.

Investigations

The neonate was under close monitoring and referred for neonatal cardio thoracic surgeon. Since the spouse was not willing for the surgery, patient was on follow up. The infant later hospitalized in 45 days with the symptom of fever and noisy breathing and persistent cyanosis as observed by its parents. Chest X-ray revealed bilateral bronchopneumonia with increased pulmonary vascular markings. Repeat echo showed Mitral atresia, 2mm restricted inter-atrial communication, 5mm muscular VSD, mild TR, Severe PAH, 2mm PDA.

Treatment

Infant was kept under Hood box oxygen at FiO2 40% and started on IV antibiotics (Ampicillin, amikacin), given for a total of 6 days. Patient's party consulted a pediatric cardio thoracic surgeon who advised staged surgical correction with guarded prognosis with suggestions, which would not carry good long term prognosis, in view of dominant RV, and tricuspid valve as the systemic AV valve. Patient had sudden cardiac death in home at the age of 8months.

DISCUSSION

Literature describes certain cases with mitral atresia and its association due to its rare occurrence. Mitral atresia could be associated with multiple anomalies as seen in literature, but reports on fetal diagnosis of the same is limited [2]. The present case had isolated mitral atresia associated with VSD, relatively small sized left ventricle well developed aorta and was detected in fetal echocardiography. Since the gestational age was above the limit for advisable MTP, the subject had to carry the pregnancy. After birth the neonatal screening confirmed the diagnosis which was then followed up for 5 months. Similar case was published by Aliyu et al.[3], which described multiple shunt lesion and hypo plastic LV with mitral atresia in a 5 month old infant.

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Diagram 1: Fetal Echocardiography of apical four chamber view showing muscular VSD, small LV and large RV

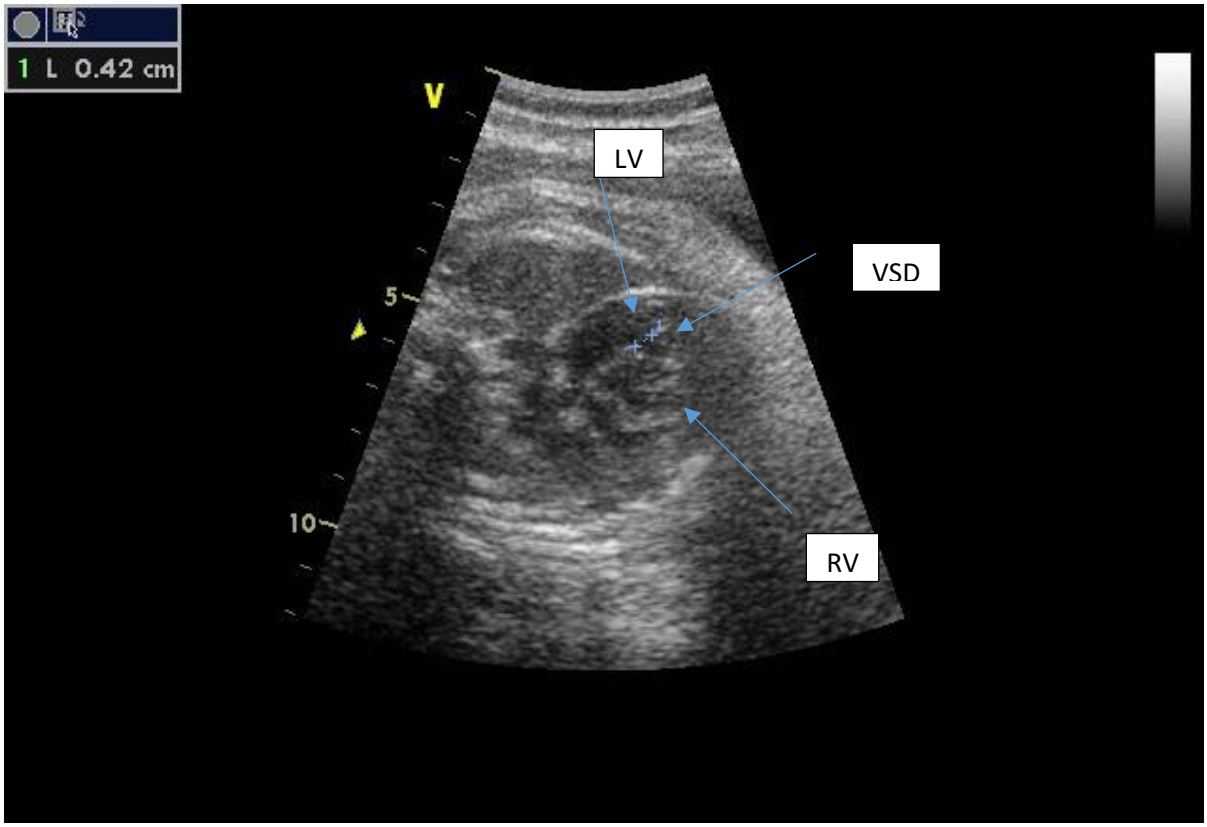

Diagram 2: Neonatal Echocardiography of apical four chamber view showing small muscular VSD, small LV and large RV and atretic mitral valve

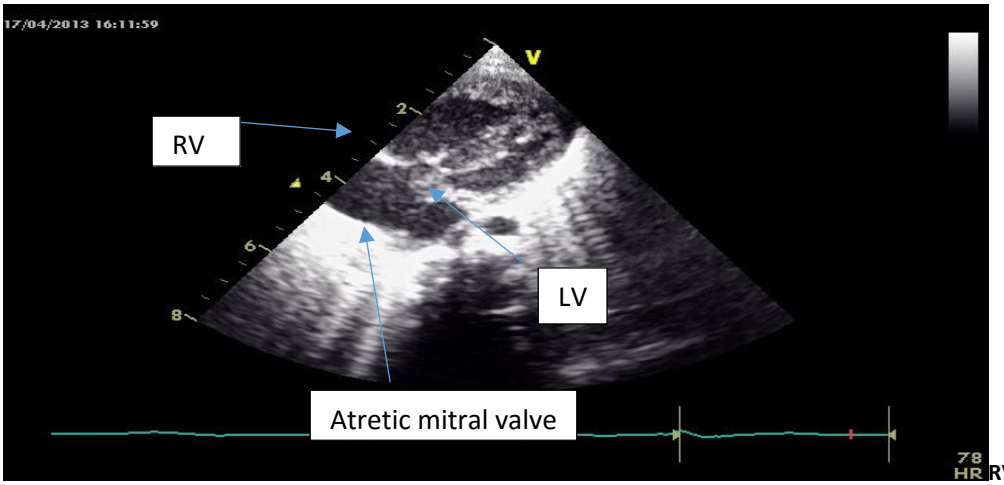

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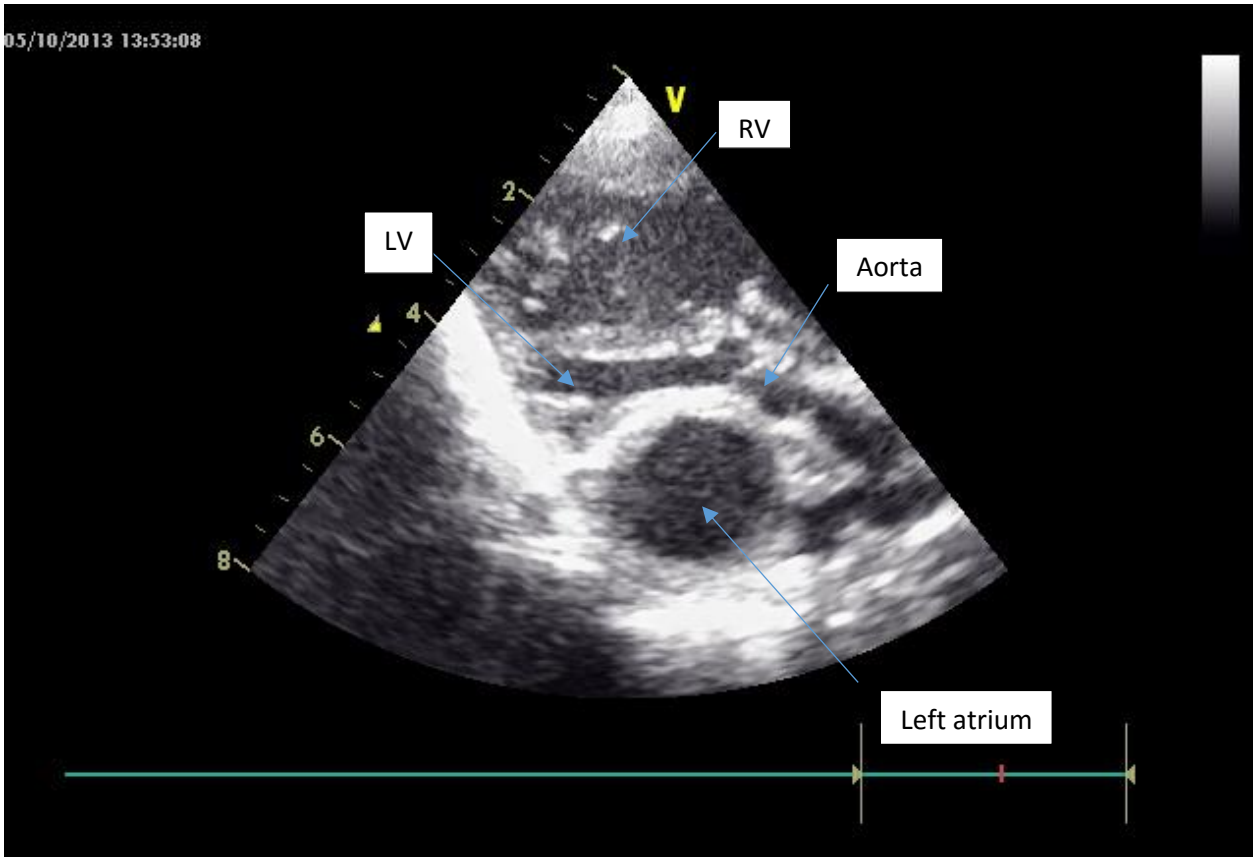
Diagram 3: PLAX view depicting mitral atresia



CONCLUSION

The congenital mitral atresia is a rare anomaly that could be associated with wide range of associated cardiac anomalies which could not be predicted by mother's risk status. The diagnosis of the same by early screening fetal echocardiography would prevent adverse outcome on the patient's course in pregnancy.

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