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PRENATAL DIAGNOSIS OF AORTO PULMONARY WINDOW BY FETAL ECHOCARDIOGRAPHY

Radio-Diagnosis

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ABSTRACT

Fetal aortopulmonary window is a rare fetal cardiac anomaly which shows a direct communication between pulmonary artery and aorta just distal to semilunar valve . It accounts for 0.2 to 0.6 % of all congenital cardiac defects1 Fetal Echocardiography is the gold standard diagnostic tool for detection of Aorto pulmonary window. We present 3 cases diagnosed antenataly in anomaly scan . In all of our cases with normal cardiac connections, three-vessel view is sufficient to make the diagnosis. However according to available literature many times additional views are required. An APW allows blood to flow from the aorta into the pulmonary artery, causing too much blood to flow through the lungs. Early diagnosis and intervention with closure of defect improves post natal outcomes significantly thus reducing mortality associated with it.

KEYWORDS

Aorto pulmonary septal defect, Transposition of great arteries, Arterial duct, Atrioventricular – ventriculoatrial concordance

INTRODUCTION :

The communication between pulmonary artery and aorta can be seen on routine second trimester level 2 scan with standard 3 vessel view However, when there are associated abnormalities affecting the spatial orientation of the great arteries, the diagnosis of an APW can be challenging and overlooked even with specialist fetal echocardiography . However there are many incidences which showed aorto pulmonary septal defect associated with other abnormalities (Kutsche and Van Mierop reported additional malformations in 52% of a series of 188 cases[3]) which further affected the spatial orientation of great arteries thus making its diagnosis a challenging task even for a specialist fetal echo cardiologist.

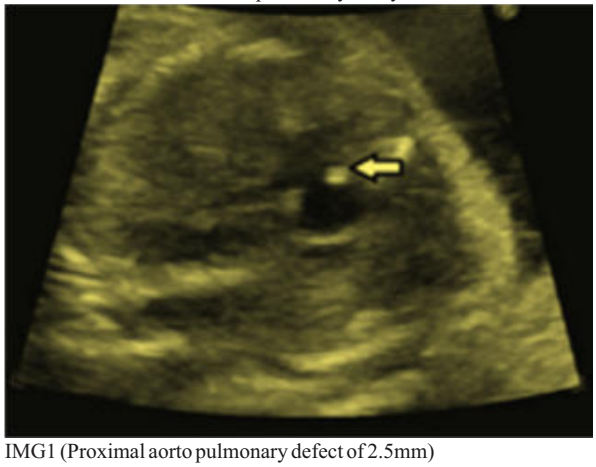
Richardson *et al*.[2] classified APW into three types

- Type 1, there is a simple defect located between the aorta and the main pulmonary artery, immediately above the sinuses of Valsalva.(most frequent form[2])
- Type 2, the defect is located more distally between the ascending aorta and the pulmonary trunk with extension into the origin of the right pulmonary artery.
- Type 3, there is anomalous origin of the right pulmonary artery from the ascending aorta.

We present 3 cases diagnosed with aorto pulmonary septal defect diagnosed antenatally in level 2 scan (2nd trimester scan) along with post natal confirmation.

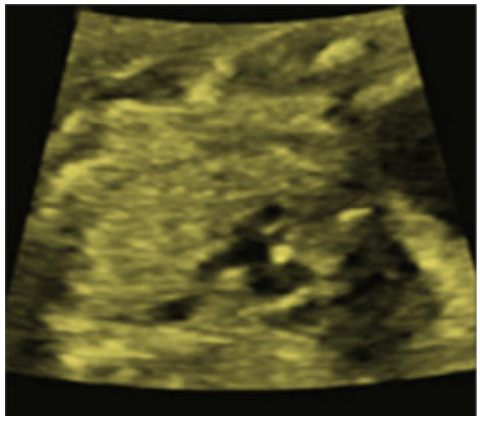
CASE 1(IMG1,2,3): The patient came in at 21 weeks for Target scan referred from primary center . The findings were :

There was a proximal aorto pulmonary septal defected measuring 2.5mm . The margins were hyperechoic as seen in the cases of ventricular septal defect . The arterial duct was tapering and flow across the defect was from the pulmonary artery to the aorta .

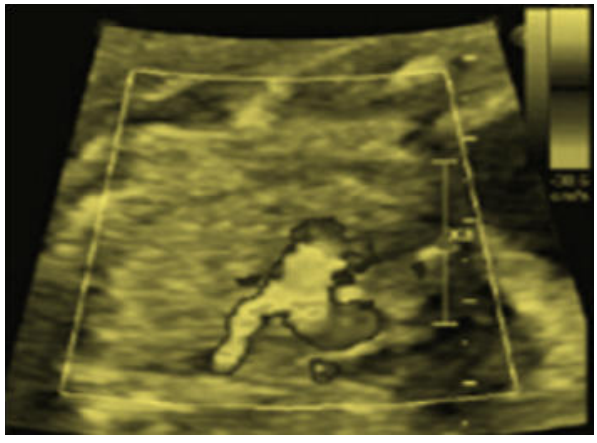


IMG1 (Proximal aorto pulmonary defect of 2.5mm)

- The arterial duct was tapering and smaller than the aortic arch. There was a proximal APW of 2.5 mm
- The margins of defect were hyperechoic as seen with ventricular septal defect (VSD). Along with the arterial duct, flow across the APW was from the pulmonary artery to the aorta.
- Aorta to pulmonary artery ratio was : 1.3 : 1 (i.e. aorta was larger in size)
- There was no flow acceleration across the semilunar valve and all other valves showed no obvious abnormality at the time of scan .
- Normal Atrio-ventricular and ventriculo – arterial concordance was noted.



IMG2 (abnormal three vessel view)



IMG3(color mode with 3 vessel view)

Post Natal Follow Up :

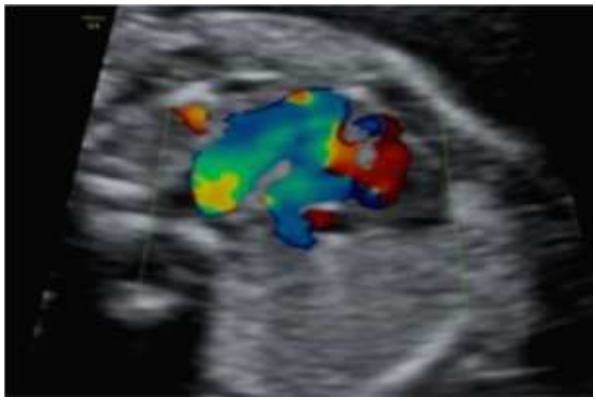
An emergency cesarean section was conducted at 32 weeks of

gestation. At birth, the male neonate, weighing 1.37 kg, presented with significant floppiness and severe respiratory distress, necessitating invasive ventilation. Postnatal two-dimensional echocardiography was carried out on the first and twenty-first day after birth. The echocardiogram revealed a large proximal atrioventricular septal defect measuring 7 mm, accompanied by a left-to-right shunt. A small arterial duct was observed on day 1 but had closed by the follow-up visit. Additionally, a small perimembranous ventricular septal defect measuring 2 mm was noted, also exhibiting a left-to-right shunt. No other cardiac or extracardiac structural anomalies were identified.

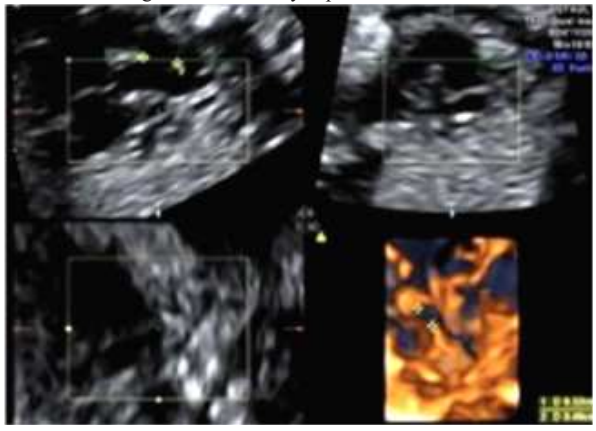
CASE 2(IMG 4,5,6) : Another 23 year old woman came in at 23 week of gestation

Findings :

- Normally positioned and sized heart with atrioventricular and ventriculoarterial concordance was seen .
- Aortopulmonary septal defect(IMG4) was noted in short axis view just above semilunar valves . blood flow was noted from right to left .
- Other findings include fetal micromelia and polyhydramnios .



IMG 4 - Showing Aorto Pulmonary Septal Defect On Color Mode



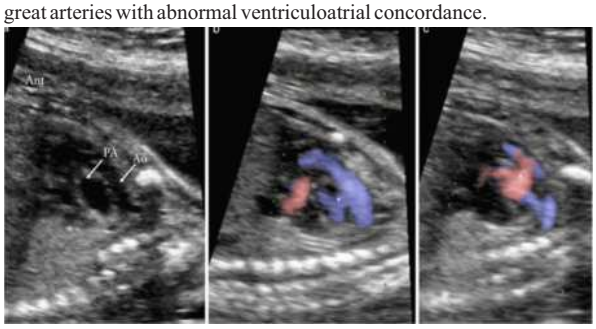
IMG 5-gray Scale Images Showing Defect

IMG 6 (POST NATAL)

Post Natal Follow Up :
800 gram male infant was delivered at 27 week of gestation through C section . Diagnosis of type 1 aorto pulmonary window was confirmed by post natal echocardiography.

CASE 3(IMG 7,8,9) : 28 year old women was referred due to abnormal 3 vessel view during a scan at primary centre .First trimester NT was >95th percentile .

Fetal echo was performed with following findings :
Abdominal situs solitus with atrioventricular concordance and normal intra cardiac anatomy. In parasagittal plane, abnormal position of great vessels close to diaphragm was seen due to complete transposition of great arteries with abnormal ventriculoatrial concordance.



IMG 7 **IMG 8** **IMG 9**

(With B-mode,APW could not be shown with certainty) (With color Doppler APW can be seen between Aorta and pulmonary artery)

Post Natal Findings :
The neonate underwent an arterial switch operation 7 days after delivery, during which the APW was identified and repaired.

The APW was identified retrospectively on sagittal views (IMG8) but could not be seen on the three-vessel view due to the abnormal relationship of the great arteries.

CONCLUSION:

Timely closure of the defect is essential to prevent the progression to irreversible pulmonary vascular obstructive disease. In the absence of surgical intervention, the mortality rate may reach 40–50% within the first year of life. The overall prognosis of aortopulmonary window (APW) is primarily influenced by the presence of associated anomalies and the development of pulmonary vascular disease.

As with ventricular septal defect (VSD), the diagnosis of APW in this case was supported by the detection of a hyper echoic specular reflection at the site of the blunted end of the aortopulmonary septum. This characteristic finding is commonly referred to as the “T sign,” analogous to that seen in VSD.

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