

Aortopulmonary window: Clues to fetal diagnosis

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ABSTRACT

Being a planar structure, fetal diagnosis of aorto-pulmonary window poses great challenge. A few echocardiographic signs can help to clinch the diagnosis.

Keywords: Antenatal diagnosis, congenital heart defect, fetal two-dimensional echocardiography, surgery for heart defect

Aortopulmonary window (APW) is a very rare congenital cardiac defect and it is still rarer to diagnose on the antenatal scan. Only a few case reports and case series have been published till date. There are no specific clues or guidelines which can help to make fetal diagnosis.^[1,2]

Present case report discusses APW suspected during second-trimester scan. It was conformed after birth. It also stresses upon possible clues that can help to identify this unique defect.

CASE

A 32-year-old primigravida was referred for fetal echo as the aorta appeared larger than the pulmonary artery during anomaly scan at 20 weeks. The cardiac scan was carried out at 21 weeks. The findings were as follows. Situs was solitus with levocardia. Atrio-ventricular and ventriculo - arterial connections were concordant. All four valves were normal and there was no flow acceleration across semilunar valves. Aorta was larger than pulmonary artery with a ratio of 1.2:1; however, Z scores were within normal range. The arterial duct was tapering and smaller than the aortic arch. There was a

proximal APW of 2.5 mm [Figures 1 and 2 and Video 1]. 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.

There was development of severe polyhydramnios during the late second and early third trimester. Emergency cesarean section was performed at 32 weeks. At birth, the male neonate (birth weight 1.37 kg) was floppy, had severe respiratory distress and required invasive ventilation. Postnatal two-dimensional echocardiography was performed on day 1 and day 21. There was a large proximal APW of 7 mm with left to right shunt. Small arterial duct was present on day 1, but it closed by on follow-up. There was a small 2 mm perimembranous VSD with left to right shunt. There was no other cardiac anomaly. There were no extracardiac structural anomalies.

There was very poor recovery in neurological function. Neonate underwent extensive evaluation, including ultrasound of skull, magnetic resonance imaging brain, metabolic screen etc. The genetic evaluation suggested X

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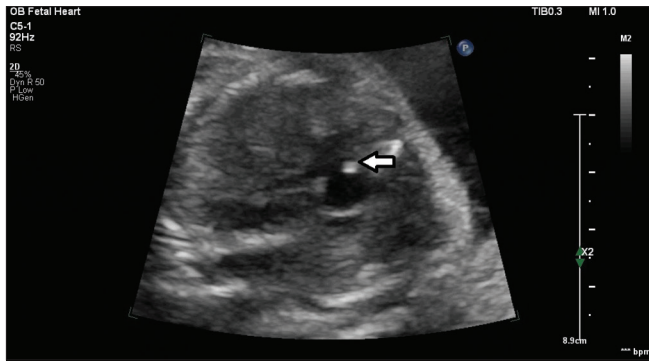


Figure 1: Proximal aorto pulmonary window. Note “T sign” with arrow

linked myotubular myopathy. The neonate could not be weaned from the ventilator as there was poor respiratory drive and succumbed at 37 days.

DISCUSSION

The tissue separating the aorta and pulmonary artery is a planar structure and has little thickness. Depending on the angle of sound waves, orientation of fetus, and origin of right pulmonary artery behind ascending aorta, this area is prone for a lot of artifacts. Furthermore, during the second trimester, pulmonary artery is larger than the aorta. If the reverse is present, there is a long list of differential diagnoses. Small pulmonary artery can be seen with tetralogy of Fallot, dysplastic pulmonary valve with pulmonary stenosis, etc. The dilated aorta may be associated with bicuspid aortic valve irrespective of aortic stenosis.^[1-4]

In fetal circulation, approximately 40% cardiac output is pumped into the ascending aorta and the remaining 60% into the pulmonary artery. As a result, the latter is larger than the former. Less than 10% of the cardiac output goes to the lung and the rest all in the pulmonary artery goes to the descending aorta through the arterial duct. APW may change these hemodynamics. Instead of the arterial duct, shunting happens through APW and more than usual blood goes through the aortic arch. Larger than the usual aortic arch and smaller arterial duct are the result of these changes in hemodynamics.^[4] This hypothesis needs to be justified in a larger case series. Just like VSD, diagnosis of APW, in this case, was aided by the presence of hyperechoic specular reflection caused by blunted end of aortopulmonary septum. It is often called as “T sign” of VSD.^[3] Obtaining a three-vessel view with aorta perpendicular to the ultrasound beam decreases the chances of artifacts. It is possible by

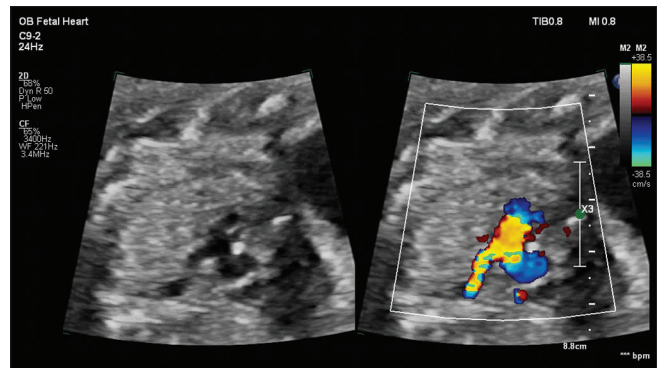


Figure 2: Three vessel view

obtaining image from either side of the thorax than from the front.^[1]

Polyhydromnios was secondary to X-linked myotubular myopathy affecting pharyngeal muscles. Above two conditions have never been reported together in literature. The association may be of chance.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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