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ORIGINAL ARTICLE

Unusual Combination of Anomalous Connection of the Left Pulmonary Veins and Aortic Arch Anomaly Presenting as Abnormal Three Vessel View in the 2nd Trimester US Scan

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Abstract Partial anomalous pulmonary venous drainage is often missed antenatally unless a complex congenital heart defect is associated. Likewise, isolated fetal coarctation of aorta has only a 60–70% chance to be diagnosed in the second trimester. The combination of these two conditions (PAPVC and COA), in the absence of situs abnormality or syndromic association is rare and not much reported in the literature. We are reporting a fetal heart which presented with apparent hypoplasia of the left ventricle and ascending aorta, as well as significant dilatation of the right atrium, right ventricle and pulmonary artery at 22 weeks of gestation. To begin with, we strongly suspected a left sided obstructive lesion until we noticed a 4th channel in the three vessel view (3VV) and dilated innominate vein in the absence of a dilated coronary sinus in the 4 chamber view. This led to a diagnosis of supracardiac PAPVC in combination with an arch anomaly. The pregnancy was continued up to 37–38 weeks when she underwent C-section for an obstetric indication. The baby underwent surgical intervention postnatally.

Keywords PAPVC · Coarctation of Aorta, Fetal Cardiac Anomaly

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Introduction

The combination of supracardiac partial anomalous pulmonary venous connection and coarctation of the aorta is an unconventional fetal diagnosis in the absence of situs anomalies and complex heart defects and is hardly reported in literature [1]. After birth blood volume in the pulmonary circulation increases and receives almost 50% of combined ventricular cardiac output (CVCO), unlike the 20% of CVCO received in fetal life. Abnormal post-left atrial space index [2] is one of the major clues for suspicion of a total anomalous pulmonary venous connection while in isolated PAPVC the findings are often subtle. Isolated fetal PAPVC is insignificant hemodynamically and remains mostly obscured before the birth.

On the other hand, coarctation of aorta (COA), narrowing of the distal aortic arch, constitutes 6–8% of all CHD. Its antenatal diagnosis is challenging and carries a high false positive rate. Prenatal diagnosis of coarctation of the aorta is critically important to avoid morbidity and mortality postnatally by facilitating 'in utero transfer' to a neonatal tertiary cardiac set-up. We report an antenatal diagnosis of PAPVC and forme-froste' coarctation of the aorta diagnosed at the 22nd antenatal week and its subsequent follow up.

Case Report

A 32 year second gravida with gestational diabetes mellitus (GDM), who was normoglycemic on metformin and had a history of one spontaneous abortion, presented to us with a normal anomaly scan done elsewhere. As per institutional protocol, she was advised a review fetal ultrasound and fetal echocardiography in view of GDM.

Cardiac evaluation showed no situs abnormality and segmental analysis revealed atrio-ventricular, ventriculo-arterial concordance. On eyeballing, the mitral valve (MV), left ventricle (LV), ascending aorta (AA) and transverse arch (TA), isthmus (ISTH) were apparently smaller when compared to the right side of the heart. The calculated fetal Z scores were lower but not abnormal for the predicted values for fetal gestational age (Table 1). Although there was a suggestion of a small posterior shelf at ISTH, Doppler velocities at ISTH, arch of aorta and ductal arch (DA) were normal. A small aorta almost the same size as the SVC in the 3VV and narrow aortic arch in the 3VT view prompted suspicion of coarctation of aorta and a sagittal view of the aortic arch (Fig. 1) confirmed a narrow isthmus. Additionally, the ratio of the aortic isthmus and ductal diameter less than one raised the suspicion of aortic arch abnormality [3]. There was no pericardial effusion, tricuspid regurgitation or cardiac dysfunction. The interatrial connection was non-obstructive and mitral and tricuspid inflow velocities were normal and biphasic. The possibility of a right sided lesion causing RV volume overload was explored. Increased distance between the LA and descending aorta (post LA space index) has been suggested as an important parameter for diagnosing TAPVC. The Post LA space index, in this case, was 1, falling short of a cut off value of 1.27 [2]. Besides, right sided pulmonary veins (PVs) were connected to the LA ruling out a TAPVC. Left sided PVs could not be profiled, initially. An additional venous channel was seen in the 3VV, to the left of the pulmonary artery (Fig. 2). However, dilated innominate (IV) in modified tracheal view (Fig. 3) and normal coronary sinus (CS) in 4 chamber view, suggested a pathology other than persistent left superior vena cava (L-SVC). On color Doppler also, this venous channel seemed like draining into the IV. Subsequently, with better fetal positioning, it was clear that the left PVs were not connected to the LA but were tributaries of the ascending 4th venous channel, suggesting a supracardiac PAPVC of left PVs. At this stage, diagnosis of a supracardiac PAPVC of the left PVs and 'forme-froste' CoA was proposed.

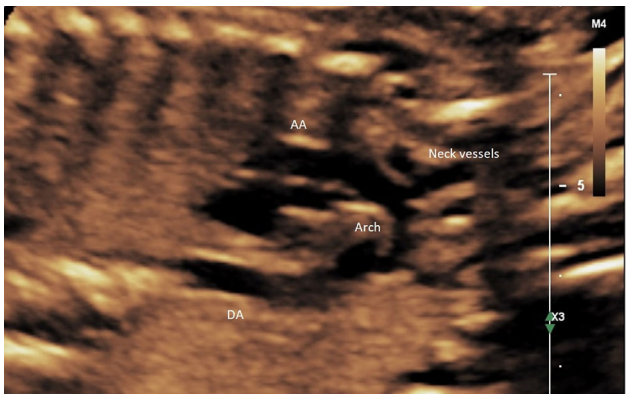

Fig. 1 Narrow aortic arch seen in sagittal view

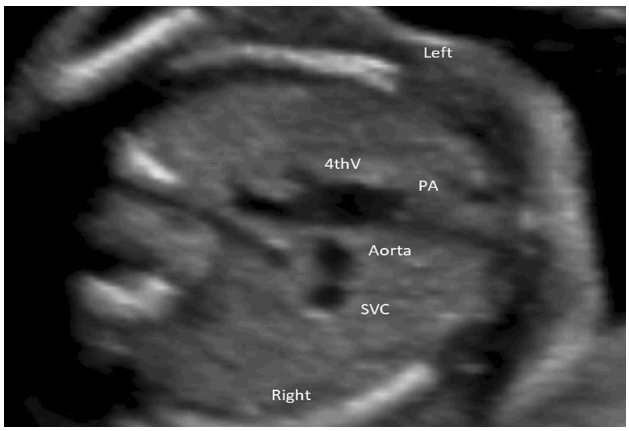

Fig. 2 Fourth vessel seen in three vessel view left to pulmonary artery

Table 1 .

Left heart	Z-score			Right heart	Z score		
	22 week	35 weeks	2nd day postnatal (38 weeks)		22 week	35 week	2nd day postnatal
MV	− 0.1	− 1.4	− 0.86	TV	+ 0.72	+ 0.9	+ 0.5
AV	− 0.69	− 1.4	− 2.07	PV	+ 2.17	+ 0.22	+ 0.77
AA	− 1.88	− 1.59	− 1.38	MPA	+ 2.1	+ 0.13	+ 1.2
Isthmus	− 1.6	− 2	− 3.1				
Distal arch			− 2.7				
LVIDD[a]			− 2.7				

MV mitral valve, *TV* tricuspid valve, *AV* aortic valve, *PV* pulmonary valve, *MPA* main pulmonary artery, *AA* ascending aorta, left ventricle
[a]diastolic dimension (M-mode)

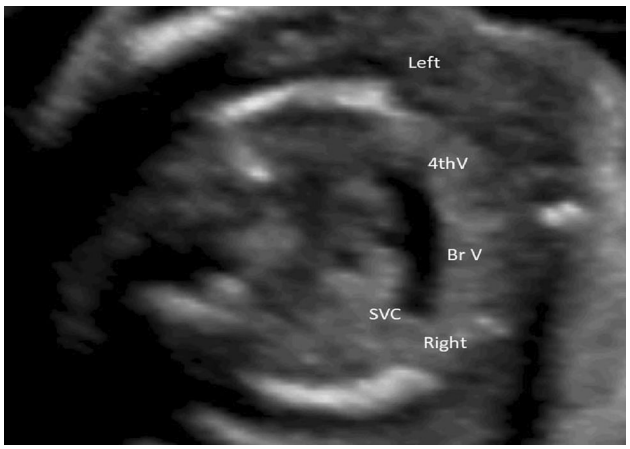

Fig. 3 Dilated brachiocephalic vein connecting vertical vein to SVC

A three-generation pedigree was constructed but no hereditary influence was detected. Couple-counselling was done about the possibility of manifestation of coarctation of aorta in the immediate postnatal period needing neonatal cardiac intervention. A close clinical and ultrasound guided follow-up was advised for timely detection of the onset of fetal compromise or fetal heart failure.

The antenatal follow up was uneventful. At 37.5 weeks, a C-section was done due to obstetric indications and mild fetal intrauterine growth retardation. A female child with a birth weight of 2.4 kg and normal apgar scores was delivered. On day 2, a first postnatal echocardiogram was done which showed an unobstructed anomalous connection of the left PVs to the IV, posterior shelf at isthmus and isthmus narrowing with no significant gradient. Juxta ductal PDA was shunting predominantly left to right. On day 4, expectedly, bilateral femoral arterial pulsations became feeble. The echocardiography was repeated which showed spontaneous closure of the ductus arteriosus, increased gradient at the isthmus (max gradient 18 mm Hg) with early diastolic spill. The baby was transferred to a neonatal cardiac surgery center where she underwent a computerized tomography (CT) angiogram (Fig. 4) and subsequent surgical repair of coarctation of the aorta. Postoperatively she did well. No attempt was made to address the PAPVC at this stage.

Discussion

PVs are relatively smaller in calibre as they handle less blood volume in the antenatal period. A diagnosis of TAPVC/PAPVC is difficult and usually done on 2 and 3 D echocardiography based on the following features: (1) bald left atrium (non-visualization of PV connections into LA) (Fig. 5); (2) abnormal 3VV or SVC/IVC dilatation; (3) icreased post-left atrial space index derived by dividing the distance between the posterior LA wall and descending aorta by the DA diameter; (4) RV/LV dimension ratio (> 1.5); (5) Abnormal TV/MV diameter ratio; (6) 3D spatiotemporal image correlation (STIC) fetal echocardiography [4].

Presence of a fourth vessel in the 3VV (Fig. 6) suggests either a persistent left superior vena cava or an ascending vertical vein (AVV) draining total or partial anomalous pulmonary veins. In our case, the fourth vessel was the vertical vein and points in favor were (1) Absence of dilated coronary sinus in 4 chamber view (2) Presence of prominent brachiocephalic bridging vein, and (3) Flow direction in the fourth vessel towards upper thorax (Fig. 7).

Fetal COA is an elusive and challenging diagnosis because of the difficult evaluation of isthmus anatomically in tracheal and sagittal view and erroneous assessment of Doppler flow velocities due to the presence of juxtaposed opening of the ductal arch (DA). The arch vessels must be located carefully to avoid confusion with the DA. Recently

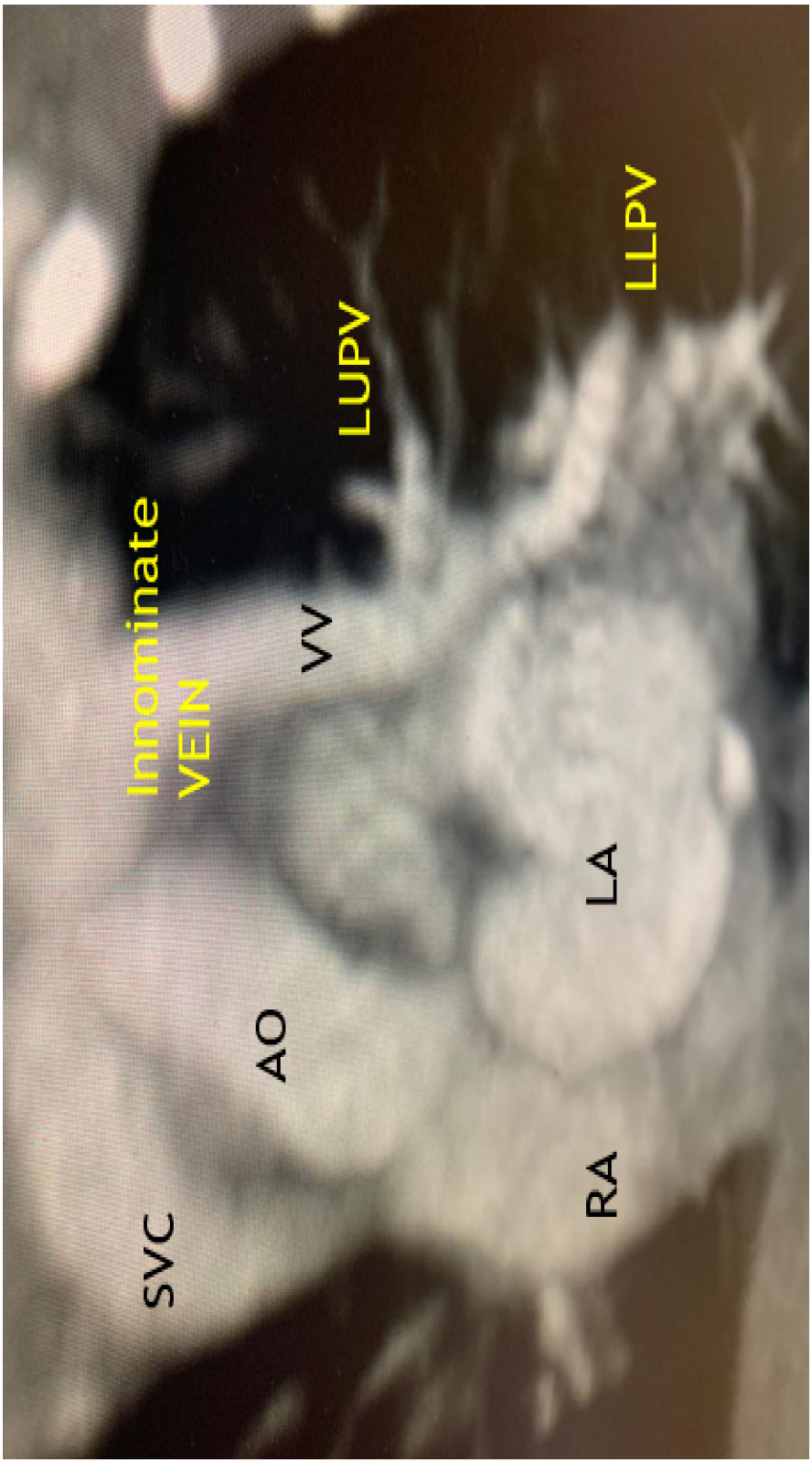

Fig. 4 CT angiogram (coronal view) ascending vertical vein (VV) draining left upper and lower veins (LUPV and LLPV) into innominate vein (IV)

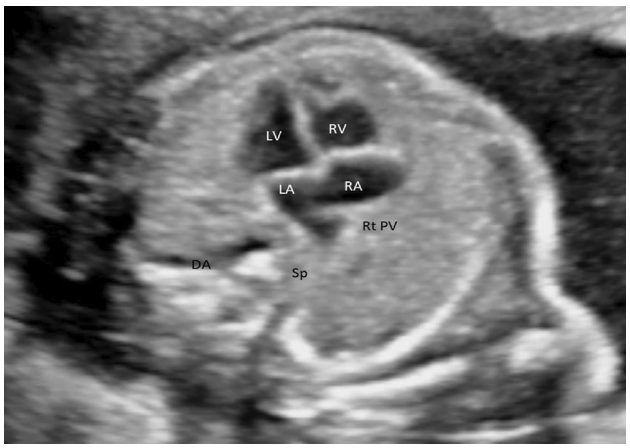

Fig. 5 Non visualization of connection of left pulmonary vein to left atrium, relatively increased retro atrial space

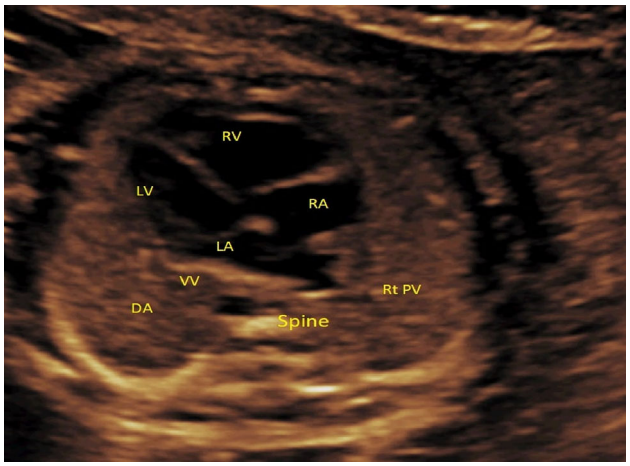

Fig. 6 Four chamber view of heart showing right pulmonary vein draning in to left atrium and vertical vein on left side

multiple parameters have been suggested for diagnostic accuracy. ISTH diameter, ductal diameter, isthmus/ductal ratio, z score of AA, AV, MV, distal AA, ISTH and arterial duct, the presence of a shelf and Doppler flow disturbance at the ISTH, altered flow across the inter-atrial connection, altered RV/LV ratio, altered TV/MV ratio. Also 3-dimensional (3D) imaging (± STIC) is a convincing method, but it is not available at all the centers.

In our case a correct diagnosis could be made in the absence of most of the indirect parameters narrated above. We suspected Heart disease due to perceived RV/LV asymmetry [5] which was not significant on measurement. Additionally, we found a small AA and ISTH which has a 80% chance of having COA physiology. Therefore, on finding a clue, a careful exploration of anatomy led us to a rare diagnosis, which helped us in planning the post-natal management efficiently.

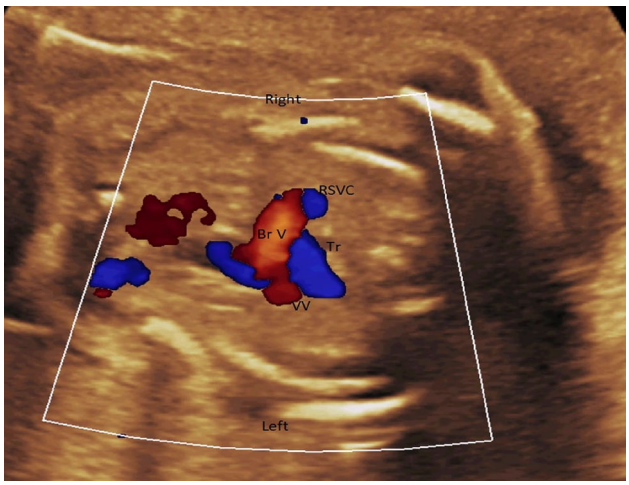

Fig. 7 Modified 3VT view in colour where vertical vein seen on left and SVC on right with opposite colour

Conclusion

Coexistent PAPVC and coarctation of the aorta are not diagnosed commonly in the antenatal period because of hemodynamically different fetal circulation. If we keep a high index of suspicion then an accurate diagnosis can be made, leading to appropriate postnatal management of the baby.

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Compliance with Ethical Standards

Conflict of interest The authors declare that there is no conflict of interest.

Ethical Statement The manuscript has not been submitted at any other place for simultaneous consideration and has not been published previously (Partially or completely. A single study is not split up into several parts. No data have been fabricated or manipulated to support our conclusion. No data, text, images or theories by others are presented. Co-author has adequately contributed to the work and manuscript has been approved by co author.

Informed Consent This is a retrospective study—"For this type of study, formal consent is not required".

References

1. Bajolle F, Zaffran S, Losay J, Ou P, Buckingham M, Bonnet D. Conotruncal defects associated with anomalous pulmonary venous connections. Arch Cardiovasc Dis. 2009;102:105–10.

2. Kawazu Y, Inamura N, Shiono N, Kanagawa N, Narita J, Hamamichi Y, et al. 'Post-LA space index' as a potential novel marker for the prenatal diagnosis of isolated total anomalous pulmonary venous connection. Ultrasound Obstet Gynecol. 2014;44:682–7.
3. Pasquini L, Mellander M, Seale A, Matsui H, Roughton M, Ho SY, Gardiner HM, et al. Z-Scores of the fetal aortic isthmus and duct: an aid to assessing arch hypoplasia. Ultrasound Obstet Gynecol. 2007;29(6):628–33.
4. Palladini D, Pistorio A, Wu LH, Meccariello G, Lei T, Tuo G, et al. Prenatal diagnosis of total and partial anomalous pulmonary venous connection: multicenter cohort study and meta-analysis. Ultrasound Obstet Gynecol. 2018;52:24–34.
5. Valsangiacomo ER, Hornberger LK, Barrea C, Smallhorn JF, Yoo SJ. Partial and total anomalous pulmonary venus connection in the fetus: two dimensional and Doppler echocardiographic findings. Ultrasound Obstet Gynecol. 2003;22:257–63.