Fetal aorto-pulmonary window: case series and review of the literature

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ABSTRACT

Aorto-pulmonary window is a rare congenital cardiac anomaly characterized by a communication between the aorta and the pulmonary artery above the semilunar valves. Prenatal diagnosis is rare. We report four fetuses with aorto-pulmonary window and review the relevant literature. Approximately half of the reported cases had additional cardiac defects; none had chromosomal abnormalities. In cases with normal cardiac connections, the diagnosis can be made prenatally on the standard three–vessel view as seen in two of our cases. In one fetus with complete transposition, the diagnosis was made retrospectively on sagittal views. In the remaining case the window was seen post-natally but could not be identified retrospectively due to the abnormal supero-inferior relationship of the ventricles and vessels.

CASE SERIES

Between January 1997 and July 2015, fetal echocardiography was performed in 11,727 fetuses in our tertiary centre for fetal cardiology. Among these, we identified four with a diagnosis of aorto-pulmonary window (APW), two of whom were diagnosed prenatally. In the other two, the diagnosis was made post-natally and could be seen retrospectively in one of these.

Case 1

Prenatal findings

A 33 year-old woman, gravida 3, para 1, was referred for fetal echocardiography at 34 weeks due to the presence of a cystic mass located behind the left atrium. Family history was unremarkable. First trimester nuchal translucency thickness was 1.6 mm. Fetal anatomical survey performed at 21 weeks of gestation was normal apart from the presence of an echo-poor lesion in the upper mediastinum, thought to represent an intra-thoracic cyst. As the pregnancy advanced and the mediastinal lesion persisted, she was referred for cardiac assessment. Fetal echocardiography showed abdominal situs solitus. The atrio-ventricular and ventriculo-arterial connections were concordant. The fetal heart was normally sited in the chest. At the level of the 4-chamber view, a relatively small cystic mass, measuring 10 x 10 x 9 mm was depicted behind the left atrium with no distortion of the cardiac structures. The intracardiac anatomy appeared normal. The 3-vessel view demonstrated normal relationship of the pulmonary artery, aorta and superior vena cava. However, cross-sectional views showed a large communication (8mm) between the ascending aorta and the pulmonary artery (Figure 1). Both branches of the pulmonary artery were identified separately and appeared normal. The arterial wall defect was proximal and separate from the origin of the right pulmonary artery (type 1 according to Richardson classification).[1] Colour flow mapping showed bidirectional shunting across the defect, thus confirming the presence of the APW. The family was informed about the diagnosis, the need for post-natal open-heart surgery and the likelihood of good long-term outcome.

A three-dimensional scan was performed at 37^{+4} weeks (Prestige V20, Medison, Korea). Static rather than STIC (Spatio-Temporal Image Correlation) volumes were obtained due to frequent fetal body and breathing movements and limitations posed by advanced gestational age. Volumes

were analyzed off-line using a dedicated computer software (Sonoview Pro, version 1.6.2, Medison). Rendered images of the fetal APW are shown in Figure 1.

Post-natal follow up

A male baby was delivered at 37^{+6} weeks through spontaneous vaginal delivery, with no complications. Birth weight was 3550g. Post-natal echocardiography on day 5 confirmed the diagnosis of isolated APW (Type 1, measuring 8 mm). Neonatal karyotype was normal and excluded 22q11 micro-deletion. The baby was initially treated with diuretics and surgical repair performed on day 36 of life. The APW was repaired through the aorta. The ascending aorta side was closed with a pulmonary artery wall flap and autologous pericardium was used to reconstruct the pulmonary trunk. The post-operative course was uneventful and the baby was discharged home without complications eight days after surgery. The child remains well, on clinical follow up for four years. There has been no need for further intervention.

Case 2

Prenatal findings

A 28 year-old woman, gravida 3, para 1, was referred to our fetal medicine unit due to abnormal three-vessel view on anomaly scan. Family history was unremarkable. First trimester scan showed nuchal translucency thickness of 2.7mm (>95^{th} centile) and her combined risk was low. Fetal echocardiography at 22^{+5} weeks of gestation showed abdominal situs solitus with atrio-ventricular concordance. The intracardiac anatomy was normal. There was ventriculo-arterial discordance in keeping with the diagnosis of complete transposition of the great arteries. The great vessels were otherwise normal with no outflow tract obstruction. No other abnormalities were found prenatally. The anatomical survey showed no extra-cardiac abnormalities. The arterial switch procedure was explained and the family was informed about the likely good outcome. A female baby was delivered vaginally at 38^{+5} weeks of gestation, after induction of labour.

Post-natal follow up

The baby was born in good condition. No resuscitation was required and elective infusion of prostaglandin E was commenced. The post-natal echocardiogram confirmed the prenatal diagnosis of simple transposition with a wide patent foramen ovale and a large ductus arteriosus. The baby underwent an arterial switch operation on day seven of life, when the APW was identified and repaired. Based on surgical description, the APW was type 1. The arterial duct was identified and ligated and the atrial septal defect was closed. Post-surgical course was uneventful and the baby was discharged home on post-operative day 15. The fetal echocardiograms were reviewed. The APW was identified retrospectively on sagittal views (Figure 2) but could not be seen on the three-vessel view due to the abnormal relationship of the great arteries. At three-year follow up, echocardiography shows increased velocity across both pulmonary arteries related to the arterial switch operation (right = 3m/s and left = 3.2m/s). The child remains clinically well and had no further interventions.

Case 3

Prenatal findings

A 36 year-old woman, gravida 2, para 1, was referred for fetal echocardiography at 20^{+4} weeks of gestation due to inability to obtain the four-chamber view. The family history was unremarkable but for gestational diabetes in her previous pregnancy. The first trimester scan was normal. Nuchal translucency was 1.9mm and combined screening test was low risk. Fetal echocardiography demonstrated normal abdominal situs. The heart was on the left side of the fetal chest and of normal size but the four-chamber view could not be obtained in any axial plane through the fetal chest. There was an abnormal spatial orientation of the ventricles, which were in a supero-inferior position, resembling a criss-cross relationship of the ventricular inlets. The atrio-ventricular connection was concordant. The two ventricles were symmetrical and normal in size. Frame by frame review of the recorded images appeared to show a large inlet ventricular septal defect with straddling of the tricuspid valve. The great vessels were also in an unusual position but the ventriculo-arterial connection was concordant. There was no obvious outflow tract obstruction, although the left pulmonary artery could not be visualized well in any of the antenatal scans. Both semilunar valves were of normal size. There was normal systemic and pulmonary venous return. Due to the abnormal spatial orientation of the arteries, which were

also in a supero-inferior relationship, a standard three-vessel view could not be obtained. The crossover of the great vessels and their unusual course within the chest could only be seen in parasagittal views (Figure 3). The aorta appeared normal with no evidence of obstruction.

There were no extra-cardiac abnormalities. The findings and the likelihood of a univentricular palliation based on presence of a straddling tricuspid valve were explained to the family. The parents opted for amniocentesis, which was performed at 20^{+4} weeks of gestation. PCR and array CGH were normal. Around 29 weeks of gestation, patient developed gestational diabetes. A male baby, weighing 2180g was born in good condition, after spontaneous vaginal delivery at 37^{+3} weeks of gestation.

Post-natal follow up

The baby was admitted to our tertiary pediatric cardiology hospital on day two of life, in good condition, for further investigations due to complex anatomy. The echocardiogram confirmed situs solitus with concordant atrio-ventricular and ventriculo-arterial connections, with the right ventricle positioned superiorly in relation to the left ventricle. The foramen ovale was widely patent but there was no ventricular septal defect or straddling of mitral or tricuspid valves, and therefore, no obvious need for a univentricular palliation. The main pulmonary artery was shown to arise from the superior aspect of the right ventricle, being to the left of the aortic root. The aorta arose from the inferior left ventricle, with the aortic valve situated more caudally. There was a large communication between the aorta and the main pulmonary artery, indicating the presence of an APW. The left pulmonary artery was confirmed to be smaller than the right. The aortic arch could not be imaged by any standard transthoracic echocardiographic view. A Computer Tomography (CT) scan confirmed the echocardiographic findings and showed no obstruction in the aortic arch. The arch was positioned very low within the chest with the take off of aortic branches almost at the level of the diaphragm. This could also be seen in retrospect in the prenatal images (Figure 3). The left main bronchus was found to be unusually long and the left upper lobe bronchus could not be identified. The baby was discharged from hospital and commenced on diuretics. Due to increased work of breathing and oxygen saturation around 80%, cardiac catheterization was performed on day 4. The APW was described as 'not classic', mainly

due to its large size and supero-inferior relationship of the vessels. However, from the available images and surgical report the APW did not seem to extend into the right pulmonary artery (type 1). The consensus was for the baby to undergo biventricular repair on cardiopulmonary bypass, which would require weight gain. Alternatively, banding of the right pulmonary artery was to be considered if the baby became symptomatic or failed to gain weight. The baby remained well and thriving for the first four months of life. Surgery to close the APW and to augment the left pulmonary artery was performed at five months. At one-year follow up, the baby remains well from the cardiac point of view but continues to receive ongoing treatment related to lung and airway problems.

Case 4

Prenatal findings

A 30-year old woman, gravida 1, para 0, was referred for to our unit because of an abnormal four-chamber view in the routine anomaly scan. The family history was unremarkable. First trimester combined screening was normal. Fetal echocardiography performed at 20^{+3} weeks of gestation showed situs solitus with concordant atrio-ventricular and ventriculo-arterial connections. Heart size was normal but it occupied a central position with an antero-posterior axis. There were no structural intracardiac abnormalities. The coronary sinus was dilated due to a persistent left superior vena cava. There was no outflow tract obstruction. At the level of the three-vessel view, there was a wide communication between the aorta and the main pulmonary artery (Figure 4). Colour flow mapping showed bidirectional shunt, confirming the presence of an APW. Both branches of the pulmonary artery were identified separately and appeared normal, classified as type 1 according to Richardson. At the level of the three-vessel trachea view, the transverse arch was smaller than the ductal arch. On sagittal views, the diagnosis of an associated interrupted aortic arch was raised. Assessment of extra-cardiac structures showed a single umbilical artery. The right kidney could not be imaged. Parents opted for amniocentesis. The PCR and array CGH analysis were normal. Follow-up scans confirmed the diagnosis of interrupted aortic arch after the left subclavian artery (Figure 4).

Post-natal follow up

The baby was born in good condition. No resuscitation was required and the baby was commenced on elective infusion of prostaglandin E. The post-natal echocardiogram confirmed the prenatal cardiac diagnosis and showed a widely patent foramen ovale and ductus arteriosus. The right kidney was identified on ultrasound in an ectopic position. The baby underwent surgery on day seven of life. The aortic arch was reconstructed and the aortic and pulmonary aspects of the APW were repaired with pulmonary homograft. The arterial duct was ligated and the atrial septal defect closed. Post-surgical course was uneventful. At three-month follow up, the child remains well with no need for further intervention.

DISCUSSION

Review of this case series and previous reports show that prenatal diagnosis of APW is feasible. The communication between the aorta and pulmonary artery can be identified using the standard three-vessel view and therefore, can be suspected during routine mid-trimester screening. 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 by specialist fetal echocardiography.

An APW, also called aorto-pulmonary septal defect is a rare congenital heart malformation. It refers to a communication between the ascending aorta and the pulmonary artery in the presence of separate semilunar valves.[2] It accounts for 0.1-0.2 % of all cardiac defects in live births [3] with a female: male ratio of 1:3.[4] Richardson classified APW into three types.[1] In Type 1 there is a simple defect located between the aorta and the main pulmonary artery, immediately above the sinuses of Valsalva. In 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 and in Type 3 there is anomalous origin of the right pulmonary artery from the ascending aorta. The most frequent form is type 1.[5] An APW occurs in isolation or associated with other cardiac defects.[2] Kutsche and van Mierop reported additional malformations in 52% in a series of 188 cases.[6] These included simple lesions such as ventricular and atrial septal defects, patent arterial duct, aortic and pulmonary stenosis as well as more complex abnormalities such as interrupted aortic arch, coarctation of the aorta, aortic valve atresia, tetralogy of Fallot and transposition of the great arteries. If untreated, isolated APW can lead to heart failure and pulmonary hypertension with irreversible pulmonary vascular disease.[7]. Therefore, early diagnosis is important to optimise medical and surgical treatment. In cases with associated malformations, clinical presentation and time of intervention will depend on the nature of the additional abnormalities.

Previous publications on prenatal diagnosis of APW are restricted to case reports, either isolated or with coexisting abnormalities (Table 1). In six of the seven previous reports, the diagnosis was made prenatally. In all, the APW could be seen at the level of the three vessel- view.[2, 4, 8-11] In the

remaining case,[12] with associated tetralogy of Fallot and severe pulmonary stenosis/atresia, the window was only identified post-natally. The authors argued that abnormal flow conditions with reversed flow in the pulmonary artery contributed to failure to identify the APW prenatally.

In this series of four cases, the APW was accurately diagnosed prenatally in two when it was clearly demonstrated in the three-vessel view. In the other two, the standard three-vessel view could not be obtained due to associated diagnosis of complete transposition in one and supero-inferior relationship of the vessels in the other. After reviewing the fetal echocardiogram in the fetus with transposition, we were able to identify the APW on sagittal views. However, we could not demonstrate the APW in the other case despite it being a large defect on post-natal scans. Although this is a small series, it highlights the importance of associated cardiac malformations, in keeping with post-natal literature.[6] The three-vessel view is the most useful cross-sectional plane to suspect an APW during routine screening and is also a diagnostic view for the fetal cardiologist. The APW may be best demonstrated with a plane that is perpendicular to the plane of the hole itself, i.e., through the side of the fetal chest rather than straight through the front or back as seen in Figures 1 and 4. This plane of insonation also allows better demonstration of the flow through the defect, an orientation that is parallel to flow. It is less likely that the APW will be identified at the level of the three-vessel view in cases with associated cardiac abnormalities that *per se* will not allow the simultaneous imaging of the aorta and pulmonary artery on the three-vessel view. In some instances, sagittal views may be diagnostic as in our case of transposition of the great arteries.

Embryologically, the beginning of the formation of the aorto-pulmonary septum can be traced to the fifth week of development, when pairs of opposing ridges appear in the truncus, called cushions or truncus sweelings. They grow towards the aortic sac, twisting around each other counter-clockwise, foreshadowing the spiral course of the future septum.[13] After complete fusion, the ridges form the aorto-pulmonary septum, dividing the truncus into separate aortic and pulmonary channels. Neural crest cells migrate to the outflow region of the heart contributing to the formation of the aorto-pulmonary septum. There are many proposed mechanisms explaining outflow tract defects, including direct insults to the truncus swellings, insults to neural crest cells

that disrupt signalling to truncus swellings or insults to neural crest cells that disrupt their contribution to fusion. All proposed mechanisms can potentially disrupt the whole process of outflow tract formation. Abnormalities in neural crest migration and signalling are linked to velocardiofacial syndrome, because neural crest cells also contribute to the craniofacial development. Although APW can be considered a conotruncal defect, we were unable to identify any report of its association with 22q11 micro-deletion (Di George syndrome) or other genetic syndromes including the velocardiofacial syndromes. Unlike other conotruncal malformations such as truncus arteriosus or interrupted aortic arch, the risk of associated chromosomal abnormalities in APW, including 22q11 deletion, seems low.[6] In our series, all fetuses but one had normal nuchal translucency in first trimester screening and all pregnancies had low combined risk. The karyotype/phenotype including 22q11 was normal in all cases. This is relevant for family counselling.

The outcome for an isolated APW is excellent.[14] Operative mortality is low. There were no surgical deaths among all cases operated in the UK in 2013-14.[15] The prognosis mainly depends on the presence of associated cardiac malformations and whether surgery is performed early.[5, 14, 16] Associated complex congenital heart disease can be a bad prognostic factor.[16] An interrupted aortic arch, for instance, was independently associated with increased mortality in one series.[14]

Patients with complex associated lesions are also at greater risk of late death. Reintervention is sometimes indicated for stenosis in both the pulmonary artery and the aorta, as well as related to the specific associated lesions.[14, 16] On follow up, residual lesions are more frequent in patients with APW Type 3, and may require early reintervention.[17]

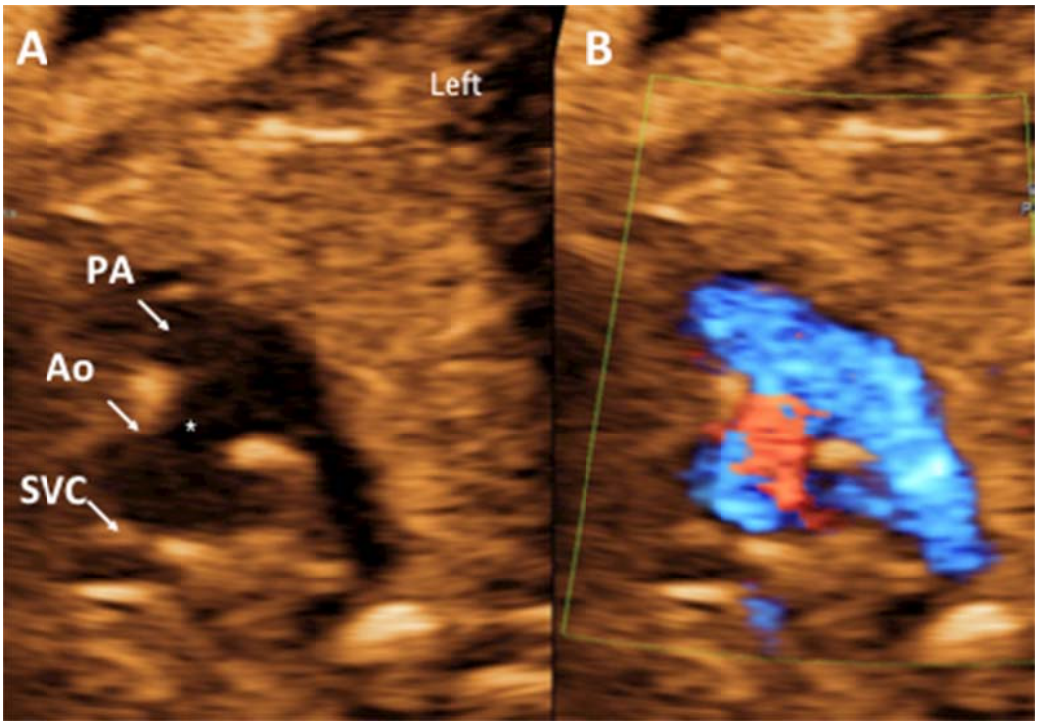
Without prenatal diagnosis, death can occur before repair is performed in cases of critical left heart obstruction. Typically, an isolated APW is large, allows a large left to right shunt leading to heart failure and is associated with high pulmonary artery systolic pressure. If there is delayed postnatal diagnosis, there is a risk of development of pulmonary vascular disease with increased morbidity and mortality. Once postnatal diagnosis is made, the APW should be repaired as early as possible, even in the presence of associated cardiovascular anomalies.[17] If the diagnosis is

known prenatally and there is no associated critical lesion, surgery is not required shortly after birth but should still be planned and performed early.

Conclusions

Although rare, APW can be diagnosed prenatally, usually at the three-vessel view level. Additional cardiac defects are common. Extra-cardiac abnormalities are uncommon. The risk of genetic or chromosomal abnormalities is low. Surgical outcome is favourable even in the presence of associated cardiac defects.

LEGENDS FOR FIGURES



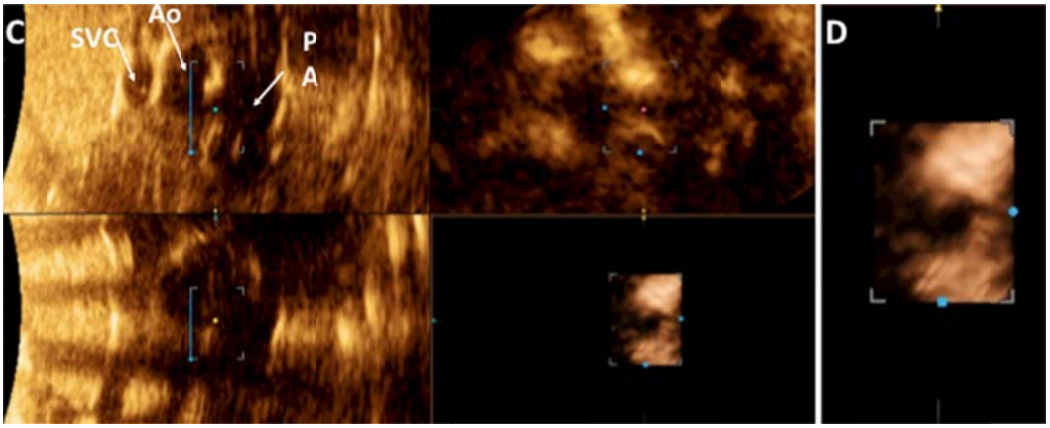


Figure 1: Fetal echo images obtained at 28^{+1} weeks of gestation for case 1. (A) and (B) show a standard three-vessel view without (A) and with (B) colour mapping. The asterisk points to the aorto-pulmonary window. (C) Multiplanar and rendered image of the window at 37^{+4} weeks. The dot corresponds to the window in the three planes. (D) shows a magnified rendered en-face view of the aorto-pulmonary window, as seen from the aortic side.

Ao: aorta, PA: pulmonary artery, SVC: superior vena cava.

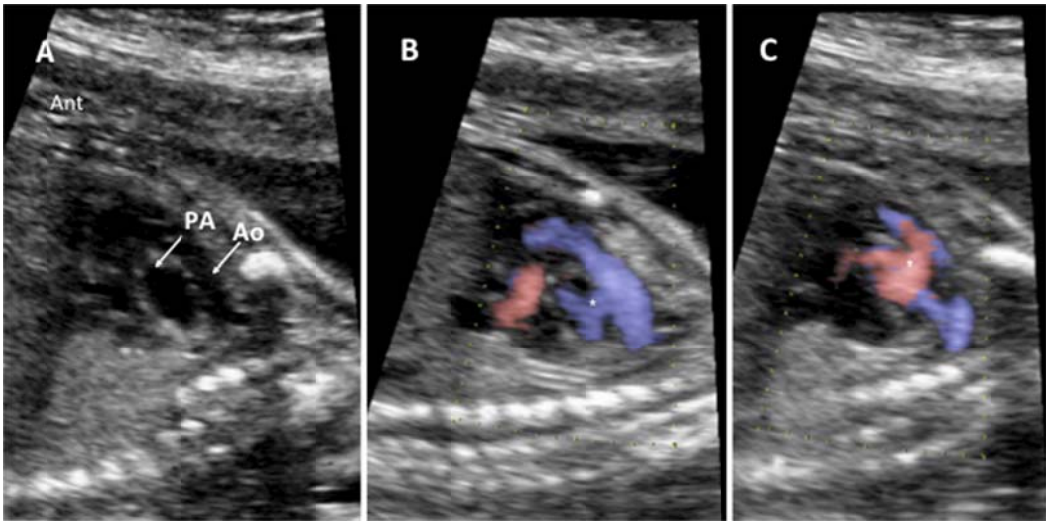


Figure 2: Fetal echo images obtained at 22^{+5} weeks of gestation in the fetus with aorto-pulmonary window and complete transposition, case 2. (A) Sagittal views of the great vessels on B-Mode. The window could not be shown with certainty. In (B and C) flow can be seen between the aorta and pulmonary artery on e-flow mapping, consistent with the post-natal diagnosis of aorto-pulmonary window (asterisk).

Ao: aorta, Ant: anterior, PA: pulmonary artery, Post: posterior.

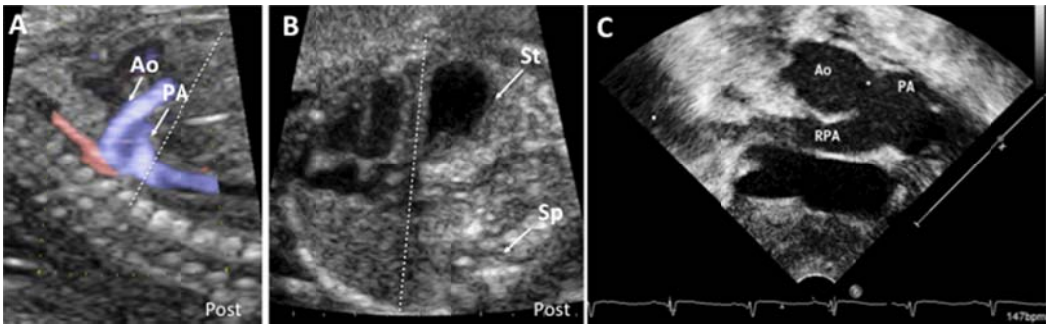


Figure 3: Fetal echo images (A) an (B) obtained at 20^{+4} weeks of gestation for case 3. (A) Parasagittal plane showing the abnormal position of the great vessels, close to the diaphragm (dotted line). (B) Oblique view show the four-chamber view and the stomach on the same plane. (C) Image obtained from post-natal echocardiography in subcostal position shows the aorto-pulmonary window (asterisk).

Ao: aorta, PA: pulmonary artery, Post: posterior, RPA: right pulmonary artery, Sp: spine, St: stomach.

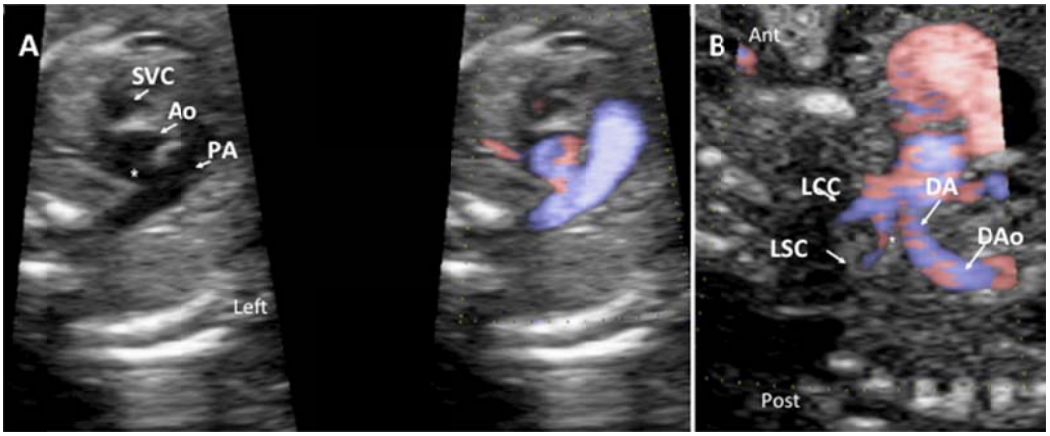


Figure 4: Fetal echo images obtained at 21^{+3} weeks of gestation for case 4. (A) at the level of the three- vessel view with and without colour. The asterisk points at the aorto-pulmonary window. (B) Sagittal view of the aortic arch, asterisk points to the site of interruption of the aortic arch.

Ao: aorta, Ant: anterior, DAo: descending aorta, DA: ductal arch, LCC: left common carotid, LSC: Left subclavian artery, PA: pulmonary artery, Post: posterior, SVC: superior vena cava.

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Table 1: Summary of cases with APW reported prenatally

	This series				Hayashi[8]	Aslan[10]	Collinet[4]	Kuehn[12]	Alvarez[2]	Kadohira[11]	Valsangiacomo[9]
Time of diagnosis	34 weeks	Postnatal	Postnatal	20 weeks	29 weeks	33 weeks	23 weeks	Postnatal	26 weeks	29 weeks	32 weeks
Other cardiac diagnosis	None	TGA	Supero-inferior ventricles, hypoplastic LPA,	IAA, LSVC	IAA	PAPVD	VSD, secundum ASD	Pulmonary atresia, VSD, LSVC, Aberrant RSA	None	IAA	RAA
Extra-cardiac findings	Thoracic cyst	None	Airway abnormalities	SUA, renal agenesis	None	Subdural hematoma & hydrocephaly	None	None	None	None	None
Karyotype/ phenotype	Normal (1)	Normal phenotype	Normal (1)	Normal (1)	Normal	Unknown	Unknown	Normal (1)	Normal (1)	Unknown	Unknown
Nuchal translucency	1.6mm	2.7mm	1.9mm	1.3mm	Unknown	Unknown	Unknown	Unknown	Unknown	Unknown	Unknown
Type of APW (Richardson classification)	Type 1	Type 1	Type 1	Type 1	Type 1	Type 1	Type 1	Type 1	Type 3	Unknown	Unknown

Abbreviations: ASD: atrial septal defect, IAA: Interrupted aortic arch, LSVC: left superior vena cava, LPA: left pulmonary artery, RAA: right aortic arch, RSA: right subclavian artery, SUA: single umbilical artery, TGA: transposition of the great arteries, VSD: ventricular septal defect. (1) includes 22q11 analysis

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