Bartsota Margarita (Orcid ID: 0000-0002-4599-2002)

Carvalho Julene S (Orcid ID: 0000-0002-5698-5828)

Double aortic arch: implications of antenatal diagnosis, differential growth of arches during pregnancy, associated abnormalities and postnatal outcome

M. Bartsota[1], V. Jowett[2], D. Manuel[1], K. Mortensen[2], J. Wolfenden[2], J. Marek[2,3] and J. S. Carvalho[1,4,5]

[1] Brompton Centre for Fetal Cardiology, Royal Brompton Hospital, London, UK

[2] Great Ormond Street Hospital, London, UK

[3] Institute of Cardiovascular Sciences, University College London, UK

[4] Fetal Medicine Unit, St George's University Hospital NHS Foundation Trust, London, UK

[5] Cardiovascular Clinical Academic Group, Molecular and Clinical Sciences Research Institute, St George's University of London, London, UK

Corresponding author: Prof. J. S. Carvalho

Brompton Centre for Fetal Cardiology, Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK

E-mail: j.carvalho@rbht.nhs.uk

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Keywords: prenatal diagnosis; echocardiography; double aortic arch; vascular ring; outcome

CONTRIBUTION

What are the novel findings of this work?

This is the largest antenatal series of double aortic arch (DAA). It shows that in the vast majority of cases both arches are patent prenatally, but postnatally the left arch is atretic in most patients, supporting the theory of differential growth of the arches. Postnatal cases of DAA with atretic left can be misinterpreted as right aortic arch and thorough assessment by computed tomography (CT) is required.

What are the clinical implications of this work?

This work will help and guide antenatal counseling and postnatal management of cases of DAA. Based on our findings, the recommendation is for these patients to be delivered at hospitals where neonatal support is available, to have early postnatal pediatric cardiology review and to undergo CT irrespective of the presence of symptoms or not.

Abstract

Objectives: This study aimed to evaluate the prenatal characteristics of double aortic arch (DAA), assess the relative size of the arches and their growth during pregnancy, describe associated cardiac, extracardiac and chromosomal/genetic abnormalities and review postnatal presentation and clinical outcome.

Methods*:* All fetuses with a confirmed diagnosis of DAA seen in five specialized referral centers between November 2012 and November 2019 were retrospectively identified from the hospitals' fetal databases. Fetal echocardiographic findings, intracardiac and extracardiac abnormalities, genetic defects, computed tomography (CT) findings and postnatal clinical presentation and outcome were evaluated.

Results*:* A total of 79 fetal cases of DAA were included. Of the whole cohort, 48.6% had an atretic left aortic arch (LAA) postnatally, with 5.1% being atretic at 1st fetal scan and diagnosed antenatally as right aortic arch (RAA). Among those who had CT scan, the LAA was atretic in 55.7%. DAA was an isolated abnormality in 91.1% of the cases, 8.9% had intracardiac (ICA) and 2.5% had extracardiac abnormalities (ECA). Among those tested, 11.5% had genetic abnormalities and specifically 22q11 microdeletion was identified in 3.8% of the patients. At a median follow up of 993.5 days, 42.5% of the patients had developed symptoms of tracheo-esophageal compression (5.5% during the first month of life) and 56.2% underwent intervention. Statistical analysis using Chi-square test showed no statistically significant correlation between patency of both aortic arches or not and need for intervention (*P*-value 0.134), development of vascular ring symptoms (*P*-value 0.350) or evidence of airway compression on CT (*P*-value 0.193)

Conclusions: Most DAA cases can be easily diagnosed in mid-gestation as both arches are patent with a dominant RAA. However, posnatally the LAA has become atretic in approximately half of the cases, supporting the theory of differential growth during pregnancy. DAA is usually an isolated abnormality; however, a thorough assessment is required, to

exclude ICA and ECA and to discuss invasive prenatal genetic testing. Postnatally, early clinical assessment is needed, and CT scan should be considered, irrespective of the presence of symptoms or not.

INTRODUCTION

Double aortic arch (DAA) is the second most common aortic arch abnormality creating vascular ring after right aortic arch (RAA) with left arterial duct and aberrant left subclavian artery (ALSA). The prevalence of DAA is reported in literature as low as 0.005%-0.007% [1,2] up to 0.01% [3] while the prevalence of RAA is estimated at 0.1% [1,2]. In the DAA cases the aortic arches can be similar in size but most frequently, the right is dominant, occurring in about 75% of the cases [4,5]. In most cases there is a left sided arterial duct.

Since the 3 vessel-trachea view (3VT) was added to the national UK cardiac protocol in 2015 [6], the number of antenatal cases of DAA increased significantly. Initially, counseling these families, especially regarding associated extracardiac and chromosomal/genetic abnormalities was based mainly on data available from RAA series, since there were no large studies of DAA cases. The aim of this study was to assess prenatal characteristics of DAA, assess the relative size of the arches and their growth during pregnancy, describe associated cardiac, extracardiac and chromosomal/genetic abnormalities and review postnatal presentation and clinical outcome.

METHODS

Study population

This is a retrospective multicenter cohort study. All fetuses with a diagnosis of DAA seen in five specialized referral centers between November 2012 and November 2019 were identified retrospectively from the hospitals' fetal databases (Figure 1). All fetal reports and if necessary, archived images were reviewed to identify cases of DAA, which constitute our study population. All cases with associated major congenital heart defects (defined as cardiac defects requiring intervention during the 1st year of life) were excluded.

Methods

All fetal echocardiograms were performed by experienced operators, using the sequential segmental analysis [7] on high-performance ultrasound systems (Aloka Alpha 10, Aloka Medical, LTd., Tokyo, Japan; Aplio i800, Canon Medical Systems Inc., Tokyo, Japan; GE Healthcare, Zipf, Austria). Images were recorded at the time of the scan and stored as digital still images and video clips (tiff, avi or DICOM).

Fetal diagnosis of DAA was made at the 3VT view at the upper mediastinum where arches were identified on both sides of the trachea (Figure 2a). Patency of both arches was also assessed on sagittal views of the aortic arch when possible (Figure 3a,b). The relative size of the arches was assessed on the first (Figure 2b) and subsequent fetal scans (Figure 2c). The side of the arterial duct was documented. Cases with prenatal diagnosis of RAA with either mirror-image branching pattern or ALSA that were proved postnatally to be cases of DAA with atretic left arch were also included in our cohort. All available electronic and medical records were reviewed to assess the presence or not of ECA, chromosomal/genetic abnormalities, computed tomography (CT) findings, postnatal clinical presentation, timing and type of intervention when required.

Categorical data were presented as number (%). Numeric data were presented as mean (standard deviation - SD) for normally distributed data while non-normal data were presented as median (range). Assessment of correlation of DAA type (patent both arches *vs* atretic left arch) with postnatal outcome was performed using Chi-square test. Correlations were deemed significant for $P \leq 0.05$. The analysis was done using IBM SPSS.

Our study was approved as service evaluation by our clinical audit department (Projects ID 003956 and 003262) and no ethical approval was required.

RESULTS

During the study period a total of 75 fetuses were diagnosed with DAA; two fetal cases of RAA with mirror-image branching pattern and two fetal cases of RAA with ALSA that postnatally were proved to be cases of DAA were also included in our cohort since one of the aims of the study was to show the differential growth of arches during pregnancy that can happen at a very early gestational age. Of these 79 patients, three (3.8%) opted for termination of pregnancy (TOP) and three were lost to follow up, so postnatal data are available in 73 cases. The median gestational age (GA) at diagnosis was 20^{+6} weeks (range, 15^{+1} to 33^{+6} weeks). Seventy-seven were singleton pregnancies and two were twin pregnancies (one dichorionic-diamniotic and one monochorionic-diamniotic pregnancy).

In the vast majority of cases (91.1%) the reason of referral to fetal cardiology services was suspected cardiac abnormality (Table 1). The number of scans in each pregnancy varied from one to four with most patients (62%) having two fetal cardiology reviews.

Antenatal findings

In the majority of the cases (83.5%) at the first fetal scan the RAA was dominant with patent smaller LAA. In four patients (5.1%) the left was atretic leading to misdiagnosis of RAA, in three (3.8%) the LAA was dominant and in six (7.6%) both aortic arches were symmetrical. The arterial duct was left-sided in all cases. Seven patients (8.9%) had minor additional cardiac abnormalities (Table 1).

Two patients (2.5%) had ECA. One had left renal agenesis and the other had multiple abnormalities including esophageal atresia, horse-shoe kidney, extra digit on hand and right choanal stenosis (Table 1). The former did not have genetic testing and the latter had normal karyotype and micro-array.

Fifty-two patients had genetic testing that included quantitative fluorescent polymerase chain reaction (QF-PCR) and chromosomal microarray analysis (either antenatally or postnatally) and two patients had non-invasive prenatal testing. Genetic abnormalities were identified in 6/52 patients (11.5%) (Table 1), including 22q11 microdeletion in two patients (3.8%), hand-foot-genital syndrome HOXA13 (n=1), small deletion on chromosome 16 (arr16p13.11) (n=1), 16p13.1 partial microdeletion Rubinstein-Taybi syndrome (n=1) and copy number loss short arm chromosome 16 (n=1).

Postnatal findings

Postnatal follow up and confirmation of the diagnosis was available in 73/76 cases. All patients with postnatal follow up were born alive and 43 (59%) of them were male. The median age at the time of 1st postnatal review was 15 days (range, 1–133 days). By the time the study was completed at a median follow up period of 993.5 days (range, 134–2733 days), 31 patients (42.5%) had developed vascular ring symptoms. Symptoms included different degrees of stridor, persistent cough, noisy breathing, dysphagia, choking episodes, feeding difficulties and one patient had intermittently signs of compromised perfusion of the left arm due to left subclavian artery (LSA) origin stenosis. Only 4 patients (5.5%) developed severe symptoms during the neonatal period warranting intervention during the first month of life and from the rest, all but one developed symptoms during the 1st year of life.

Computed tomography (CT) findings

CT was performed in 52 patients (71.2%) at a median age of 57.5 days (range, 1–1437 days). CT scans were all performed in awake patients, with the child immobilized during a 'feed and wrap' in a state of the art CT scanner with high pitch scanning excluding false negatives as a result of intubation/positive pressure ventilation with large airways expansion. When

comparing symptomatic and asymptomatic patients, 94% of the symptomatic cases had CT at a median age of 52 days (range, 1–445 days) and 53% of the asymptomatic patients had CT at a median age of 58 days (range, 13–1437 days). Regarding size of arches, in the majority of patients (44.2%) the CT showed a dominant RAA and an atretic left arch either distal to the LSA (91.3%) or between the left common carotid and the LSA (8.7%). In 16 (30.8%) patients the RAA was dominant with a smaller patent LAA, in three (5.8%) the aortic arches had similar size, in four (7.7%) the left arch was dominant and in six patients (11.5%) with antenatal diagnosis of DAA the initial CT report was interpreted as RAA with ALSA. Since fetal echocardiogram can be considered gold standard examination for diagnosis of double aortic arch with patency of both arches and after reviewing the fetal images that confirmed double aortic arch, we requested re-evaluation of the CT findings of these specific cases; after re-assessment these cases were regraded with high level of certainty to DAA with atretic LAA giving a final number of 29 (55.7%) patients with atretic LAA on the CT postnatally.

When we also included the patients who did not have CT but the patency of the arches was assessed postnatally by echocardiography only, then in our whole postnatal cohort 48.6 % of the patients had atretic LAA.

Regarding airway compression, in 17 patients (32.7%) there was at least moderate narrowing of the trachea and in two (3.8%) there was significant narrowing of the left main bronchus. Of note, in 3/29 (10.3%) symptomatic patients who had CT there were no signs of airway or esophageal compression and in 8/24 (33.3%) asymptomatic patients who had CT there was evidence of mild or moderate tracheal stenosis. However, none of our patients required tracheal reconstruction.

Intervention

Forty-one patients (56.2%) underwent intervention at a median age of 132 days (range, 5–966 days) and one had been referred for non-urgent surgery by the time the study was

completed. With the exception of one patient who had cardiac catheterization and balloon dilatation of the origin of the LSA, the remaining patients had surgical intervention that involved division of the non-dominant or atretic arch and of the ligament of the arterial duct with or without oversewing of the diverticulum of Kommerell. In three patients the LSA was translocated and re-implanted to the left common carotid artery, two patients required aortopexy and two patients had anterior pexy of the LSA. All symptomatic patients and all patients who had at least moderate stenosis of the trachea or the left main bronchus on the CT irrespective to the presence of symptoms or not, underwent intervention. Two with moderate tracheal stenosis and one with marked left main bronchus stenosis were asymptomatic. In patients with mild compression of the airway on the CT, only those who were symptomatic had surgery and only one without vascular ring symptoms and without airway compression underwent surgical intervention.

Statistical analysis using Chi-square test showed no statistically significant correlation between type of DAA (both arches patent vs atretic LAA) and need for surgery, presence of vascular ring symptoms or evidence of airway compression on CT (Table 4).

DISCUSSION

This is the largest retrospective cohort study of fetal DAA cases. It has shown differential growth of the arches during pregnancy evidenced by patency of both arches in ~95% of the whole cohort at first scan, whilst postnatally nearly 50% had atretic LAA (Table 2). In a small proportion (~5%) the left arch was already atretic at mid-gestation leading to misdiagnosis of RAA.

Similarly, DAA with atretic left can be misinterpreted postnatally as RAA. Thus, with prenatal diagnosis of DAA, the fetal echocardiogram remains the gold standard examination. The possibility of DAA with atretic LAA should be taken into consideration and excluded by thorough assessment of CT in patients with no antenatal imaging, who present postnatally as RAA especially with mirror-image branching pattern and vascular ring symptoms. CT signs indicative of DAA with LAA atresia have been described in the literature[8] (Figure 4).

Associated abnormalities

In this study we found ICA in ~9% of cases, similar to 10% reported in another series of 50 fetal cases [9], but in contrast to other studies that showed 16.6% [10] up to 31% [3] (Table 3). We showed that only 2.5% had ECA in contrast to other studies that showed higher prevalence (8% [9] to 14% [3]) (Table 3). This discrepancy cannot be explained by difference in the time of initial assessment (our study: median GA at 1st assessment 20^{+6} weeks - range 15^{+1} to 33, Vigneswaran *et al.*: 21 weeks - range 12 to 31 [9], Guo *et al.*: 29 weeks - range 23 to 31 [3]).

We observed genetic abnormalities in ~12% of our patients, which compares to 4% [9] and 6% [3] in other antenatal series. Specifically, 22q11 microdeletion accounted for only 3.8% of all cases, compared to a prevalence of 0% [9] and 6% [3] reported in previous studies. This relatively low prevalence of 22q11 microdeletion contrasts with a postnatal series that reported this microdeletion in 14% of their cases and interestingly, higher risk in patients with atretic minor arch [14]. However, this study had ascertainment bias as it included only symptomatic patients

requiring cardiopulmonary evaluation. Interestingly, in our series 3/6 affected patients (5.7%) had deletion on chromosome 16, not previously reported in association with DAA.

The association with overall chromosomal/genetic abnormalities (11.5%) and 22q11.2 microdeletion (3.8%) in this series is lower than that reported antenatally for RAA (14.1–15.3% [11,12,13] and 6.4–10% [11,12,13], respectively).

Outcome

In our study 42.5% of patients were symptomatic, which is similar to the findings of Guo *et al*. study (41%) [3] and less than the Vigneswaran *et al*. (65%) [9] and Trobo *et al*. studies (72.4%) [10] (Table 3). Based on our findings and available literature, we can conclude that symptoms occur in ~ 40–70% of patients with DAA. These findings support the theory that DAA is the tightest type of vascular ring since the reported percentage of symptomatic patients in RAA is 5.6–25.2% [2,11,15].

In our cohort 56.2% of the cases underwent intervention, a decision based on the presence of symptoms and/or significant airway and/or esophageal compression on the CT. This is higher than 41% in the Guo *et al*. study [3] and lower than 87% in Vigneswaran *et al*. study [9] (Table 3). This could be explained by the fact that in Guo *et al*. study none of the asymptomatic patients had significant tracheal or esophageal compression as opposed to Vigneswaran *et al*. study where 13/47 (27.6%) patients were asymptomatic with abnormal tracheal appearances. We did not show any statistically significant correlation between type of DAA and outcome. Based on our study and available literature, we can conclude that ~ 50-80% of DAA cases require surgical intervention.

In our study only a small number of neonates (~5%) developed symptoms significant enough to warrant neonatal intervention. This contrasts with a much higher reported 21% of patients having symptoms from birth [9]. This discrepancy may be related to severity of symptoms. Whilst our 5% relates to symptoms requiring neonatal intervention, in Vigneswaran's study, it is not clarified whether all required surgical intervention during neonatal period and 40% of these

neonates had additional pathology that may have contributed to respiratory distress [9]. Based on our findings we would recommend that patients with antenatal diagnosis of DAA are delivered at hospitals with available neonatal support. A small percentage of patients will develop significant symptoms during neonatal period and will require urgent assessment. For the many neonates who are asymptomatic we would recommend early review by pediatric cardiologist since DAA is the tightest type of vascular ring and even asymptomatic patients can have significant airway compression. CT should be performed in all patients irrespective of the presence of symptoms or not since the absence of symptoms does not exclude significant airway compression that may warrant intervention.

Study limitations

The limitations of this study include the retrospective nature of the study, the fact that genetic testing and CT imaging had not been performed in all patients by the time that the study was completed and that 15% of the patients had follow up time less than a year.

Conclusion

In summary, DAA can be easily diagnosed at mid-gestation during fetal life as in most patients with DAA both arches are patent at this stage. Postnatally the LAA has become atretic in most of the patients, supporting the theory of differential growth of arches. In postnatal diagnosis of RAA cases, the possibility of DAA with atretic LAA should be considered and excluded. DAA is usually an isolated abnormality; however, thorough assessment for exclusion of ICA and ECA is required. The probability of associated genetic/chromosomal abnormalities is relatively low but not negligible and the option of invasive prenatal genetic testing should be discussed and offered to the families. There is no evidence of any statistically significant correlation between type of DAA and postnatal outcome. The recommendations for these patients are delivery at hospitals with available neonatal support, early postnatal pediatric cardiology review and performance of CT irrespective of the presence of symptoms or not.

Bartsota Margarita (Orcid ID: 0000-0002-4599-2002)

Carvalho Julene S (Orcid ID: 0000-0002-5698-5828)

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FIGURE LEGENDS

Figure 1. Number of cases of double aortic arch detected antenatally per calendar year

Figure 2. (a) 3VT view of double aortic arch (DAA) at mid-gestation with Color Doppler. (b) 3VT view of DAA at mid-gestation. (c) 3VT view of DAA at 3rd trimester. RAA, right aortic arch. LAA, left aortic arch.

Figure 3. (a,b) Sagittal views of both arches in double aortic arch.

Figure 4. Computed tomography (CT) images of double aortic arch (DAA) with atretic left arch CT in a 4-year-old with CT performed awake as a non-ECG-gated study. Principal features of CT diagnosis of DAA with atretic left: a relatively acute angle and rather posterior course of the patent and anterior part of LAA in the course towards the left CCA origin (a, arrow), a trapezoid-like position of the four aortic arch branches in an axial plane (b), a tubular Kommerell's diverticulum (c, arrow) with suggested ligamentous tethering between the diverticulum and the patent anterior part of the LAA (d, arrow).

Table 1: Antenatal and postnatal data in 79 fetuses with double aortic arch

Reason for referral	
Suspected cardiac abnormality	72/79 (91.1%)
Family history of congenital heart disease	3/79 (3.8%)
Increased nuchal translucency	1/79 (1.3%)
Inadequate/difficult heart views	3/79 (3.8%)
Other fetal cardiac findings	**7/79 (8.9%)**
Small muscular ventricular septal defect	2 /79 (2.5%)
Multiple small muscular ventricular septal defects	1/79 (1.3%)
Bilateral superior vena cava	3/79 (3.8%)
Small aorto-pulmonary window	1/79 (1.3%)
Extracardiac fetal abnormalities	**2/79 (2.5%)**
Left renal agenesis	1/79 (1.25%)
Multiple abnormalities	1/79 (1.25%)
Genetic Findings†	
Abnormal genotype	6/52 (11.5%)
22q11 microdeletion syndrome	2/52 (3.8%)

Data are given as n/N (%). †Includes only those with genetic testing.

Table 2: Antenatal and postnatal characteristics of morphology of arches in cases assessed by computed tomography

	Antenatally	Postnatally
Dominant RAA with patent LAA	83.5%	30.8%
Dominant RAA with atretic LAA	5.1%	55.7%
Dominant LAA	3.8%	7.7%
Symmetrical arches	7.6%	5.8%

Data are given as %. LAA, left aortic arch; RAA, right aortic arch.

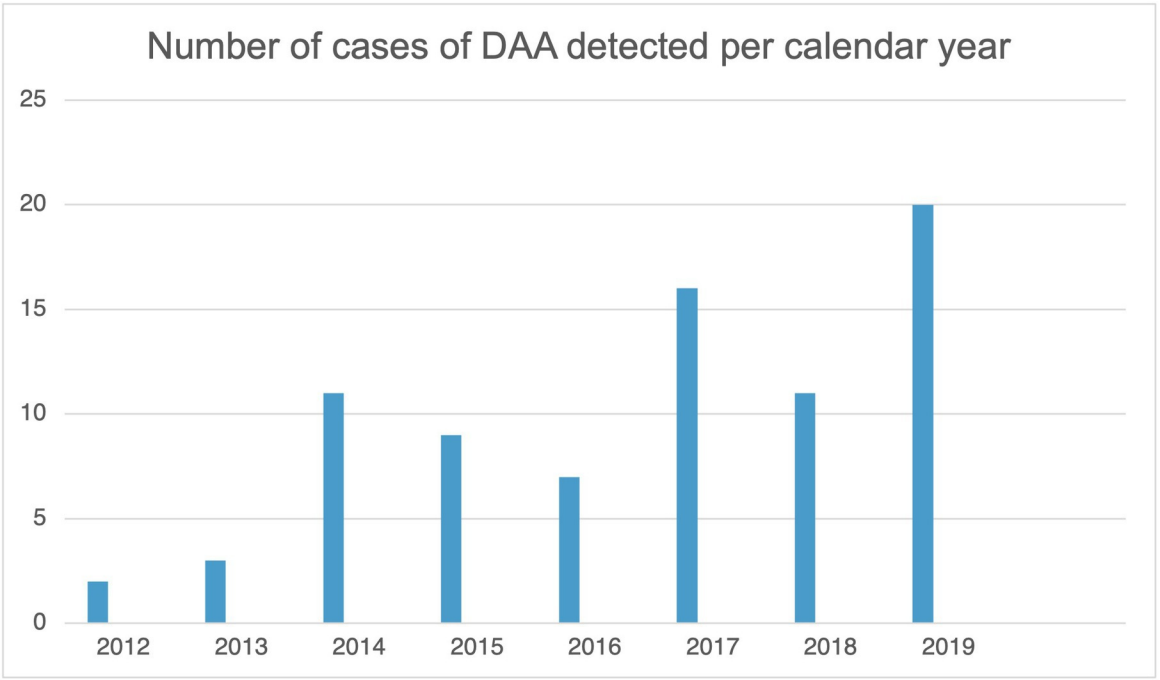
Table 3: Largest antenatal series of double aortic arch

	This series	Guo 2020	Vigneswaran 2020
	N= 79	N= 36	N= 50
Isolated	91.1%	69%	90%
Associated cardiovascular abnormality	8.9%	31%	10%
Associated extracardiac abnormality	2.5%	14%	8%
Associated genetic abnormality	11.5%	6%	4%
22q11 microdeletion	3.8%	6%	0%
Symptomatic	42.5%	41%	65%
Intervention	56.2%	41%	87%

Table 4: Correlation of morphology of arches with postnatal outcome

	Patent both arches	Atretic left arch	*P*-value
Postnatal intervention	18/36 (50%)	23/34 (67.6%)	0.134
Vascular ring symptoms	14/36 (38.8%)	17/34 (50%)	0.350
Airway compression on CT	16/23 (69.5%)	15/29 (51.7%)	0.193

CT, computed tomography.



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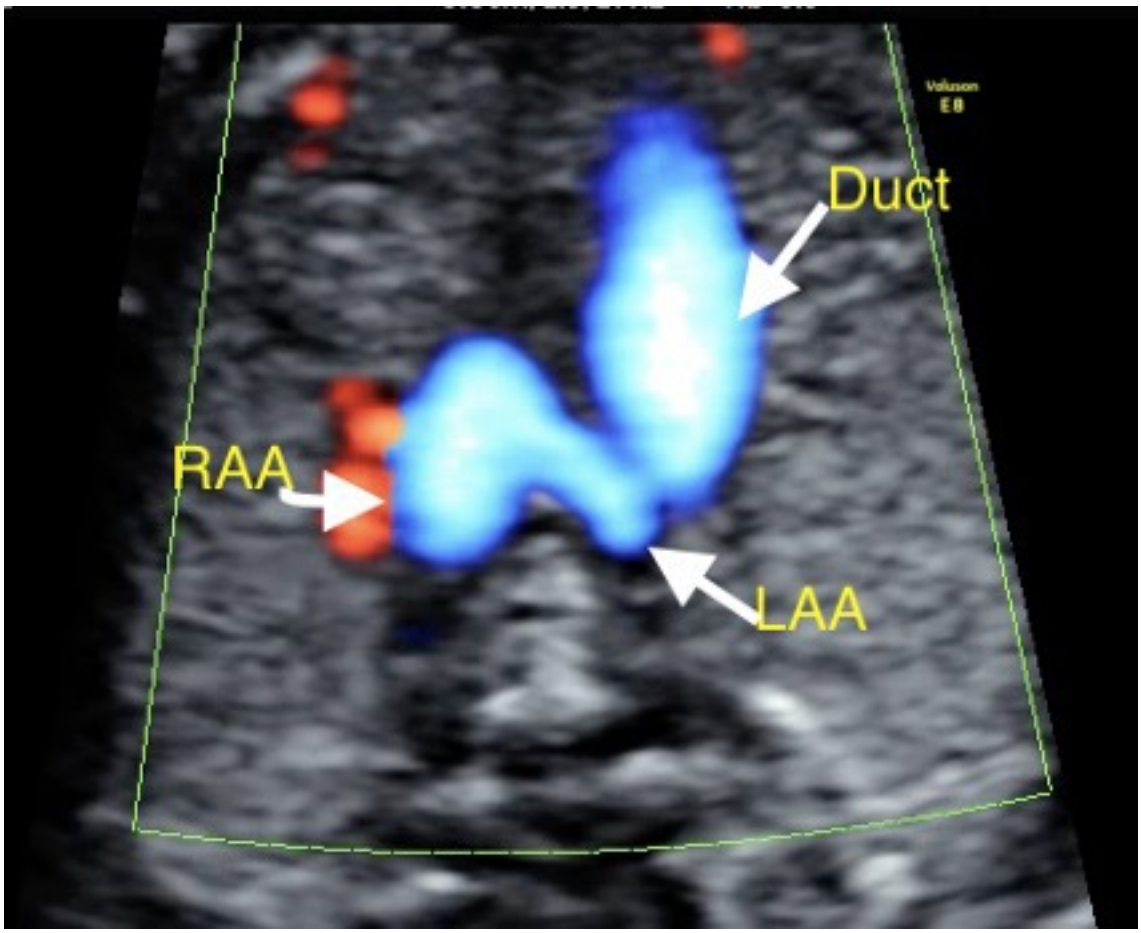

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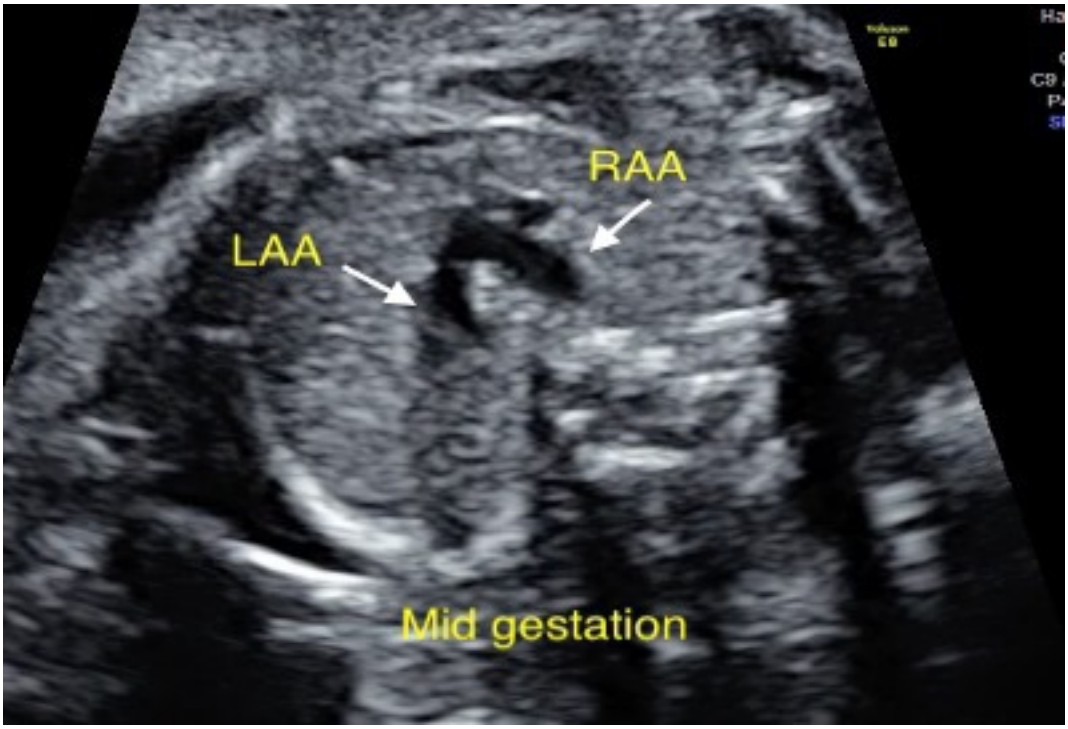

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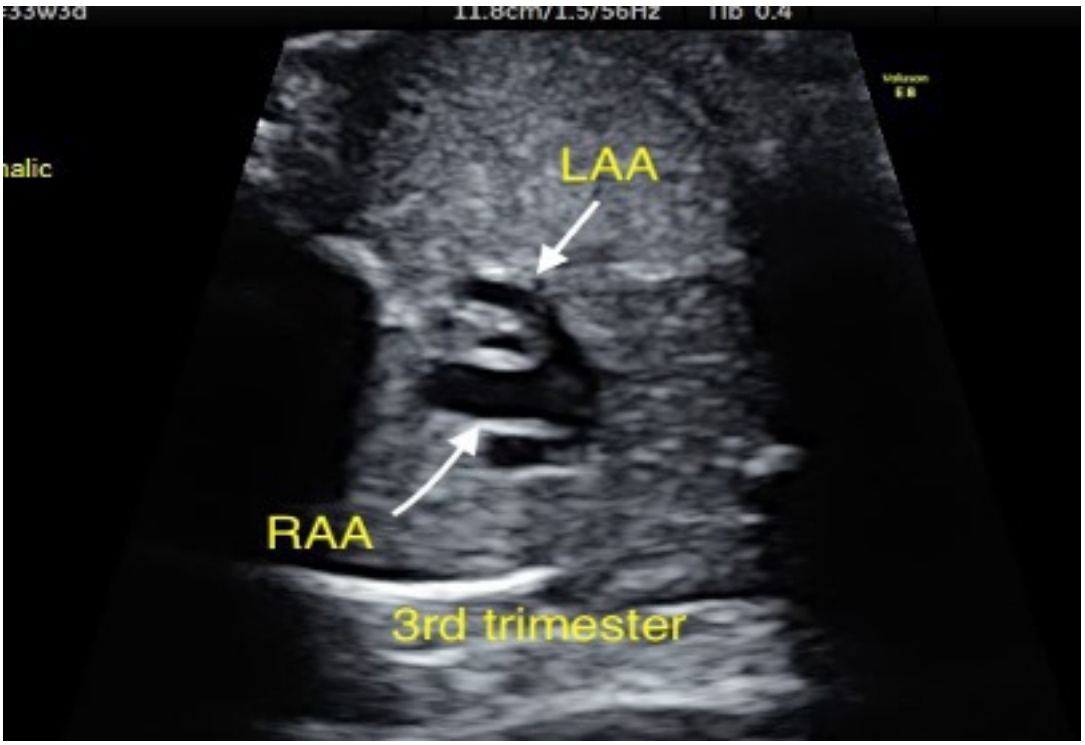

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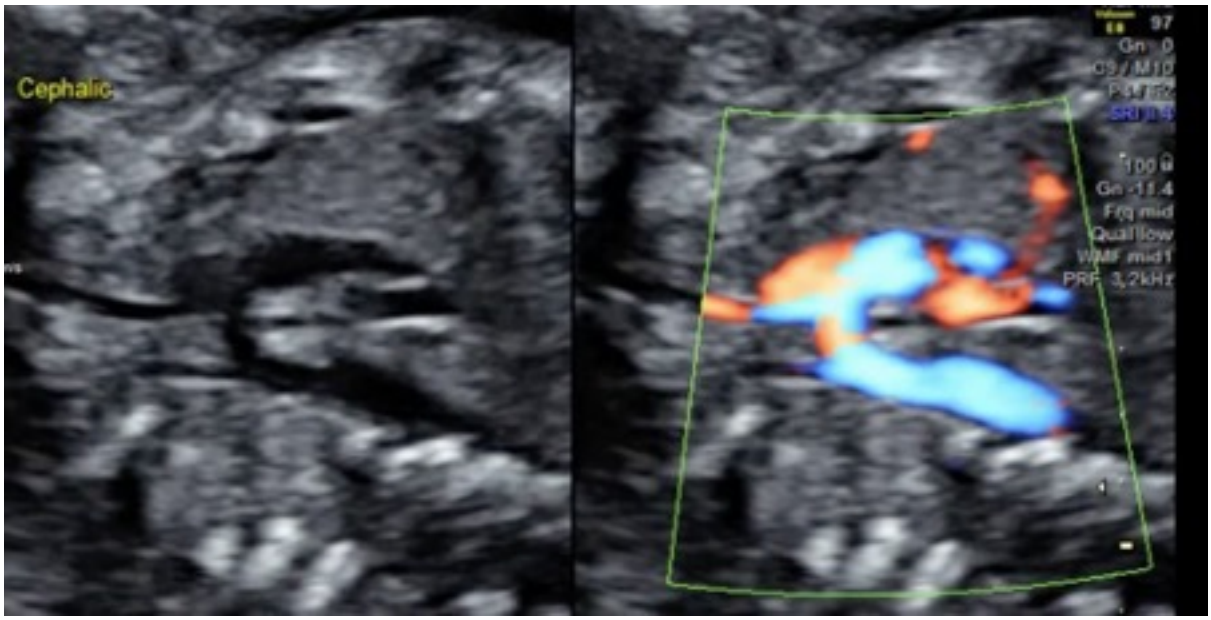

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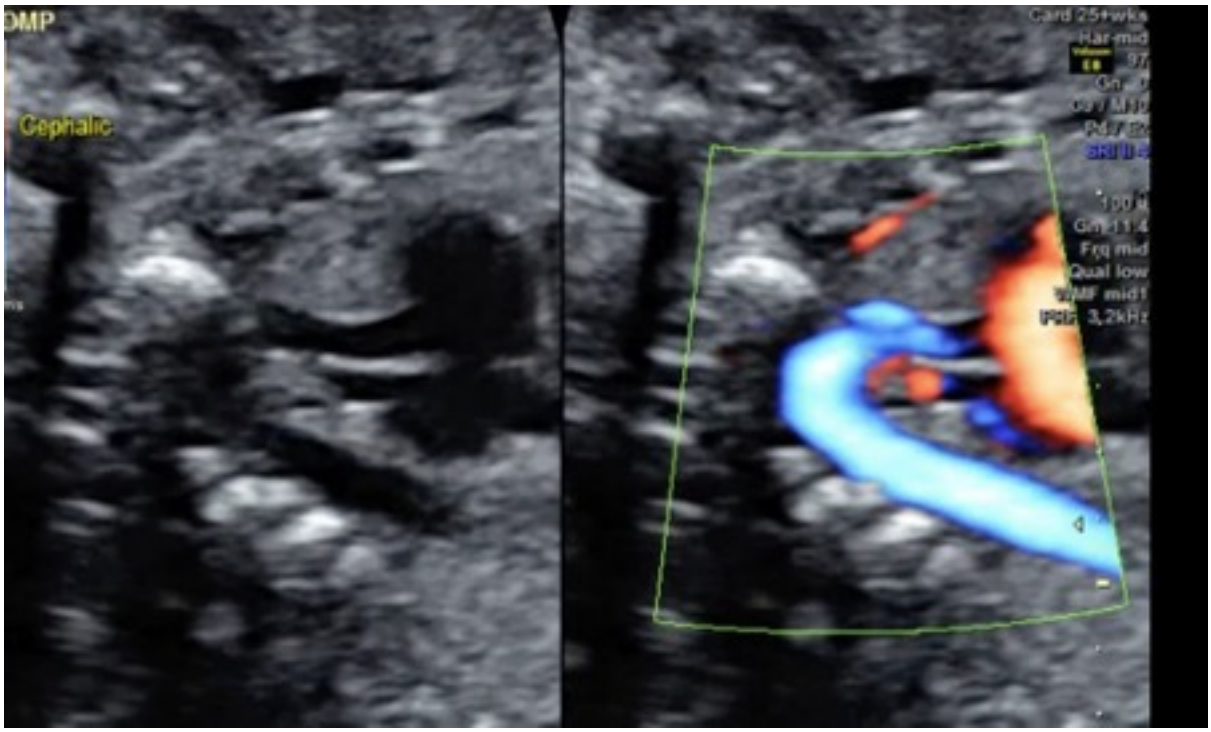

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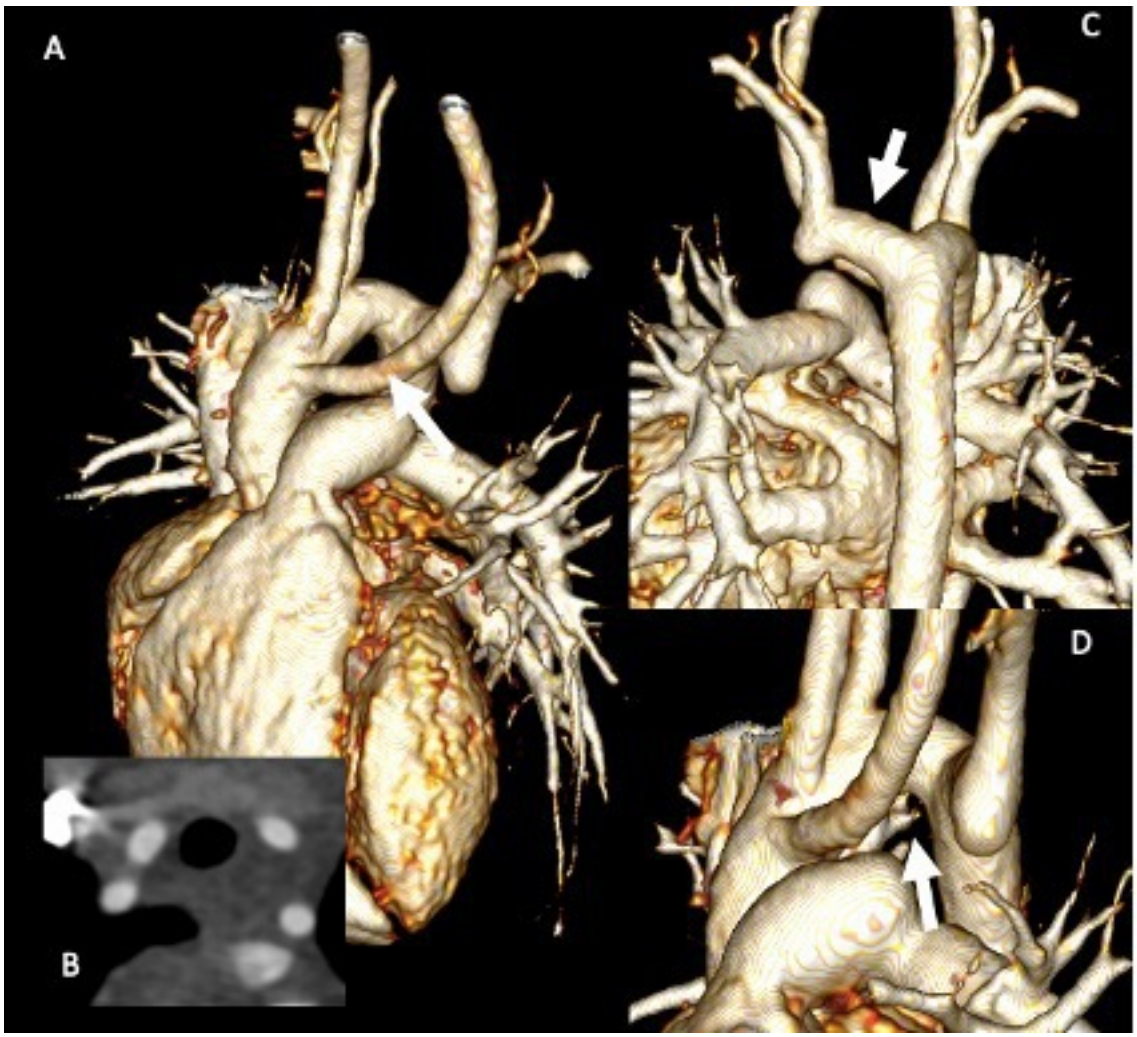

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