Ultrasound Obstet Gynecol 2023; **62**: 69–74
Published online 9 June 2023 in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.26186.

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

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KEYWORDS: double aortic arch; echocardiography; postnatal outcome; prenatal diagnosis; vascular ring

CONTRIBUTION

What are the novel findings of this work?
This is the largest reported antenatal series of double aortic arch (DAA). Both arches were patent in the majority of cases prenatally, but most patients had atretic left arch postnatally, supporting the theory of differential growth of the arches during pregnancy. Postnatal cases of DAA with atretic left arch may be misinterpreted as right aortic arch, and thorough assessment using computed tomography (CT) is required.

What are the clinical implications of this work?
This work should guide antenatal counseling and postnatal management of DAA cases. Based on our findings, we recommend that affected patients are delivered at hospitals in which neonatal support is available, receive early postnatal pediatric cardiology review and undergo CT evaluation, irrespective of the presence of symptoms.

ABSTRACT

Objectives *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 *This was a retrospective cohort study of all fetuses with a confirmed diagnosis of DAA seen in five specialized referral centers in London, UK, between October 2012 and November 2019. Cases were 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 fetuses with DAA were included. Of those assessed postnatally, 48.6% had an atretic left aortic arch (LAA), while 5.1% had an atretic LAA at the first fetal scan and were misdiagnosed antenatally with right aortic arch (RAA). The LAA was atretic in 55.8% of those who underwent CT. DAA was an isolated abnormality in 91.1% of cases; 8.9% of patients had an additional intracardiac abnormality and 2.5% had both intra- and extracardiac abnormalities. Among the 52 cases that underwent genetic testing, 11.5% had genetic abnormalities and, specifically, the 22q11 microdeletion was identified in 3.8% of patients. At a median follow-up of 993.5 days, 42.5% of patients had developed symptoms of tracheoesophageal compression (5.5% during the first month after birth) and 56.2% had undergone intervention. Statistical analysis using the* χ*-square test showed no significant relationship between morphology of DAA (patency of both aortic arches* vs *atretic LAA) and the need for intervention (*$P = 0.134$*), development of vascular ring symptoms (*$P = 0.350$*) or evidence of airway compression on CT (*$P = 0.193$*).*

Conclusions *Most cases of DAA can be diagnosed easily at midgestation, as typically both arches are patent with a*

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Accepted: 13 February 2023

dominant RAA at this stage. However, we found that the LAA had become atretic in approximately half of the cases postnatally, supporting the theory of differential growth of the arches during pregnancy. DAA is usually an isolated abnormality; however, thorough assessment is required to exclude associated intra- and extracardiac anomalies and to determine the need for invasive prenatal genetic testing. Postnatally, early clinical assessment is needed and CT scan should be considered, irrespective of the presence of symptoms. © *2023 The Authors. Ultrasound in Obstetrics & Gynecology published by John Wiley & Sons Ltd on behalf of International Society of Ultrasound in Obstetrics and Gynecology.*

INTRODUCTION

Double aortic arch (DAA) is the second most common form of aortic arch abnormality that creates a vascular ring, after right aortic arch (RAA) with left arterial duct and aberrant left subclavian artery (ALSA). The prevalence of DAA reported in the literature varies from 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]. The two aortic arches can be similar in size but the right arch is dominant in approximately 75% of cases[4,5]. In most cases of DAA, there is a left-sided arterial duct.

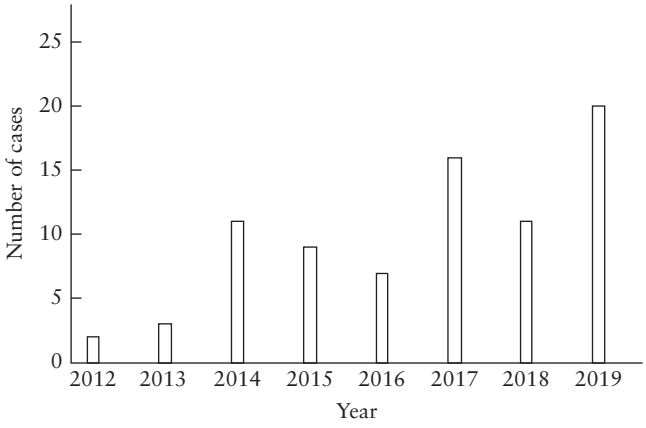

Figure 1 Number of cases of fetal double aortic arch detected per year in five referral centers between October 2012 and November 2019.

Since the three-vessel-and-trachea view was added to the UK national cardiac screening protocol in 2015[6], the number of cases of DAA detected antenatally has increased significantly. Initially, parental counseling, especially regarding associated structural and chromosomal/genetic abnormalities, was based mainly on data available from RAA series, since no large studies of DAA had been conducted. The aims of this study were: to evaluate the prenatal characteristics of DAA; to assess the relative size of the arches and their growth during pregnancy; to describe associated intracardiac (ICA), extracardiac (ECA) and chromosomal/genetic abnormalities; and to review postnatal presentation and clinical outcome.

METHODS

Study population

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

Echocardiography and data review

All fetal echocardiograms were performed by experienced operators using 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; Voluson, GE Healthcare, Zipf, Austria). Images were recorded at the time of the scan and stored as digital still images and videoclips (tiff, avi or DICOM).

Fetal diagnosis of DAA was made upon identification of arches on both sides of the trachea in the three-vessel-and-trachea view at the upper mediastinum (Figure 2). Patency of both arches was assessed on sagittal views when possible (Figure 3). The relative size of the arches was assessed on the first (Figure 2b) and

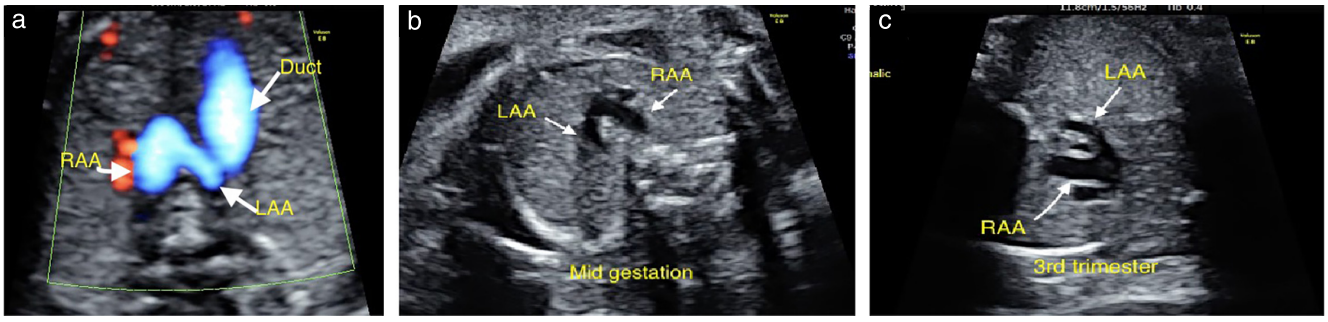

Figure 2 Color Doppler (a) and grayscale (b,c) ultrasound images in three-vessel-and-trachea view in fetuses with double aortic arch at midgestation (a,b) and in third trimester (c). (b) and (c) were taken in same fetus. LAA, left aortic arch; RAA, right aortic arch.

subsequent (Figure 2c) fetal scans. The side of the arterial duct was documented. Cases with a prenatal diagnosis of RAA with either a mirror-image branching pattern or ALSA that were determined postnatally to have DAA with atretic LAA were also included in our cohort. All available electronic and medical records were reviewed to ascertain the presence of ICA, ECA and chromosomal/genetic abnormalities, to evaluate findings on computed tomography (CT), and to review postnatal clinical presentation and the timing and type of intervention, if required.

Statistical analysis

Categorical data are presented as *n* (%) and continuous data as median (range). The relationship between type of DAA (both arches patent *vs* atretic LAA) and postnatal outcome was assessed using the χ-square test. Statistical analysis was performed using SPSS software (IBM Corp., Armonk, NY, USA); $P \leq 0.05$ was considered to indicate statistical significance.

This study was classified under the definition of service evaluation by our clinical audit department (project IDs: 003956 and 003262) and therefore ethical approval was not required.

RESULTS

During the study period, a total of 75 fetuses were diagnosed with DAA. Two fetal cases of RAA with a mirror-image branching pattern and two of RAA with ALSA that were determined postnatally to be cases of DAA were also included in our cohort, since one of the aims of this study was to evaluate the differential growth of arches during pregnancy, which can happen at an early gestational age. Of these 79 patients, three (3.8%) opted for termination of pregnancy and three were lost to follow-up, so postnatal data were available for 73 cases. The median gestational age 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).

In the vast majority (91.1%) of cases, the reason for referral to fetal cardiology services was suspected cardiac abnormality (Table 1). The number of scans per pregnancy varied from one to four, with most patients (62.0%) having two fetal cardiology reviews.

Antenatal findings

In the majority (83.5%) of cases, the RAA was dominant at the first fetal scan, with patent smaller LAA (Table 2). In four (5.1%) patients, the LAA was atretic leading to misdiagnosis as RAA; in three (3.8%), the LAA was dominant; and in six (7.6%), the aortic arches were symmetrical. The arterial duct was left-sided in all cases. Seven (8.9%) patients had minor additional ICA (Table 1).

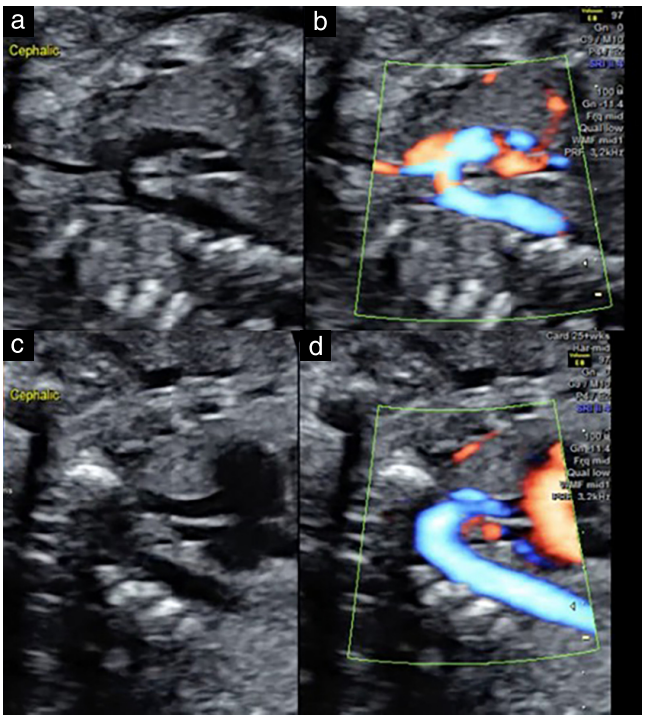

Figure 3 Grayscale (a,c) and color Doppler (b,d) ultrasound images in sagittal view in fetus with double aortic arch, showing right and left arches.

Table 1 Reason for referral and associated structural and genetic abnormalities in 79 fetuses with double aortic arch

Characteristic	*Value*
Reason for referral	
Suspected cardiac abnormality	72 (91.1)
Family history of CHD	3 (3.8)
Increased nuchal translucency	1 (1.3)
Inadequate/difficult heart views	3 (3.8)
Other fetal cardiac abnormalities	7 (8.9)
Small muscular VSD	2 (2.5)
Multiple small muscular VSDs	1 (1.3)
Bilateral superior vena cava	3 (3.8)
Small aortopulmonary window	1 (1.3)
Extracardiac fetal abnormalities	2 (2.5)
Left renal agenesis	1 (1.3)
Multiple abnormalities	1 (1.3)
Genetic abnormalities*	6/52 (11.5)
22q11 microdeletion syndrome	2/52 (3.8)

Data are given as *n* (%) or *n*/*N* (%). *Includes only patients who underwent invasive genetic testing. CHD, congenital heart disease; VSD, ventricular septal defect.

Table 2 Morphology of arches on antenatal echocardiography and postnatal computed tomography in cases with double aortic arch

Parameter	*Antenatal (n = 79)*	*Postnatal (n = 52)*
Dominant RAA with patent LAA	66 (83.5)	16 (30.8)
Dominant RAA with atretic LAA	4 (5.1)	29 (55.8)
Dominant LAA	3 (3.8)	4 (7.7)
Symmetrical arches	6 (7.6)	3 (5.8)

Data are given as *n* (%). LAA, left aortic arch; RAA, right aortic arch.

Two (2.5%) patients were diagnosed with ECA (Table 1). One had left renal agenesis and the other had multiple abnormalities, including esophageal atresia, horseshoe kidney, polydactyly of the hand and right choanal stenosis. The former did not undergo genetic testing and the latter had normal karyotype and microarray analysis.

Fifty-two patients underwent genetic testing comprising quantitative fluorescent polymerase chain reaction and chromosomal microarray analysis (either ante- or postnatally) and two patients had non-invasive prenatal testing. Genetic abnormalities were identified in 6/52 (11.5%) patients (Table 1), including two (3.8%) cases of 22q11 microdeletion and single cases, respectively, of hand–foot–genital syndrome (*HOXA13*), small deletion on chromosome 16 (arr16p13.11), Rubinstein–Taybi syndrome (16p13.1 partial microdeletion) and copy-number loss on the short arm of chromosome 16.

Postnatal findings

Postnatal follow-up and confirmation of diagnosis were available for 73 cases. All patients with postnatal follow-up were liveborn and 43 (58.9%) were male. The median age at first postnatal review was 15 days (range, 1–133 days). By the time the study was completed, with a median follow-up period of 993.5 days (range, 134–2733 days), 31 (42.5%) patients had developed symptoms of vascular ring. Symptoms included varying degrees of stridor, persistent cough, noisy breathing, dysphagia, choking episodes, feeding difficulties and, in one patient, intermittent signs of compromised perfusion of the left arm due to stenosis of the left subclavian artery (LSA) origin. Only four (5.5%) of these neonates developed severe symptoms that warranted intervention during the first month after birth; of the remaining 27, all but one developed symptoms during the first year after birth.

Computed tomography findings

CT was performed in 52 (71.2%) patients at a median age of 57.5 days (range, 1–1437 days). All CT scans were performed without anesthesia, using a 'feed and wrap' technique to immobilize the infant, and were conducted using a state-of-the-art CT scanner with high-pitch scanning, which excludes false negatives resulting from intubation or positive pressure ventilation with large airways expansion. CT was performed in 93.5% of symptomatic patients, at a median age of 52 days (range, 1–445 days), and in 54.8% of asymptomatic patients, at a median age of 58 days (range, 13–1437 days). Regarding the size of arches on CT (Table 2), the majority (44.2%) of patients were found to have a dominant RAA and an atretic LAA, either distal to the LSA (91.3%) or between the left common carotid artery 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 were similar in size; in four (7.7%), the LAA was dominant; and in six (11.5%) patients with an antenatal diagnosis of DAA, the initial CT report was interpreted as RAA with ALSA. Since fetal echocardiography can be considered the gold-standard examination for the diagnosis of DAA with patency of both arches, and after confirming DAA on review of the fetal images, we requested re-evaluation of the CT findings in these cases. Following reassessment, these six cases were regraded with a high level of certainty to DAA with atretic LAA, giving a total of 29 (55.8%) patients with atretic LAA on CT postnatally.

When we included additionally patients who did not undergo CT but for whom the patency of the arches was assessed postnatally on echocardiography only, then the proportion of our whole postnatal cohort with atretic LAA was 48.6%.

Airway compression was observed in 31/52 (59.6%) patients on CT. At least moderate narrowing of the trachea was noted in 17 (32.7%) patients and significant narrowing of the left main bronchus was noted in two (3.8%). Of 29 symptomatic patients who underwent CT, three (10.3%) showed no signs of airway or esophageal compression; and among 23 asymptomatic patients who underwent CT, eight (34.8%) had evidence of mild or moderate tracheal stenosis. However, none of the patients in our cohort required tracheal reconstruction.

Intervention

By the time the study was completed, a total of 41 (56.2%) patients had undergone intervention, at a median age of 132 days (range, 5–966 days), and one had been referred for non-urgent surgery. With the exception of one patient who underwent cardiac catheterization and balloon dilatation of the origin of the LSA, surgical intervention 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 reimplanted 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 with at least moderate stenosis of the trachea or left main bronchus on CT, irrespective of the presence of symptoms, underwent intervention. Two patients with moderate tracheal stenosis and one with marked left main bronchus stenosis were asymptomatic. Among patients with mild compression of the airway on CT, only those who were symptomatic had surgery. Only one patient with neither vascular ring symptoms nor airway compression underwent surgical intervention.

Statistical analysis using the χ-square test showed no significant relationship between the 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 3).

Table 3 Morphology of arches on postnatal echocardiography and postnatal outcome in infants with double aortic arch

Outcome	*Both arches patent*	*Atretic left arch*	P
Postnatal intervention	18/36 (50.0)	23/34 (67.6)	0.134
Vascular ring symptoms	14/36 (38.9)	17/34 (50.0)	0.350
Airway compression on CT	16/23 (69.6)	15/29 (51.7)	0.193

Data are given as *n*/*N* (%). Information regarding patency of left arch on postnatal echocardiography was available in 70 cases. Postnatal computed tomography (CT) was performed in 52 cases.

DISCUSSION

To our knowledge, this is the largest retrospective cohort study of fetal DAA. We demonstrated differential growth of the arches during pregnancy; patency of both arches was observed in around 95% of the whole cohort at the first scan, whereas postnatally, nearly 50% had atretic LAA. In a small proportion of fetuses (around 5%), the LAA was already atretic at midgestation, leading to misdiagnosis as RAA. Similarly, DAA with atretic LAA can be misinterpreted as RAA postnatally. Thus, fetal echocardiography remains the gold-standard examination for the prenatal diagnosis of DAA; however, in patients with a postnatal diagnosis of RAA for whom no antenatal imaging is available, especially those with a mirror-image branching pattern and vascular ring symptoms, the possibility of DAA with atretic LAA should be taken into consideration and excluded by thorough assessment of CT findings. CT signs indicative of DAA with LAA atresia have been described in the literature[8] (Figure 4).

Associated abnormalities

In this study, we observed associated ICA in around 9% of cases, which is similar to the 10% rate reported by Vigneswaran *et al.*[9] in a series of 50 fetal cases of DAA, but in contrast to the 31% rate observed in a cohort of 36 DAA cases diagnosed antenatally by Guo *et al.*[3] (Table 4). We found that only 2.5% of fetuses had ECA, while other studies have reported a higher prevalence (8%[9] to 14%[3]). This discrepancy cannot be explained by a difference in the timing of the initial assessment; median gestational age at first assessment was 20 + 6 weeks (range, 15 + 1 to 33 + 6 weeks) in the present study, 21 weeks (range, 12–31 weeks) in the study of Vigneswaran *et al.*[9] and 27 weeks (range, 23–31 weeks) in that of Guo *et al.*[3].

We observed genetic abnormalities in around 12% of our patients, which compares with 4%[9] and 6%[3] in other antenatal series. Specifically, 22q11 microdeletion accounted for only 3.8% of all cases in our cohort, compared with 0%[9] and 6%[3] in previous studies. The relatively low prevalence of 22q11 microdeletion in the present cohort contrasts with that found in a postnatal series[10], which reported this anomaly in 14% of cases and noted a higher risk in patients with atretic minor arch. However, this study had ascertainment bias, as it included only symptomatic patients requiring cardiopulmonary evaluation. In our series, 3/6 patients with abnormal genotype (5.8% of those who underwent genetic testing) had a deletion on chromosome 16, which has not previously been reported in association with DAA. Notably, the prevalence of overall chromosomal/genetic abnormalities (11.5%) and 22q11 microdeletion (3.8%) in this series is lower than that reported antenatally for RAA (14.1–15.3% and 6.4–10%, respectively)[11–13].

Outcome

In our study, 42.5% of patients were symptomatic, which is similar to the rate reported by Guo *et al.*[3] (41%)

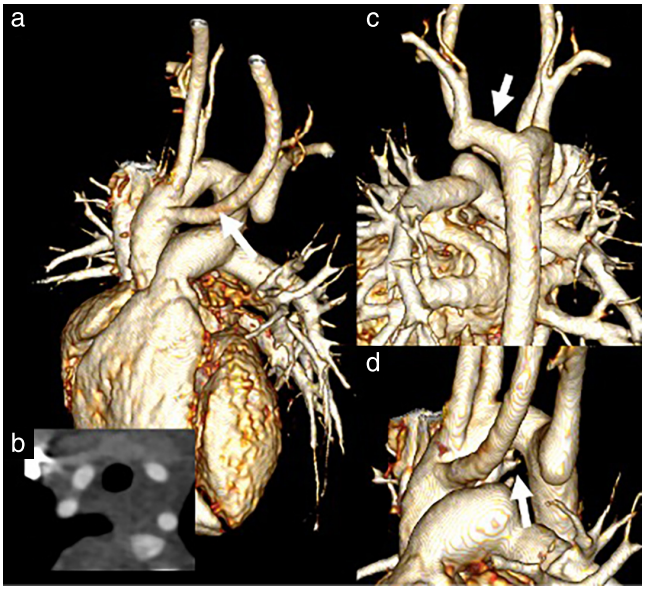

Figure 4 Computed tomography (CT) images of double aortic arch (DAA) with atretic left aortic arch (LAA), taken in 4-year-old infant while awake, without electrocardiogram gating. Principal features of DAA with atretic LAA on CT include: (a) relatively acute angle and rather posterior course of patent anterior part of LAA towards origin of left common carotid artery (arrow); (b) trapezoid-like positioning of four aortic arch branches in axial plane; (c) tubular Kommerell's diverticulum (arrow); and (d) ligament (arrow) suggesting tethering between diverticulum and patent anterior part of LAA.

Table 4 Clinical characteristics of cases in largest antenatal series of double aortic arch

Parameter	*This series (n = 79)*	*Guo (2020)[3] (n = 36)*	*Vigneswaran (2021)[9] (n = 50)*
Isolated abnormality	91.1	69	90
Associated cardiac 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 necessary	56.2	41	87

Data are given as %.

and lower than that reported by Vigneswaran *et al.*[9] (65%) and Trobo *et al.*[14] (72.4%) (Table 4). Based on our findings and the available literature, we can conclude that symptoms occur in approximately 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 patients with RAA who develop symptoms is 5.6–25.2%[2,11,15].

In our cohort, 56.2% of cases underwent intervention, a decision based on the presence of symptoms and/or observation on CT of significant airway and/or esophageal compression. This is higher than the 41% intervention rate reported by Guo *et al.*[3] and lower than the 87% rate reported by Vigneswaran *et al.*[9] (Table 4). This may be explained by the fact that in the study of Guo *et al.*[3], none of the asymptomatic patients had significant tracheal or esophageal compression, while 13/47 (27.7%) patients in the study of Vigneswaran *et al.*[9] were asymptomatic with an abnormal tracheal appearance. Furthermore, we did not find a statistically significant relationship between the type of DAA and outcome. Based on our study and the available literature, we can conclude that around 50–80% of DAA cases require surgical intervention.

In the present study, only a small number of neonates (around 5%) developed symptoms significant enough to warrant neonatal intervention. While Vigneswaran *et al.*[9] recorded symptoms in 21% of patients from birth, it is not clear whether all these cases required surgical intervention during the neonatal period, and 40% of these neonates had additional pathology that may have contributed to respiratory distress. Based on our findings, we recommend that patients with an antenatal diagnosis of DAA should be delivered at hospitals with available neonatal support. A small percentage of patients will develop significant symptoms during the neonatal period and will require urgent assessment. For the many neonates who are asymptomatic, we recommend early review by a pediatric cardiologist, since DAA is the tightest type of vascular ring and even asymptomatic patients can have significant airway compression. CT should be performed on all patients irrespective of the presence of symptoms, because the absence of symptoms does not exclude significant airway compression that may warrant intervention.

Study limitations

The limitations of this study include its retrospective nature, the fact that genetic testing and CT imaging had not been performed in all patients by the conclusion of the study and that 15% of patients had a follow-up time period of less than 1 year.

Conclusions

In summary, DAA can be diagnosed easily in midgestation because, in most patients, both arches are patent at this stage. Postnatally, we found that the LAA had become atretic in most patients, supporting the theory of differential growth of the arches during pregnancy. When RAA is diagnosed postnatally, 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 with and offered to affected families. There is no evidence of a statistically significant relationship between the type of DAA and postnatal outcome. The recommendations for these patients are delivery at a hospital with available neonatal support, early postnatal pediatric cardiology review and performance of CT, irrespective of the presence of symptoms.

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