Fetal double aortic arch: prenatal sonographic and postnatal computed tomography angiography features, associated abnormalities and clinical outcomes

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Research article

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Abstract

Background: Fetal double aortic arch (DAA) malformation is a rare congenital heart disease. There is a lack of large sample on comparative study of prenatal ultrasound diagnosis with postnatal computed tomography angiography (CTA) diagnosis, surgery, and autopsy results.

Methods: A retrospective cohort study was carried out on cases of fetal DAA that underwent a comprehensive ultrasound examination and were seen during a six-year period at seven tertiary referral centers.

Results: A total of 36 cases out of 40 prenatally diagnosed DAA fetuses were confirmed by postnatal diagnosis (fetal autopsy, CTA, and surgery). In this cohort of 36 confirmed cases, 24 (67%) were isolated anomalies, while 12 (33%) were associated with intracardiac or extracardiac anomalies, and 2 (6%) had a 22q11.2 chromosome deletion. There were 9 cases of pregnancy termination with a fetal autopsy. Among the remaining 27 live births, 16 (59%) were asymptomatic and 11 (41%) received surgical treatment due to tracheal or esophageal compression symptoms, all with satisfactory outcomes. Prenatal echocardiography showed that DAA was mainly characterized by a bifurcation of the ascending aorta into the right and left aortic arch and the formation of a complete O-shaped vascular ring around the trachea on the three-vessel tracheal view. A variant in the aortic arch branching pattern was found for the first time. The airway obstruction, branching pattern and atretic arch of DAA were clearly shown by postnatal CTA.

Conclusions: Fetal DAA has unique features on prenatal echocardiography and postnatal CTA, and steps and tips for systematic prenatal examination are required. The clinical outcomes of isolated DAA are favorable, even if surgery is performed due to symptoms. Determining whether other malformations or chromosomal anomalies exist is crucial for prognosis evaluation and prenatal decision-making.

Background

A right aortic arch is a type of congenital aortic arch anomaly, and the prevalence is estimated to be 0.1% [1]. Among them, a double aortic arch (DAA) is much rarer, affecting approximately 0.005% ~ 0.007% of fetuses [2, 3]. If the growth of the bilateral fourth arch and dorsal aorta persists, a DAA forms [4]. DAA refers to the continuity of the left and right aortic arch. After arising from the ascending aorta, the two arches surround the trachea and esophagus to form a complete vascular ring and drains into the descending aorta. Generally, the left aortic arch gives rise to the left common carotid artery (LCCA) and the left subclavian artery (LSA), and the right aortic arch gives rise to the right common carotid artery (RCCA) and the right subclavian artery (RSA) [5]. Although the two arches are symmetrical, most cases will have one that is larger and extends higher toward the head. In rare cases, there is atresia of the arch, usually the left arch. The atretic segment is usually located at the distal end of the LSA but may also occur between the LCCA and LSA [6]. Generally, only one arterial duct is open, predominantly on the left, but a right or bilateral arterial duct has also been documented [7]. The descending aorta is usually located...
on the same side of the unobstructed arterial duct and opposite to the dominant aortic arch [8]. Because of the vascular ring that encircles the trachea and esophagus, although some fetuses may be asymptomatic after birth, some may suffer from wheezing, dyspnea, dysphagia and other related compression symptoms [9].

Fetal DAA is generally diagnosed based on the characteristic complete vascular ring in the three-vessel tracheal view on ultrasound, and postnatal diagnosis can be confirmed by computed tomography angiography (CTA) [2, 10]. At present, reports in the literature on DAA are mostly about children or adults, and the reports addressing fetuses all have small sample sizes, with the largest study comprising only 9 cases [11]. Due to the few studies with large sample sizes and the lack of detailed systematic examination approaches, an accurate and comprehensive prenatal diagnosis of DAA remains difficult, and misdiagnoses or missed diagnoses can easily occur. In our study, we enrolled 36 cases of postnatally confirmed DAA from seven tertiary referral centers, making our study the largest series to date. A retrospective analysis was performed to summarize the prenatal diagnostic approaches of DAA and describe the features of postnatal CTA diagnosis to assist in prenatal consultations and clinical evaluations.

Methods

All cases of DAA in the fetal ultrasound examination databases of seven tertiary referral centers for prenatal diagnosis from January 2013 to December 2018, were retrospectively retrieved. Prenatal and postnatal medical records, including echocardiographic and CT scan images, videos and reports, fetal autopsy findings, neonatal records of the newborns, and operation records, were reviewed. This study was approved by the hospital ethics committee, and informed consent was obtained from pregnant women.

The high-quality GE Voluson E10, GE Voluson E8 Expert, and Toshiba Aplio500 Color Doppler ultrasound diagnostic instruments were used, and the probes equipped were RM6C, RAB4-8-d, C1-5-D, and PVT-375BT. Imaging was performed with fetal OB examination mode with the thermal index and medical index each set at < 1.0 and the ALARA principle was followed. Each pregnant woman was positioned in the supine or lateral position, underwent a routine obstetric ultrasound examination and then underwent a fetal cardiac assessment in which a complete fetal echocardiography by a fetal echocardiography expert was performed based on the ISUOG Practice Guidelines (updated) for sonographic screening examination of the fetal heart [12]. Volume sonography (spatiotemporal image correlation (STIC)) could also be incorporated into a more detailed anatomical and functional assessment of the fetal heart if necessary, as previously described [13]. Under normal circumstances, the three-vessel tracheal view shows that the aortic arch and ductus arteriosus together form a V shape, converging into the descending aorta and that the trachea is located behind the right arch, with no vascular constriction in front of the trachea. The diagnosis of DAA was made on the presence of two aortic arches, one on each side of the trachea, and forms a complete vascular ring, joining posteriorly to the descending aorta, with the common carotid and subclavian arteries arising separately and symmetrically, one from each arch. After
the determination of DAA malformation, the inner diameter of the aortic arch was measured. According to the Backer's classification standard, DAA was divided into the right arch dominant type, left arch dominant type and double arch balanced type [14]. On this basis, we set the ratios of the inner diameter of the right arch to that of the left arch as follows: between 0.9 and 1.1 for a double arch balanced type, greater than 1.1 for a right arch dominant type and less than 0.9 for a left arch dominant type.

For fetuses with ultrasonographic findings of DAA, careful examination was carried out to determine whether there was any other intracardiac or extracardiac malformation. All parents received detailed counseling regarding the diagnosis and therapeutic options after the ultrasound scan. Chromosome karyotype analysis by amniotic fluid or umbilical cord blood puncture was suggested. With the consent of the parents of the fetuses, autopsy and pathological examinations were performed for those who chose to terminate their pregnancies, and follow-up was conducted after delivery for those who continued their pregnancies.

For every pediatric patient, multidetector-row computed tomography (MDCT) angiography examinations were performed with a 320-detector volume CT system (Aquilion ONE, Toshiba, Japan) or a 256-row MDCT (GE Revolution, USA). Patients fasted for 46 h and were then anesthetized (chloral hydrate: 0.5 mg/kg). Imaging data were acquired after an intravenous injection of 1.5-2 ml/kg nonionic iodinated contrast agent (iopromide; Ultravist; Schering AG, Berlin, Germany) at a rate of 2-2.5 ml/s. For three-dimensional image reconstruction, the raw MDCT data were processed on a separate workstation (VITREA or Advanced Workstation 4.7, GE Revolution) with multiplanar reformatting (MPR), maximum intensity projection (MIP), minimum intensity projection (MinIP) and volume rendering (VR).

The data are presented as the mean ± standard deviation (SD) or frequency distributions (%).

**Results**

1. **Associated abnormalities and pregnancy outcomes of DAA fetuses**

A total of 358,815 fetuses with complete prenatal ultrasound data from seven centers during six years were analyzed, among which 40 cases were diagnosed with DAA prenatally and 36 cases had postnatal confirmation (9 cases by autopsy (Fig. 1) and 27 cases by CTA; 11 cases were further diagnosed by cardiac surgery), with an incidence of 0.01% (36/358,815). There were two misdiagnosed cases, a right aortic arch with mirror-imaging branching and left posterior ductus arteriosus connecting descending aorta (MRAA-LPDA-DAO) and a right aortic arch with left posterior ductus arteriosus and aberrant left subclavian artery (RAA-LPDA-ALSA), respectively. Besides, there was one case of pregnancy termination that lacked an autopsy and one case was lost to follow-up (Tab.1). The mean maternal age was 29 (range, 23-35) years, and the mean gestational age (GA) was 27 (range, 23-31) weeks. According to the Backer's classification and specific standards mentioned above, among the 36 fetuses, 25 (69%) were right arch dominant, including 2 with an atretic left arch; 5 (14%) were left arch dominant, and 6 (17%) were double arch balanced (Tab.2). All cases had left-sided ductus arteriosus. Of the 36 DAA cases, 24
(67%) were isolated abnormalities, 11 (31%) were associated with intracardiac malformations, and 5 (14%) were associated with extracardiac malformations, among which 4 (11%) had intracardiac and extracardiac malformations simultaneously (Tab.2, 3). Karyotyping was performed in 33 cases, and there were two cases of chromosome 22q11 deletion. Finally, 9 pregnancies were terminated, and 27 fetuses were delivered alive. Of the 27 live births, 16 (59%) were asymptomatic during the study period, and 11 (41%) suffered from varying degrees of stridor, dyspnea, recurrent upper respiratory tract infections and dysphagia for surgical treatment. The mean duration of follow-up was 38.0 ± 17.0 months and the mean age for symptom occurrence was 4.0 ± 2.8 months (Tab.2).

Table 1. Diagnostic accuracy of prenatal ultrasound in the fetal DAA cohort (n=40)

|             | N (%) |
|-------------|-------|
| Confirmed diagnosis | 36 (90%) |
| ▲ Autopsy | 9/36 (25%) |
| ▲ Live births | 27/36 (75%) |
| ▲▲ CTA | 27/27 (100%) |
| ▲▲ CTA, and surgery | 11/27 (41%) |
| Misdiagnosis | 2 (5%) |
| ▲ MRAA- LPDA-DAO | 1 (3%) |
| ▲ RAA-LPDA-ALSA | 1 (3%) |
| Lost to follow-up | 1 (3%) |
| Lack of autopsy | 1 (3%) |

*Note: DAA, double aortic arch; CTA, computed tomography angiography; MRAA- LPDA-DAO, right aortic arch with mirror-image branching and left posterior ductus arteriosus connecting descending aorta; RAA-LPDA-ALSA, right aortic arch with left posterior ductus arteriosus and aberrant left subclavian artery.

Table 2. The features and outcomes of the fetal DAA group with a postnatal confirmed diagnosis (n=36)
### Table 3. Associated anomalies in the fetal DAA group with a postnatal confirmed diagnosis (n=36)

| Characteristics                          | Mean±SD or N(%) |
|------------------------------------------|-----------------|
| Maternal age, y                          | 29.0 ± 6.0      |
| Gestation age at diagnosis, wks          | 27.0±4.1        |
| Follow-up period, months                 | 38.0 ± 17.0     |
| Age for symptoms, months                 | 4.0 ± 2.8       |
| Backer's Classification                   | /               |
| ▲ Right arch dominant                    | 25 (69%)        |
| ▲▲ Atretic left arch                     | 2 (6%)          |
| ▲ Left arch dominant                     | 5 (14%)         |
| ▲ Double arch balanced                    | 6 (17%)         |
| Isolated DAA                             | 24 (67%)        |
| Intracardiac anomalies                   | 11(31%)         |
| Extracardiac anomalies                   | 5 (14%)         |
| Chromosomal abnormalities                | 2/33 (6%)       |
| TOP                                      | 9(25%)          |
| Live births                              | 27 (75%)        |
| ▲ Asymptomatic                           | 16/27 (59%)     |
| ▲ Symptomatic for surgery                | 11/27 (41%)     |

*Note: DAA, double aortic arch; TOP, termination of pregnancy*
| Condition                                      | N (%) |
|-----------------------------------------------|-------|
| **Cardiac**                                   |       |
| ▲ Ventricular septal defect                    | 5 (14%) |
| ▲ Double outlet right ventricle                | 4 (11%) |
| ▲ Permanent left superior vena cava           | 4 (11%) |
| ▲ Left ventricular dysplasia                   | 1 (3%) |
| ▲ Partial atrioventricular septal defect       | 1 (3%) |
| ▲ Mirror image dextrocardia                    | 1 (3%) |
| ▲ Dextrocardia                                | 1 (3%) |
| **Extracardiac**                              |       |
| ▲ Gastrointestinal tract                       | 2 (6%) |
| ▲ Thorax                                      | 2 (6%) |
| ▲ Central nervous system                       | 1 (3%) |
| ▲ Facial                                      | 1 (3%) |
| ▲ Spine                                       | 1 (3%) |

*Note: DAA, double aortic arch.

2. **Main echocardiography characteristics of DAA fetuses**

The main echocardiography characteristics of DAA are as follows: (1) A bifurcation of the ascending aorta into the right and left arch is confirmed on the left ventricular outflow tract view and the three-vessel tracheal view (Fig.2a, 2b); (2) For a right-sided arch, an aortic arch is visible on the right side of the trachea on the three-vessel tracheal view (Fig.2b); (3) The three-vessel tracheal view can show that the left and right arches form a complete O-shaped vascular ring around the trachea and that the ductus arteriosus is connected to it to develop a 9-shaped configuration (Fig.2b). However, if an arch is atretic, the color flow is interrupted and the O-shaped vascular ring is not completed, and a fibrous cord could be seen (Fig.3); (4) The three-vessel tracheal view or sagittal view may show that both the left and right arches converge into the descending aorta (Fig.2b, 4a, 4b); (5) The sagittal view shows two aortic branches (the LCCA and LSA) of the left arch and two symmetrical branches (the RCCA and the RSA) of the right arch (Fig.4a, 4b); (6) The coronal view of the aortic arch shows the symmetric appearance of the common carotid artery and subclavian artery originating from the ipsilateral aortic arch simultaneously (Fig.4c). However, a variant in the aortic arch branching pattern was found in the examination, namely, only one branch of the left aortic arch and three branches of the right aortic arch (Fig.5); and (7) Three-
dimensional color-rendered images with spatiotemporal image correlation may help display the spatial structure of the complete vascular ring and branches((Fig.6).

3. Postnatal CTA results and main features

A total of 29 infants underwent CTA between 1 week and 1 year after birth. Twenty-seven cases were confirmed to be DAA, including 2 cases with atresia of the left arch. Two cases were found to be an MRAA-LPDA-DAO and an RAA-LPDA-ALSA. The main features of DAA on CTA are as follows: (1) On the transaxial view with MIP, the ascending aorta divides into two arches surrounding the trachea and esophagus, which mimic the prenatal three-vessel tracheal view on ultrasound (Fig.7a); (2) VR image processing for great vessels clearly shows the double arches forming a complete vascular ring and both connecting with the descending aorta and two branches arising from each arch (Fig.7b), and a proximal posteroinferiorly distorted LSA and descending aortic diverticulum indicates a DAA with an atretic left aortic arch (Fig.7c); and (3) MPR or MinIP or VR image processing for the airway can accurately demonstrate the site and severity of the tracheal obstruction (Fig.7d).

Discussion

To the best of our knowledge, this is the largest cohort to describe the prenatal diagnostic approaches and clinical prognosis of fetuses with DAA. Our study is the first to report a new branching pattern variant of DAA. In contrast to the usual pattern wherein each aorta arch gives rise to two branches, in this case, only one branch originates from the left aortic arch, namely, the LCCA, but three branches originate from the right aortic arch, namely, from proximal to distal, the RCCA, RSA, and LSA. However, additional similar cases are needed to confirm this discovery.

DAA is a mostly isolated abnormality but can also be associated with other abnormalities. We found that 31% of cases had DAA with intracardiac malformations, in contrast to 16.6% of cases in another report [8], with ventricular septal defect, double outlet right ventricle, and persistent left superior vena cava being the most common. Extracardiac malformations were rarely mentioned previously, but 5 cases were found in this study, highlighting that a careful and thorough fetal examination outside of the cardiovascular system is also needed. The two cases (6%) of 22q11.2 chromosome deletion were both accompanied by thymus dysplasia. The prevalence of chromosome 22q11.2 microdeletions (DiGeorge syndrome) in fetuses with right aortic arches has been reported to be between 6.1% and 10% [15–17], but the correlation with DAA seems to be lower [18]. Ultrasound assessment of dysplasia or the absence of a fetal thymus is useful for predicting 22q11.2 microdeletion [19]. Therefore, for fetuses diagnosed with DAA by ultrasound, thymus size assessment should be performed routinely. In this study, 9 women had terminations of pregnancy, among which 7 were associated with intracardiac or extracardiac malformations or 22q11.2 microdeletion, indicating that whether DAA is associated with other malformations or chromosomal abnormalities has an important impact on pregnancy outcomes and maternal decision-making.
Based on the abovementioned sonographic characteristics of DAA, most DAAs can be diagnosed by prenatal echocardiography. The three-vessel tracheal view is the most characteristic view for DAA diagnosis, but it alone is not enough, especially when an arch shows atresia, in which a vascular ring is not always typical as a result of an interruption in the blood flow of the atretic segment and then differential diagnosis with a right aortic arch is challenging. At this time, multiple views should be performed to better observe the origin, course, and branches of the two arches. The following steps and key points for systematic examination are recommended. (1) Whether the right aortic arch is present on the three-vessel tracheal view should be confirmed. (2) A search for bifurcation of the ascending aorta in the left ventricular outflow tract view is needed; it is necessary to trace the ascending aorta far enough into the arch as the bifurcation is usually not at the origin of the ascending aorta. (3) The three-vessel tracheal view and sagittal view of the aortic arch should be used to confirm that the left and right arches both arise from the ascending aorta and are connected to the descending aorta. (4) The three-vessel trachea view should be used to find the left and right arches surrounding the trachea and esophagus to form a complete O-shaped vascular ring; because the two arches may not be at the same level, the probe will need to be tilted slightly. (5) To determine the type of DAA, the inner diameter of the left and right arches need to be accurately measured. (6) The branching patterns of the left and right aortic arches on the sagittal views and coronal views of the aortic arch need to be confirmed. (7) Careful examination of the heart and other systems, including the thymus, should be performed to determine the existence of associated intracardiac or extracardiac malformations. (8) Chromosome and gene tests should be performed when necessary. Besides, adjusting the parameters of the color Doppler mode is necessary to reduce the scale of blood flow velocity to display a small nondominant arch fully. It is difficult to identify the fibrous cord formed by partial atresia of the nondominant arch by ultrasound examination, so the vascular ring needs to be diagnosed indirectly through the formation of a blind end or diverticulum of the arch. When the left arch is small and difficult to display, the only visible right arch should not be mistaken for the right pulmonary artery, which would also be going in the right direction.

Regarding cases before birth, very few reports have suggested that tracheal compression by the complete vascular ring of the DAA could lead to CHAOS (congenital high airway obstruction syndrome), which can cause intrauterine fetal respiratory distress or stillbirth [11, 20]. However, the sonographic characteristics for predicting perinatal complications of DAA, especially for tracheal compression requiring airway ex utero intrapartum therapy (EXIT), are not clear. In our study, none of the fetuses showed significant airway obstruction before delivery, consistent with most reports.

After birth, some DAA patients have compression due to the vascular ring surrounding the trachea and esophagus, resulting in wheezing, dyspnea, dysphagia and other symptoms, most of which occur within the first year. Currently, it is believed that surgical treatment is necessary for patients with respiratory or digestive symptoms [9, 21]. The proportion of children with symptomatic DAA is reported to be approximately 72.4% [8]. However, during the follow-up after birth in this study, 16 cases (59%) were asymptomatic, and 11 symptomatic cases (41%) underwent surgical treatment. All of them survived with good prognoses.
Echocardiography is considered to be the first-line postnatal imaging method for DAA. However, rings with special anatomical features, such as a fibrous cord and tracheal compression, cannot be recognized by ultrasound, and pulmonary air easily interferes with image quality. Therefore, MRI or CT is considered to be the gold standard for identifying such variations. However, MRA may require prolonged sedation of pediatric patients [22]. Moreover, image reconstruction, density, and time resolution are worse with MRA than with CT. Compared to MRA, MDCT is a superior imaging modality, as it requires less time for a child to calm down and provides more detailed information, including vascular structures and spatial relationships with adjacent organs, especially the airways and esophagus [23]. MDCT combined with various postprocessing options, such as VR, MIP, MinIP, and MR, can display clear details of the compressed trachea, esophagus, and vascular ring, even in cases in which the ring comprises the atretic aortic arch and arterial ligament. Generally, evidence of inferior and posterior convexity of the initial course of LSA and a descending aortic diverticulum suggests the presence of an imperforate vessel or fibrous cord connecting the structures of the atretic aortic arch [24].

The two cases of misdiagnosis in this study should be noted. Except for the familiar RAA-LPDA-ALSA forming a U-shaped vascular ring, DAA especially the dominant right arch type should also be identified with MRAA-LPDA-DAO. When the left arch is small and tortuously curved, it is easily mistaken for the left innominate artery, and the O-shaped vascular ring is not obvious and typical. Repeated multiple cross-sectional examinations show that MRAA-LPDA-DAO fails to demonstrate the connection between the left branch of the aortic arch (the left innominate artery) and the ductus arteriosus or descending aorta. However, in DAA, the left arch is usually connected with the left ductus arteriosus and descending aorta.

The limitations of this research are the retrospective nature of the analysis and the incidence of DAA, which was 0.01%, higher than that in the unselected population according to the literature (0.005%-0.007%) [2, 3]. This discrepancy may be because the research centers in this study were provincial or municipal tertiary referral centers, where many women with high-risk pregnancies or who were suspected of a fetal heart anomaly were referred and selectively examined. Moreover, since we included only fetuses with prenatal diagnoses of DAA, false-negative diagnoses could not be derived and analyzed.

**Conclusions**

In summary, DAA has clear characteristic manifestations on fetal echocardiography. A detailed and systematic prenatal ultrasound examination can effectively improve the accuracy and comprehensiveness of the prenatal diagnosis and assist in prenatal counseling and clinical evaluations. CTA may be the optimal imaging method to evaluate DAA postpartum.

**Abbreviations**

DAA
Declarations

Ethics approval and consent to participate

The study was approved by the ethics committee of The Second Xiangya Hospital of Central South University in China, Xiangya Hospital of Central South University in China, and the First Affiliated Hospital of University of South China, the First People's Hospital of Yueyang, Maternal & child health center of Changsha, Zhuzhou and Changde. Written informed consent was obtained from all of the families.

Consent for publication

Written informed consent was obtained from parents for publication of clinical details, clinical images.

Availability of data and materials

All data generated or analysed during this study are included in this published article. The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors’ contributions

QG and QCZ designed the whole study. QG, YFK, and QCZ drafted the manuscript. QG, SZ, JWZ, JZ, HXY, LW, LLT, AJY and QCZ performed the fetal echocardiography and fetal ultrasound. QLS performed the
MDCT. QG, YFK and XFW made the data analysis. All authors read and approved the final manuscript.

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Figures
Figure 1

Fetal anatomical pathology after termination of pregnancy. a. The right and left aortic arches both arise from the ascending aorta. The left aortic arch gives rise to the LCCA and LSA, and the right aortic arch gives rise to the RCCA and RSA. The left ductus arteriosus connects to descending aorta. b. After the trachea is cut off, the left and right arches form a complete vascular ring, which is connected to the descending aorta. (AAO: ascending aorta; R: right aortic arch; L: left aortic arch; RCCA: right common carotid artery; RSA: right subclavian artery; LCCA: left common carotid artery; LSA: left subclavian artery; PA: pulmonary artery DA: ductus arteriosus; DAO: descending aorta; T: trachea).
Figure 2

Bifurcation of the ascending aorta and complete vascular ring of the DAA on fetal echocardiography. a. Left ventricular outflow tract view: The distal ascending aorta bifurcation is confirmed as the right arch and left arch. b. Three-vessel tracheal view: A bifurcation of the ascending aorta into the right aortic arch and left aortic arch to form a complete O-shaped ring encircling the trachea, together with a number 9 configuration connecting with the left-sided ductus arteriosus. Both aortic arches demonstrate antegrade blood flow. (AO or AAO: ascending aorta; R: right aortic arch; L: left aortic arch; PA: pulmonary artery; DA: ductus arteriosus; T: trachea).
Figure 3

DAA with distal left aortic arch atresia on fetal echocardiography. a. Nonstandard three-vessel tracheal view of gray-scale imaging: The vascular echo at the distal end of the left aortic arch is interrupted and replaced by a fibrous cord (arrowhead). B. Nonstandard three-vessel tracheal view of color Doppler imaging shows the interruption of color flow at the distal end of the left aortic arch (arrowhead) and an incomplete vascular ring. (L-ARCH: left aortic arch; DAO: descending aorta; T: trachea).
Figure 4

Branching pattern of DAA on fetal echocardiography. a. Sagittal view of the right aortic arch: The dominant right arch gives rise to the RCCA and RSA. b. Sagittal view of the left aortic arch: The small left aortic arch gives rise to the LCCA and LSA. c. The coronal view of the aortic arch shows the symmetric appearance of the common carotid artery and subclavian artery originating from the ipsilateral aortic arch simultaneously. (R-ARCH: right aortic arch; L-ARCH: left aortic arch; RCCA: right common carotid artery; RSA: right subclavian artery; LCCA: left common carotid artery; LSA: left subclavian artery).
Figure 5

Branching pattern variant of DAA on fetal echocardiography. A. Sagittal view of the left aortic arch: One branch can be seen from the left aortic arch (H: heart, 1: left common carotid artery). Sagittal view of the right aortic arch: Three branches can be seen from the right aortic arch (H: heart, 1: right common carotid artery, 2: right subclavian artery, 3: left subclavian artery, L-ARCH: left aortic arch; R-ARCH: right aortic arch; DAO: descending aorta).
Figure 6

Three-dimensional color-rendered image with spatiotemporal image correlation of DAA: The left aortic arch and the dominant right aortic arch develop a complete vascular loop and join the left ductus arteriosus and converge into the descending aorta together. Symmetrical initial parts of two branches can probably be seen on each arch. (R-ARCH: right aortic arch; L-ARCH: left aortic arch; RCCA: right common carotid artery; RSA: right subclavian artery; LCCA: left common carotid artery; LSA: left subclavian artery; DA: ductus arteriosus; DAO: descending aorta)
Figure 7

Postnatal confirmed diagnosis of DAA by multidetector-row CTA with volume-rendering processing. a. Transaxial view: The ascending aorta divides into two arches surrounding the trachea and esophagus. b: Left view of volume-rendered image: The spatial structure mimics Fig 6, but the branches (LCCA and LSA originating from the left arch, RCCA and RSA originating from the right arch) and the surrounding of the trachea and esophagus appear clearer. c. Proximity of the posteroinferiorly distorted left subclavian artery (arrow) and descending aortic diverticulum (*) suggests a possible fibrous connection (arrowhead) between the two structures and demonstrates a double aortic arch with an atretic left aortic arch distal to the origin of the left subclavian artery. d. A compressed trachea by the vascular ring is shown (arrowhead) (AAO: ascending aorta; R or R-ARCH: right aortic arch; L or L-ARCH: left aortic arch; RCCA: right common carotid artery; RSA: right subclavian artery; LCCA: left common carotid artery; LSA: left subclavian artery; DAO: descending aorta; T: trachea; LPB: left principal bronchus, RPB: right principal bronchus)