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Case Report

Prenatal aortic arch development in double aortic arch: Understanding postnatal closure of left aortic arch: A case report ☆,☆☆

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ABSTRACT

Double aortic arch (DAA) is a rare congenital abnormality characterized by a vascular ring that often requires surgical intervention due to respiratory complications. The DAA and right aortic arch with mirror-image branches (RAA-MB) represent abnormalities in development of the aortic arch. However, prognosis differs significantly, as the DAA forms vascular rings, whereas the RAA-MB typically does not. Distinguishing between the conditions becomes particularly challenging in cases of DAA with closure of the posterior portion of the left aortic arch (LAA) because the postnatal manifestations closely resemble those of RAA-MB. Herein, we present a case of DAA in which longitudinal observation of the LAA and RAA diameters during pregnancy aimed in predicting postnatal closure of the LAA. A 37-year-old female with suspected DAA was referred to our hospital at 26 weeks of gestation. Initial measurements revealed comparable diameters for the LAA and RAA; however, the LAA diameter decreased to approximately half that of the RAA by term owing to growth restrictions. Postnatal contrast computed tomography confirmed the closure of the posterior portion of the LAA and RAA with Kommerell diverticulum. Our findings suggest that careful monitoring of DAA throughout fetal development, especially during the third trimester, may aid in predicting atretic changes in the nondominant arch after birth, allowing an easy distinction between the DAA and RAA-MB after birth.

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Introduction

The double aortic arch (DAA) is a rare congenital anomaly affecting approximately 0.005% of fetuses (Fig. 1) [1,2], characterized by a vascular ring. The vascular ring may cause respiratory failure due to stenosis of the tracheal cartilage ring, which may manifest as wheezing, dyspnea, and dysphagia; most of these complications occur within the first year of life [3]. Approximately 50%–75% of infants with symptomatic DAA are reported to require surgical intervention to relieve tracheal compression causing respiratory or digestive symptoms [4,5]. Therefore, prenatal diagnosis of DAA is crucial.

The normal left aortic arch (LAA) develops when the right aortic arch (RAA) developing between the right subclavian artery and the descending aorta regresses during embryonic development (Fig. 1A). In the DAA, both the RAA and the LAA persist from the ascending aorta. These structures pass through either side of the trachea and esophagus and converge posteriorly to form a descending aorta (Fig. 1B). Although the LAA is not patent in an incomplete DAA with LAA atresia, this situation results in a vascular ring composed of a fibrous cord secondary to atresia of the distal portion of the LAA (Fig. 1C). An RAA with mirror-image branching (RAA-MB) occurs when the LAA between the left subclavian artery and descending aorta regresses (Fig. 1D). In most cases, involution occurs between the left ductus and descending aorta, without forming a vascular ring. Rarely, involution occurs between the left subclavian artery and ductus, creating a vascular ring that may cause respiratory symptoms [6,7].

Due to the existence of a fibrous cord, a vascular ring forms in patients with DAA and LAA atresia (Fig. 1C) [8]. In contrast, the RAA-MB does not usually form this ring (Fig. 1D). However, after birth, the distinction between the two conditions becomes particularly difficult in cases of DAA with closure of the posterior portion of the LAA, because the postnatal manifestations are similar to those of RAA-MB [4,7,8].

A previous report suggested that accurate measurement of the inner diameter of both the LAA and RAA is crucial for determining the type of DAA [4]. However, longitudinal observations of the diameters of the LAA and RAA during fetal development in the DAA have not been reported. In this study, we present a case of DAA in which longitudinal observation of the LAA and RAA diameters throughout pregnancy predicted LAA closure after birth.

Case report

A 37-year-old woman (primigravida) conceived via in vitro fertilization. DAA was suspected at 23 weeks and the patient was referred for perinatal management at 26 weeks. Fetal ultrasound showed DAA, with an RAA diameter of 3.8 mm, LAA diameter of 3.0 mm, and RAA/LAA diameter ratio (RA ratio) of 1.27 [≤0.9: left dominance, 0.9–1.0: bilateral equality, ≥1.0: right dominance [4]], indicating mild RAA dominance (Figs. 2 and 3). In the present case, the angle between the two aortic arches was 45° (Fig. 2A). During fetal life, the RAA developed well, while the growth of LAA was impaired (Fig. 3). As a result, the RAA diameter was 5.9 mm, the LAA 3.1 mm, and the RA ratio 1.90 at 37 weeks (Fig. 2D). At 40 weeks, labor induction was attempted; however, cesarean section was performed owing to labor arrest. The neonate, a girl weighing 3370 g, was born with an Apgar score of 8 and 8 at 1 and 5 min, respectively. She initially showed cyanosis, but her condition improved immediately with routine care. A postnatal echocardiography, performed on the day of birth, confirmed the diagnosis of right-dominant DAA with an RAA diameter of 5.6 mm, LAA diameter of 3.5 mm, and RA ratio of 1.6, and blood flow was observed in bilateral AA. A contrast computed

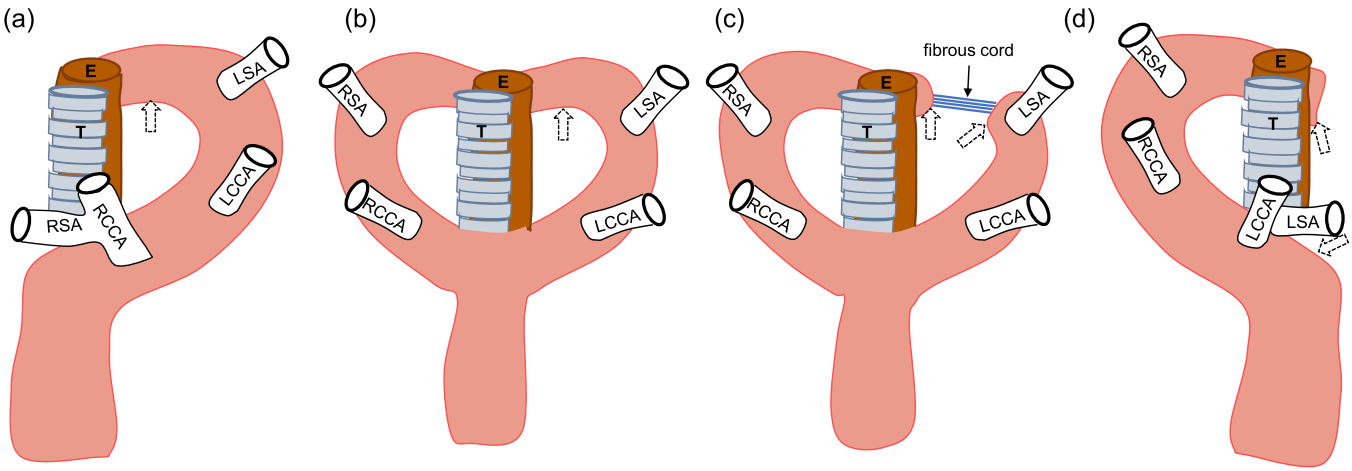

Fig. 1 – Schematics of the normal left aortic arch (A), double aortic arch (B), double aortic arch with distal left arch atresia (C), and right aortic arch with mirror-image branching (D). The dotted arrow indicates the ductus arteriosus after involution. There are 2 patterns of ductus arteriosus location in the double aortic arch with distal left arch atresia (C) and the right aortic arch with mirror-image branching (D).
LCCA, left common carotid artery; LSA, left subclavian artery; RCCA, right common carotid artery; RSA, right subclavian artery; DA, ductus arteriosus; T, trachea; E, esophagus.

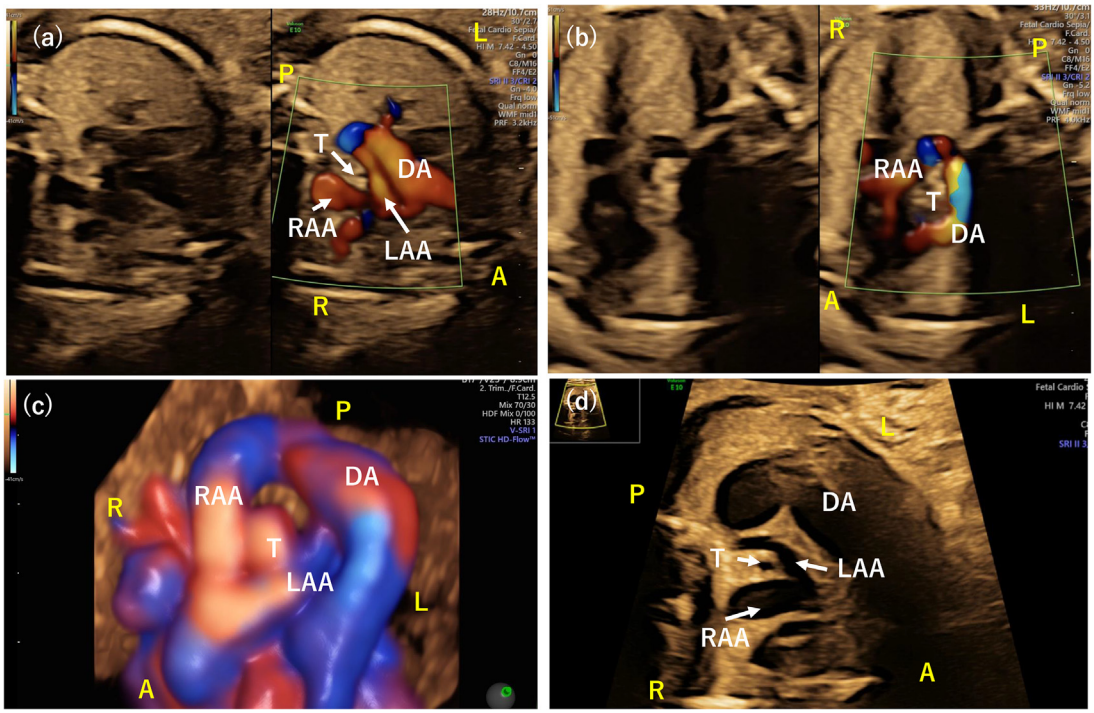

Fig. 2 – Prenatal sonographic findings of DAA. DAA was diagnosed at 26 weeks (A-C). At 26 weeks, the size of the two aortic arches is comparable (A), the angle between the arches is 45° (A), and the O-shaped vascular ring is observed in the three-vessel-trachea view (B). Three-dimensional color-rendered image illustrating the spatial relationships of the great arteries in the DAA (C). At 37 weeks, a right-dominant DAA is observed (D). RAA, right aortic arch; LAA, left aortic arch; DA, ductal arch; T: trachea, A: anterior, P: posterior, L: left.

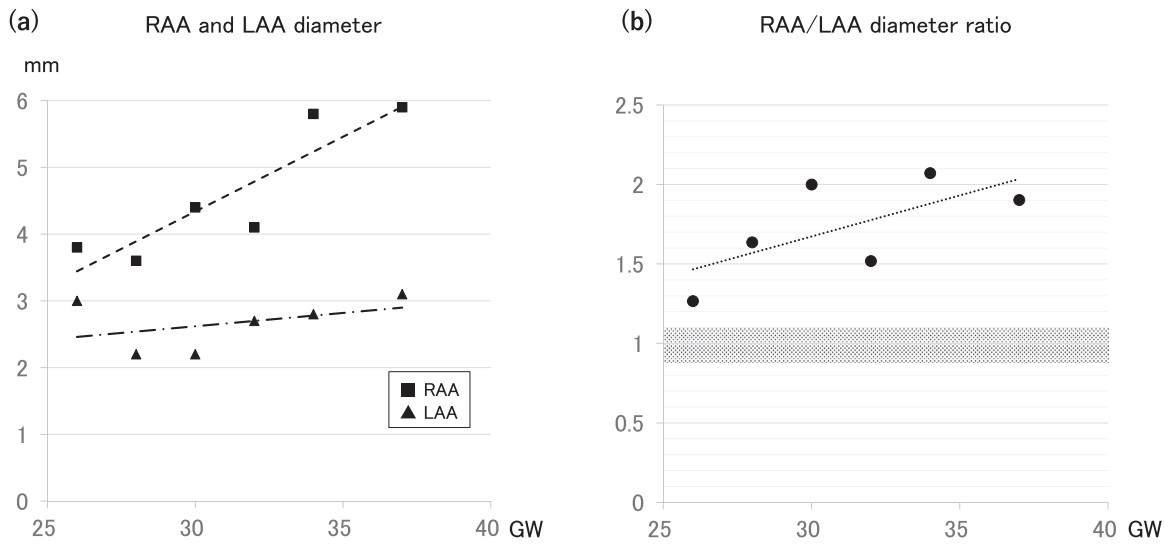

Fig. 3 – Diameters of RAA and LAA (A) and RAA/LAA ratio (B). LAA proper development with RAA growth impairment during fetal life. RAA, right aortic arch; LAA, left aortic arch.

tomography scan on postnatal day four confirmed the closure of the posterior portion of the LAA and RAA with Kommerell diverticulum (Fig. 4). At 1 month of age, she did not show respiratory distress, and surgery was planned after weight gain.

Discussion

In neonates with DAA, the transverse aortic arches are typically unequal in caliber; 70%–90% of individuals have a dom-

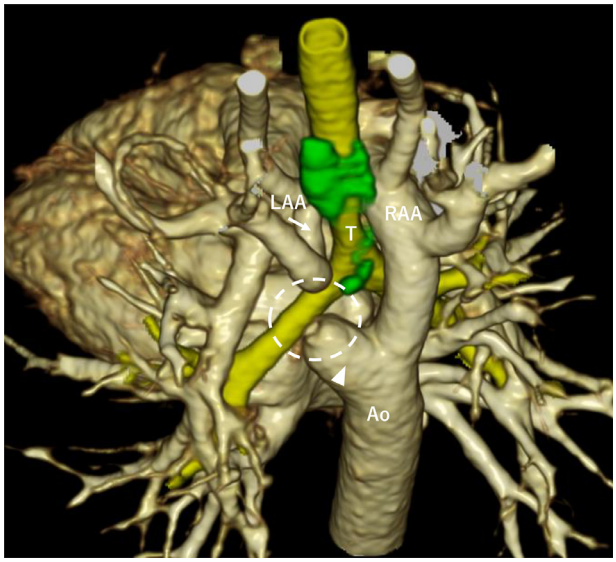

Fig. 4 – Postnatal contrast computed tomography images on postnatal day 4. The LAA is closed at its posterior portion (dotted circle), and RAA with Kommerell diverticulum (arrowhead) is confirmed. RAA, right aortic arch; LAA, left aortic arch; Ao, descending aorta; T, trachea.

inant RAA [3,9], and approximately one-third of the patients have incomplete DAA [9]. In another study, 10 of 66 patients with DAA had Kommerell diverticum, and all of these patients presented with LAA atresia [10]. However, the timing in which differences between the LAA and RAA diameters were observed was not defined. In our patient's case, development of the LAA was markedly impaired during the third trimester, although the LAA and RAA diameters were comparable in the second trimester (Fig. 3). DAA with congenital heart disease, including tetralogy of Fallot, double-outlet right ventricle, transposition of the great arteries, and common arterial trunk, has been described in approximately one-sixth of cases [3,4]; however, no intracardiac malformation was observed in our case. Thus, during pregnancy, attention should be paid to potential intracardiac malformations and differences in the diameters of the 2 aortic arches in patients with DAA. Monitoring these differences might help predict postnatal closure of the LAA.

In this case, we performed ultrasound examinations at 3 key points to achieve accurate prenatal diagnosis of DAA. First, the "O"-shaped vascular ring was observed in the three-vessel-trachea view (Fig. 2B). This formation can be present in DAA, but not in RAA-MB [6]. Second, the angle between the 2 aortic arches was 45° in our case (Fig. 2A). Recently, 2 groups suggested that measuring the angle of the first branch of the aorta helps distinguish between the DAA and RAA-MB [8,11]. Both groups showed that the angle in a fetus with DAA was significantly lower than with RAA-MB, and the recommended cut-off values were 62.8° and 71°, respectively. Finally, three-dimensional sonography with spatiotemporal image correlation illustrated the spatial relationships of the great arteries, supporting the prenatal diagnosis of DAA (Fig. 2C). The main difference between DAA and RAA-MB lies in whether the first branch of the ascending aorta is connected to the descending aorta, however, the 2 arches in DAA may not be at the same level [4,8]. Consistently with previous findings [6], adding three-dimensional modalities to fetal echocardiography improved the diagnostic accuracy of arch anomalies in our case.

In conclusion, prenatal diagnosis of DAA and meticulous follow-up during fetal life, especially during the third trimester, may aid in predicting atretic changes in the non-dominant arch, resulting in an easy distinction between DAA and RAA-MB after birth.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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