

Case Report

Right aortic arch with aberrant left subclavian artery—prenatal diagnosis and evaluation of postnatal outcomes: Report of three cases

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Abstract

Objective: To present prenatal diagnosis of a right aortic arch (RAA) with an aberrant left subclavian artery (LSCA) by the three vessels and trachea (3VT) view using routine ultrasound and the newer technique of three-dimensional power Doppler ultrasound (3D-PDU) together with a discussion of the postnatal outcome.

Case Reports: Three fetuses having an RAA with an aberrant LSCA were diagnosed prenatally between January 2004 and June 2009. They were all detected at between 20 and 24 weeks by routine ultrasound examination at our hospital. All of them were diagnosed via the 3VT view of the fetal upper mediastinum using ultrasound, which revealed an abnormal U-shaped pattern instead of a normal V-shaped confluence; furthermore, 3D-PDU showed an abnormal RAA and an aberrant LSCA with a vascular ring. These abnormalities were not combined with any other congenital cardiac defects in our three cases. A normal chromosome complement was present without microdeletion of chromosome 22q11.2 in all three cases. Two of the cases were genetically assessed prenatally and the other was assessed postnatally. After delivery, diagnosis was confirmed by echocardiography and three-dimensional 64-slices helical computed tomography angiography. Two of the three fetuses were asymptomatic postnatally, whereas one fetus presented with symptoms of tracheoesophageal compression caused by the vascular ring, but this had improved by 8 months of age.

Conclusion: The 3VT view in routine prenatal ultrasound examination is important and essential for the prenatal diagnosis of an RAA with an aberrant LSCA. Moreover, 3D-PDU is able to provide a more clear-cut cardiovascular structure, which helps with the diagnosis.

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Keywords: Aberrant left subclavian artery; Prenatal diagnosis; Right aortic arch; Three-dimensional power Doppler ultrasound; Three vessels and trachea view

Introduction

Right aortic arch (RAA) is one of a number of types of congenital aortic arch abnormalities. It was divided into two types: mirror-image branching and aberrant left subclavian artery (LSCA) [1]. The incidence of RAA is about 0.1% [2,3]. In a normal case, the aorta arises from the left ventricle upward to the right side, becomes aortic arch, and runs backward to the left in front of the trachea, and then left

downward to the diaphragm. The first normal branch is innominate artery, which divides into the right common carotid and right subclavian arteries, whereas the second branch is left carotid artery; and the third is LSCA (Fig. 1). In RAA with aberrant LSCA, the aorta ascends from the left ventricle toward the right side, then turns posterior to the trachea, and in most cases descends left to the diaphragm. The left carotid artery is the first branch of aortic arch and this is followed by the right carotid artery, the right subclavian artery, and an aberrant LSCA (Fig. 2). Thus, a vascular ring is formed around the trachea and esophagus by aortic arch, aberrant LSCA, ductus arteriosus, and pulmonary arterial trunk. This type seldom combines with other congenital cardiac anomalies but may cause a compression of the trachea and esophagus. In

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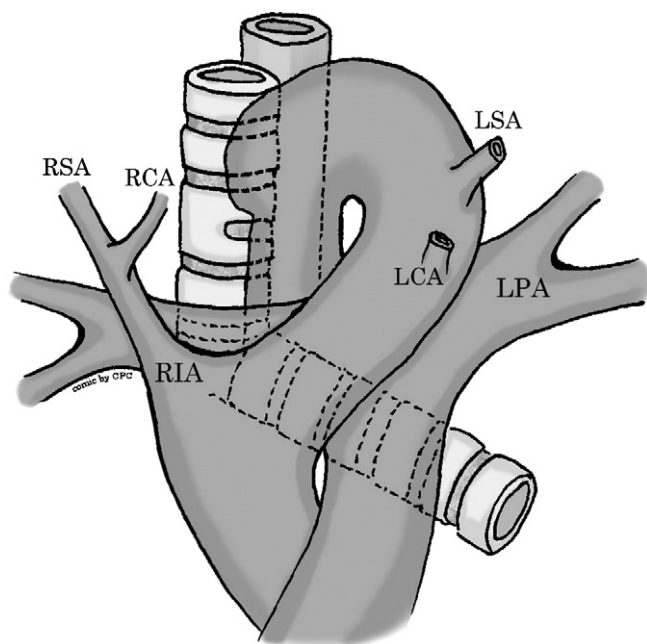


Fig. 1. The anatomy of the normal aortic arch. LCA = left carotid artery; LPA = left pulmonary artery; LSA = left subclavian artery; RCA = right carotid artery; RIA = right innominate artery; RSA = right subclavian artery. Illustration by Pei-Ching Chen.

RAA with mirror-image branching, the aorta derives from the left ventricle upward to the right side, turns downward at the right side of trachea, and descends on the right side or at the middle line. It does not form a vascular ring or sling. The first branch of the aortic arch is the left common innominate artery, which divides into the left common carotid artery and

LSA. This is followed by the right common carotid artery and the right subclavian artery in succession (Fig. 3). This type is commonly associated with other cardiac defects or chromosomal anomalies, such as a microdeletion affecting chromosome 22q11.2 [4].

The prenatal diagnosis of RAA with aberrant LSCA is based on the targeted sonographic three vessels and trachea (3VT) view of the fetal upper mediastinum. The 3VT view is a transverse view of fetal upper mediastinum [5]. In this plane, the superior vena cava, ascending aorta, and main pulmonary artery line up from right to the left. The trachea is located right and posterior to the aorta. In RAA with aberrant LSCA, the 3VT view shows the aortic arch, the proximal part of the aberrant LSCA, the left-sided ductus, and the pulmonary artery, which form a “U” shaped loop around the trachea [2,6]. Three-dimensional Power Doppler ultrasound (3D-PDU) is another method for the prenatal diagnosis of RAA with aberrant LSCA. This allows the three-dimensional reconstruction of the vessels after their visualization using power Doppler ultrasound. It is thus able to reveal an RAA with a vascular ring and an aberrant LSCA [7,8]. We present three cases of prenatal diagnosis of RAA with aberrant LSCA by 3VT view/3D-PDU and follow-up of these cases to their postnatal outcome.

Case reports

We have performed routine prenatal fetal ultrasonographic scans at 20–24 weeks of gestation with a commercially available GE Voluson 730 Expert Ultrasonography System Scanner (GE Medical Systems, Milwaukee, WI, USA) equipped with a multifrequency transabdominal volumetric

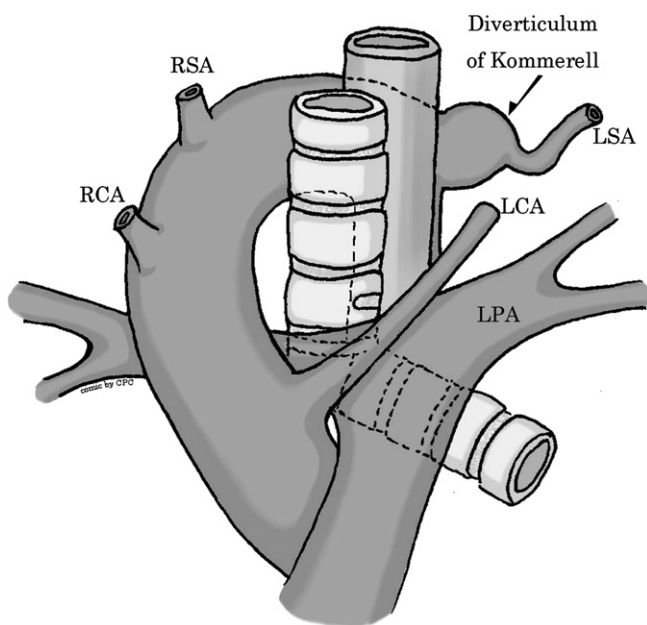


Fig. 2. The anatomy of the right aortic arch with aberrant left subclavian artery. LCA = left carotid artery; LPA = left pulmonary artery; LSA = left subclavian artery; RCA = right carotid artery; RSA = right subclavian artery. Illustration by Pei-Ching Chen.

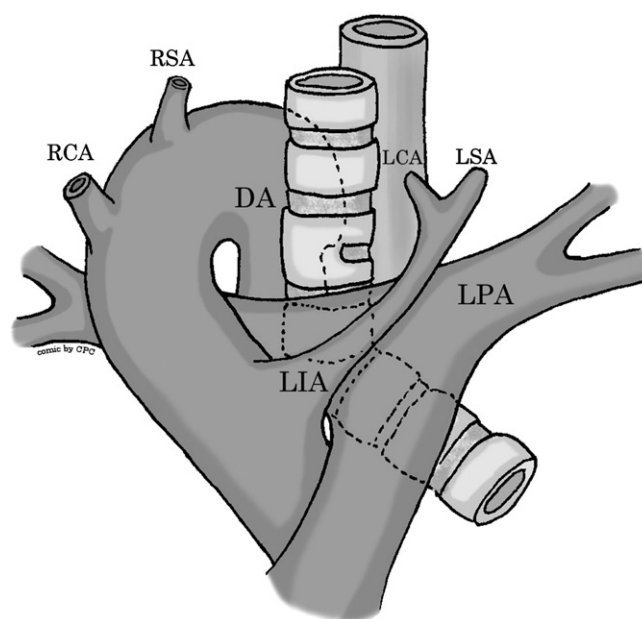


Fig. 3. The anatomy of the right aortic arch with mirror-image branching. DA = descending aorta; LCA = left carotid artery; LIA = left innominate artery; LPA = left pulmonary artery; LSA = left subclavian artery; RCA = right carotid artery; RSA = right subclavian artery. Illustration by Pei-Ching Chen.

probe (4–8 MHz) in our hospital since January 2004. One of the most important parts of the routine ultrasonographic scan is a thorough fetal cardiac examination, which includes the transverse view of the fetal upper abdomen, the four-chamber view, the left ventricular outflow tract view, the right ventricular outflow tract view, the 3VT view, and the longitudinal view of aortic arch [9]. From January 2004 to June 2009, three fetuses having RAA with an aberrant LSCA were found prenatally. The characteristics and postnatal outcome of our three fetuses are summarized in Table 1.

The all three cases had received regular prenatal care at our hospital since early gestation, and the routine ultrasound examination at 20–24 weeks of gestation was performed normally. The ultrasound showed an abnormal 3VT view, but no other cardiac abnormalities or structural anomalies. The 3VT view showed a U-shaped configuration with the trachea inside rather than a normal V-shaped confluence with trachea laterally placed (Fig. 4). At this point, 3D-PDU was performed. We applied the transducer to obtain a coronal view from the ventral side of the fetus and visualized the descending aorta on the left side of trachea using two-dimensional (2D) power Doppler. Then a mechanic sweep was made and this collected a volume acquisition of the 3D vasculature. After reconstruction, an abnormal vessels structure with an RAA and an aberrant LSCA, which formed a vascular ring, was reconstructed (Fig. 5). A diagnosis of fetal RAA with aberrant LSCA was made. Amniocentesis was performed at 21 weeks of gestation in Case 1 and Case 3. Screening for microdeletion of chromosome 22q11.2 was performed by fluorescence *in situ* hybridization. One patient (Case 2) refused amniocentesis because of the risk associated with this procedure. This baby received karyotyping and a fluorescence *in situ* hybridization survey after delivery. A normal karyotype without microdeletion of chromosome 22q11.2 was found in all three cases. We performed serial sonographic follow-up and found that the fetuses were growing normally. All three fetuses were delivered by vaginal birth at term with normal birth body weight, length, and Apgar scores. After delivery, echocardiography and 3D reconstruction using 64-slices spiral computed tomography (CT) angiography confirmed our diagnosis (Fig. 6).

The babies in Case 1 and Case 2 remained well and no complications have been identified so far. One male is now

three and half years of age (Case 1), and the other is one and half years of age (Case 2). They still receive clinical follow-up at our pediatric cardiology department. The fetus in Case 3 presented with stridor and dysphagia after delivery. She had also suffered from recurrent respiratory tract infections (five times) and often needed to receive bronchial secretion drainage. It was suggested that she receive clinical follow-up and conservative management without the need for surgical intervention because of the mild severity of the problems. The dysphagia improved at 6 months of age, and the stridor improved at 8 months of age. Her condition is stable at present at 9 months of age.

Discussion

The embryological origin of the various different anomalies of the aortic arch is based on the theory of the hypothetical double aortic arch, which was described by Edwards et al. [10]. In the hypothetical double aortic arch theory, the ascending aorta is divided into two RAAs and two left aortic arches, which surround each side of the trachea and esophagus and connect together to form descending aorta. A common carotid artery and subclavian artery are derived from each aortic arch. Each side has a ductus connected to the aortic arch and pulmonary artery (Fig. 7). Normally, the regression occurs between the origin of right subclavian artery and descending aorta in the RAA. The regression also includes the right ductus. Thus, the normal aortic arch is formed. If the regression occurs between the origin of LSCA and the descending aorta in the left aortic arch, it then becomes an RAA with mirror-image branching. If the interruption presents between the left carotid artery and the LSCA in the left aortic arch, it will form an RAA with aberrant LSCA [11]. There can be a dilated segment at the proximal part of the aberrant LSCA. It is known as the diverticulum of Kommerell. The proximal part of the aberrant artery carries the blood flow from the ductus into the descending aorta; and, therefore, the proximal part of the aberrant artery is as wide as the ductus and descending aorta. Depending on the presence of inappropriate persistence or the different regression segments, there are other types of aortic arch anomalies, including double aortic arch, left aortic arch with aberrant right subclavian artery, and circumflex retroesophageal aortic arch [11].

Table 1
Characteristics and postnatal outcome of the fetal RAA with aberrant LSCA in our study

Characteristics	Case 1	Case 2	Case 3
GA at diagnosis (wk)	20	22	21
Karyotype of fetus	46,XY	46,XY	46,XX
Associated with other congenital cardiac anomaly	No	No	No
Delivery age (wk)	38	39	38
Type of delivery	Vaginal delivery	Vaginal delivery	Vaginal delivery
Postnatal outcome	Well and alive at three and half years of age	Well and alive at one and half years of age	Stridor and dysphagia, which had improved at 8 months of age
Maternal age (yr)	32	29	33

GA = gestational age; LSCA = left subclavian artery; RAA = right aortic arch.

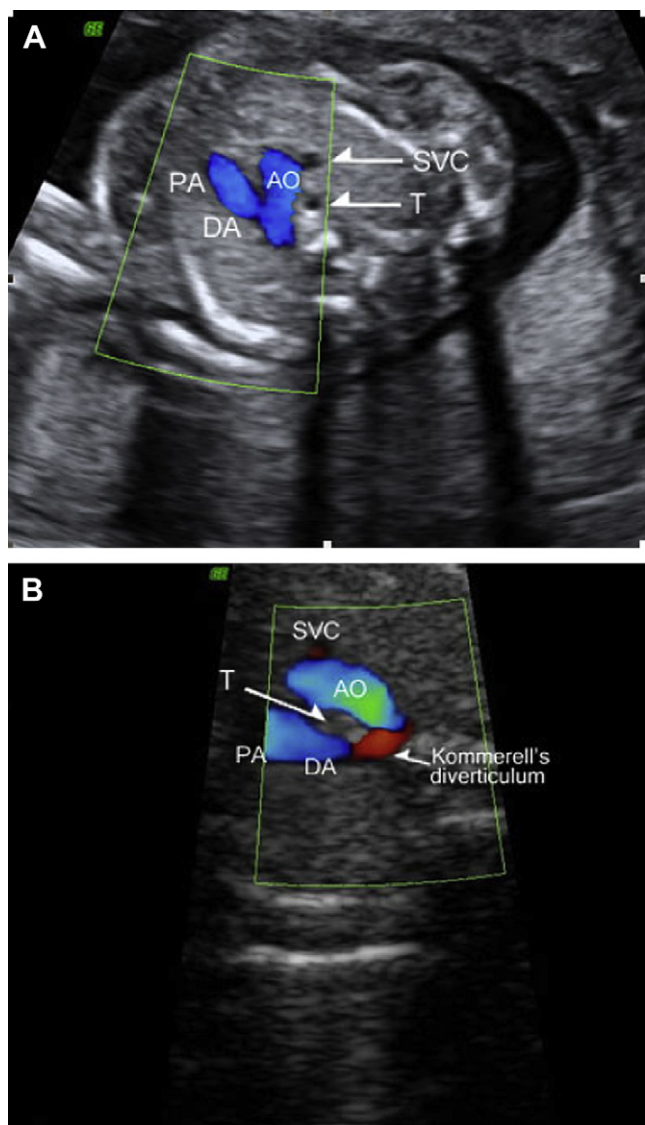


Fig. 4. (A) The normal V-shaped confluence by 3VT view. Notice the trachea is located right laterally and posterior to the vessel junction. (B) The abnormal U-shaped configuration by 3VT view. The trachea is entrapped between aortic arch and pulmonary artery. 3VT=three vessels and trachea; AO=aorta; DA=ductus arteriosus; PA=pulmonary artery; SVC=superior vena cava; T=trachea.

The incidence of the RAA is 0.1%; this has been reported using prenatal diagnosis and for the adult population [2,3]. RAA with aberrant LSCA is rarely concomitant with any other congenital heart disease, and this accounts for just about 10%. On the other hand, the risk of association with other congenital heart disease in the mirror-image branching type is more than 90% [3]. The most common association is Tetralogy of Fallot. Hastreiter et al. [3] described that the RAA with mirror-image branching was 13%–34% associated with Tetralogy of Fallot, 20% associated with double-outlet right ventricle, and 13%–35% associated with truncus arteriosus. In our cases, all the fetuses having RAA with aberrant LSCA were not combined by other congenital heart anomalies.

RAA may be combined with chromosomal anomalies, such as a microdeletion of chromosome 22q11.2. In our cases, all

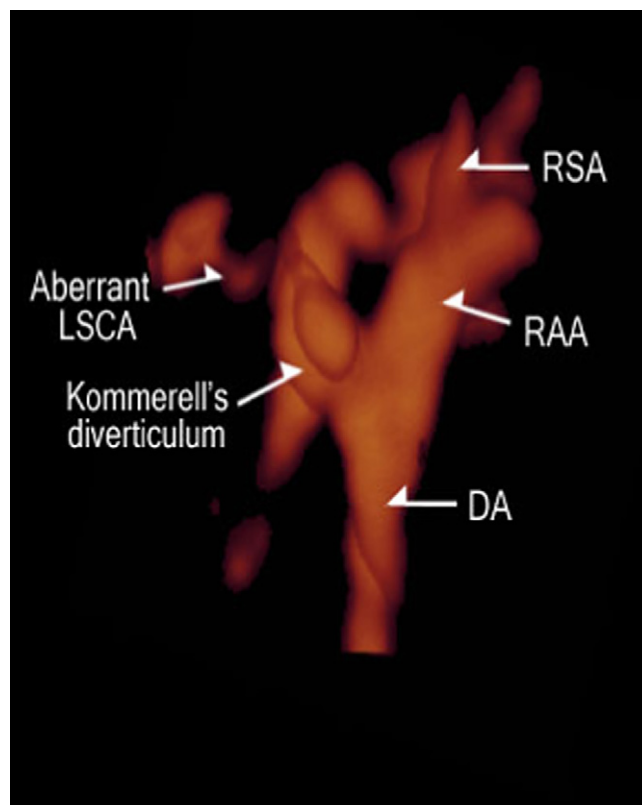


Fig. 5. 3D-power Doppler of the right aortic arch with aberrant left subclavian artery in Case 1. Notice the vascular ring formed by right aortic arch and aberrant left subclavian artery. Kommerell's diverticulum and right subclavian artery can also be visualized. 3D=three-dimensional; DA=descending aorta; LSCA=left subclavian artery; RAA=right aortic arch; RSA=right subclavian artery.

the three cases of RAA with aberrant LSCA had a normal karyotype. Deletions within chromosome 22 are thought to impair migration and/or the development of cells from the cardiac neural crest and thus this interferes with the formation of the central cardiovascular system. McElhinney et al. [4] published that six of eleven patients with RAA with mirror-image branching but without other cardiac anomalies had DiGeorge syndrome. He mentioned that RAA is frequently associated with microdeletion of chromosome 22q11.2, regardless of the presence of associated cardiac anomalies.

The prenatal diagnosis of RAA was based on the 3VT view. From the four-chamber view plane, the transducer is moved upward along the long axis of the fetal body to the upper mediastinum. Then the 3VT view is obtained. In this plane, it is possible to see the superior vena cava, main pulmonary artery, and ascending aorta. These three vessels are aligned in a straight line from the right posterior aspect to the left anterior aspect of the thorax. Furthermore, the trachea is filled with fluid when the child is a fetus and therefore the trachea is visible and dorsal to the aorta. Therefore, the term 3VT view was established [5]. Tracing to the aortic arch and ductus arteriosus at the descending aorta, it will form a V-shaped confluence. The right arm of the “V” represents the aortic arch continuous with the descending aorta, and the left arm

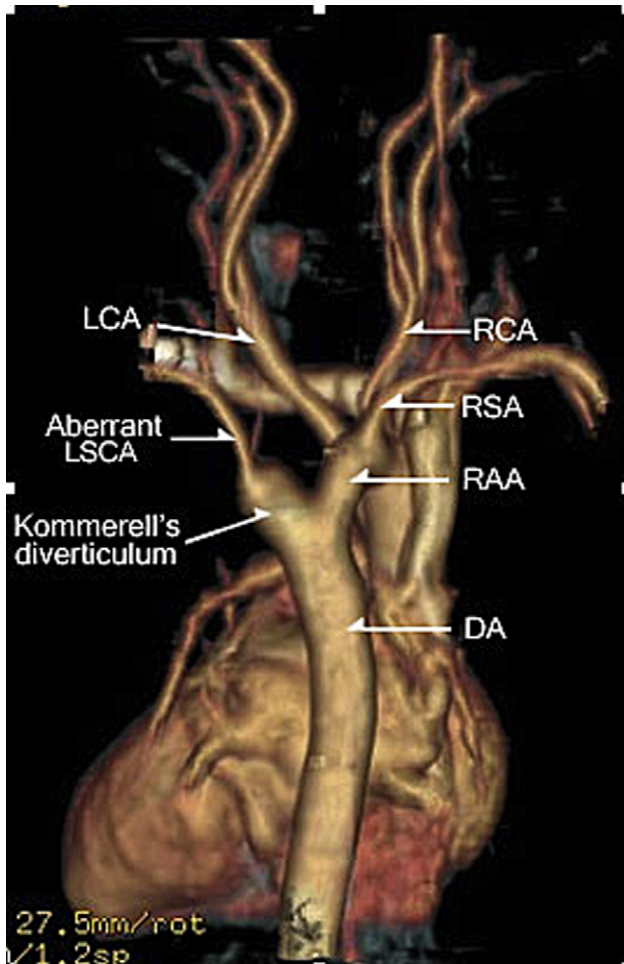


Fig. 6. The 3D reconstruction of 64-slices CT angiography of the right aortic arch with aberrant left subclavian artery postnatally. 3D = three-dimensional; CT = computed tomography; DA = descending aorta; LCA = left carotid artery; LSCA = left subclavian artery; RAA = right aortic arch; RCA = right carotid artery; RSA = right subclavian artery.

represents the pulmonary artery connected with the ductus arteriosus. In this view, the trachea is located laterally and posterior to the vessel junction. This is situated on the right side of aorta. In RAA, the aortic arch and pulmonary artery are connected with ductus arteriosus to form a U-shaped configuration. Both are connected by Kommerell's diverticulum. The trachea is trapped between the aortic arch, and the pulmonary artery is connected with the ductus arteriosus and is ventral to the Kommerell's diverticulum. This is located on the left side of aortic arch [2].

3D-PDU is another method for the prenatal diagnosis of RAA. The 3D-PDU is an ultrasound technology that reconstructs the 3D structure of the vessels after they are visualized using power Doppler ultrasound. The anatomy of great vessels can be visualized well enough to detect any abnormal vascular structures [7,12]. In our case, the two-dimensional power Doppler was first set to obtain the longitudinal ventrodorsal coronal view with visualization of the descending aorta and trachea. Secondly, the color box size was selected for the needed image information. Afterward, a mechanic sweep was

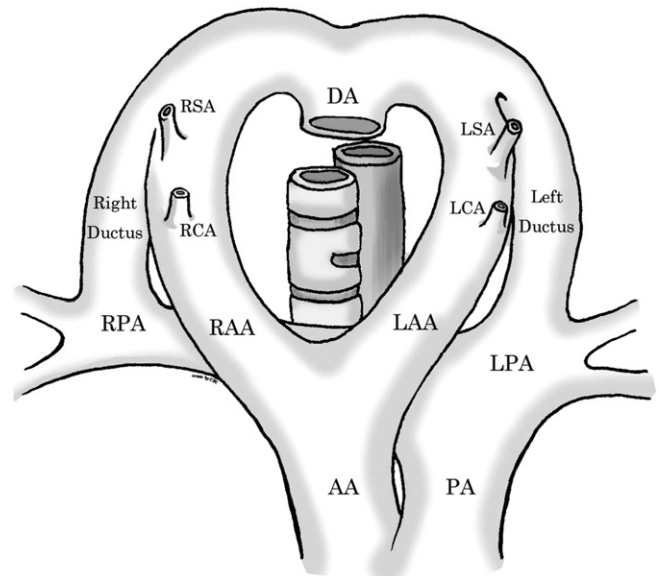


Fig. 7. The embryological origin of the hypothetical double aortic arch. AA = ascending aorta; DA = descending aorta; LAA = left ascending aorta; LCA = left carotid artery; LPA = left pulmonary artery; LSA = left subclavian artery; RAA = right ascending aorta; RCA = right carotid artery; RPA = right pulmonary artery; RSA = right subclavian artery. Illustration by Pei-Ching Chen.

made and followed by reconstruction to give a 3D structure for the vessels. The RAA, aberrant LSCA, Kommerell's diverticulum, and descending aorta were visualized. Although the trachea and esophagus cannot be seen by this method, the vascular ring is clearly visible and how it goes around the trachea/esophagus is easily imagined.

In our cases, a confirmed diagnosis of RAA with aberrant LSCA after delivery was mainly by 64-slice 3D helical CT angiography. A 3D reconstruction using the multi-slice helical CT scan images is a newer imaging technique. This minimally invasive procedure can clearly visualize the detailed anatomy of complicated vascular structures as well as the interrelationship with other systems [13]. Therefore, the 3D multislice helical CT not only provides a precise diagnosis but also aids the cardiac surgeon when planning a surgical strategy if surgical intervention is indicated.

In RAA with aberrant LSCA, the trachea and esophagus are enclosed by a vascular ring formed by aortic arch, aberrant LSCA, ductus arteriosus, and pulmonary arterial trunk. The vascular ring, especially the aberrant LSCA with Kommerell's diverticulum, may compress the trachea and/or esophagus and cause symptoms, such as stridor and dysphagia. Donnelly et al. [14] described 9 of 12 cases of RAA with aberrant LSCA where there was airway compression at the level of the arch and aberrant subclavian artery, which was contributed to by the Kommerell's diverticulum. The severity of symptoms at presentation depends on the degree of vascular compression. Fortunately, the vascular ring is usually loose, so the clinical symptoms seldom appear. Achiron et al. [2] reported that only 1 of 18 children with RAA was symptomatic at the age of 6 months and all other cases showed a normal course on follow-up to 60 months. In our cases, one of three babies

showed symptoms of tracheoesophageal compression. However, our case number is too small to reflect the incidence of symptomatic cases. In the mirror-image branching type, it does not form a vascular ring or sling. Therefore, there will be no tracheoesophageal compression in theory. However, when a left ductus arteriosus connects the left pulmonary artery to the upper descending aorta (the left ductus connects the left pulmonary artery to the LSCA mostly), this will form a vascular ring and is often symptomatic if the descending aorta passes to the left side of the vertebral column [15].

For those symptomatic patients with compression of the vascular ring, especially where there is severe respiratory distress with stridor, recurrent respiratory tract infections and/or severe dysphagia with a failure to thrive, surgical intervention is indicated. The surgical management mainly involves release of the compression of vascular ring and includes division of the left ductus arteriosus and/or division of the origin of aberrant LSCA followed by anastomosis to the aorta or to the left common carotid when there is a potential risk of rupture of the Kommerell's diverticulum [3,16,17]. For mildly symptomatic patients, conservative management, such as drainage of bronchial secretions, a humidified air supply, antibiotics for suspected upper respiratory tract infections, and a soft diet are suggested. For asymptomatic patients, long-term follow-up is suggested.

Prenatal diagnosis of RAA with aberrant LSCA is important, especially if detection of associated congenital cardiac anomalies is carried out. For those cases that are not combined with another anomaly, most are asymptomatic and have a satisfactory outcome. Nevertheless, precise prenatal diagnosis is able to alert clinicians so that they can intensively manage the infants, which avoids misdiagnosis or delay in treatment. In conclusion, we emphasize the importance of the 3VT view in routine prenatal ultrasound examinations. In addition, the use of 3D-PDU can help with the visualization of any malformations of the fetal vascular system and provides a more precise prenatal diagnosis for these congenital cardiovascular abnormalities.

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References

- [1] Weinberg PM. Aortic arch anomalies. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, editors. *Moss and Adams' heart disease in infants, children, and adolescents including the fetus and young adult*. 7th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008. p. 730–60.
- [2] Achiron R, Rotstein Z, Heggesh J, Bronshtein M, Zimand S, Lipitz S, et al. Anomalies of the fetal aortic arch: a novel sonographic approach to in-utero diagnosis. *Ultrasound Obstet Gynecol* 2002;20:553–7.
- [3] Valletta EA, Pregarz M, Bergamo-Andreis IA, Boner AL. Tracheoesophageal compression due to congenital vascular anomalies (vascular rings). *Pediatr Pulmonol* 1997;24:93–105.
- [4] McElhinney DB, Hoydu AK, Gaynor JW, Spray TL, Goldmuntz E, Weinberg PM. Patterns of right aortic arch and mirror-image branching of the brachiocephalic vessels without associated anomalies. *Pediatr Cardiol* 2001;22:285–91.
- [5] Yoo SJ, Lee YH, Kim ES, Ryu HM, Kim MY, Choi HK, et al. Three-vessel view of the fetal upper mediastinum: an easy means of detecting abnormalities of the ventricular outflow tracts and great arteries during obstetric screening. *Ultrasound Obstet Gynecol* 1997;9:173–82.
- [6] Chaoui R, McEwing R. Three cross-sectional planes for fetal color Doppler echocardiography. *Ultrasound Obstet Gynecol* 2003;21:81–93.
- [7] Chaoui R, Kalache KD, Hartung J. Application of three-dimensional power Doppler ultrasound in prenatal diagnosis. *Ultrasound Obstet Gynecol* 2001;17:22–9.
- [8] Chaoui R, Schneider MBE, Kalache KD. Right aortic arch with vascular ring and aberrant left subclavian artery: prenatal diagnosis assisted by three-dimensional power Doppler ultrasound. *Ultrasound Obstet Gynecol* 2003;22:661–3.
- [9] Yoo SJ, Lee YH, Cho KS, Kim DY. Sequential segmental approach to fetal congenital heart disease. *Cardiol Young* 1999;9:430–44.
- [10] Edwards JE. Vascular rings and slings. In: Moller JH, Neal WA, editors. *Fetal, neonatal, and infant cardiac disease*. Norwalk, CT: Appleton & Lange; 1990. p. 745–54.
- [11] Yoo SJ, Min JY, Lee YH, Roman K, Jaeggi E, Smallhorn J. Fetal sonographic diagnosis of aortic arch anomalies. *Ultrasound Obstet Gynecol* 2003;22:535–46.
- [12] Chaoui R, Kalache KD. Three-dimensional power Doppler ultrasound of the fetal great vessels. *Ultrasound Obstet Gynecol* 2001;17:455–6.
- [13] Katz M, Konen E, Rozenman J, Szeinberg A, Itzhak Y. Spiral CT and 3D image reconstruction of vascular rings and associated tracheobronchial anomalies. *J Comput Assist Tomogr* 1995;19:564–8.
- [14] Donnelly LF, Fleck RJ, Pacharn P, Ziegler MA, Fricke BL, Cotton RT. Aberrant subclavian arteries: cross-sectional imaging findings in infants and children referred for evaluation of extrinsic airway compression. *Am J Roentgenol* 2002;178:1269–74.
- [15] Schlesinger AE, Mendeloff E, Sharkey AM, Spray TL. MR of right aortic arch with mirror-image branching and a left ligamentum arteriosum: an unusual cause of a vascular ring. *Pediatr Radiol* 1995;25:455–7.
- [16] Lunde R, Sanders E, Hoskam JAM. Right aortic arch symptomatic in adulthood. *Neth J Med* 2002;60:212–5.
- [17] Babu R, Pierro A, Spitz L, Drake DP, Kiely EM. The management of esophageal atresia in neonates with right-sided aortic arch. *J Pediatr Surg* 2000;35:56–8.