

Case Report

Prenatal diagnosis of fetal double aortic arch: report of a case

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Abstract: Double aortic arch (DAA) is the most common congenital anomaly of the aortic arch system, in which the trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches, often resulting in variable airway compression. We present a case of fetal DAA prenatally diagnosed by fetal echocardiography and clearly confirmed at autopsy. The autopsy visualization allowed for better understanding of this rare cardiac anomaly and facilitated improving the prenatal diagnostic rate.

Keywords: Fetal echocardiography, double aortic arch, autopsy

Introduction

Double aortic arch (DAA) is the most common congenital anomaly of the aortic arch system, in which the trachea and esophagus are completely encircled by connected segments of the aortic arch and its branches, often resulting in invariable airway compression [1]. This is a case report of the prenatal diagnosis of fetal DAA identified by fetal echocardiography with confirmation at autopsy.

Case report

A 26-year-old woman was referred for detailed fetal cardiovascular evaluation at 24 weeks gestation because of fetal aortic arch system abnormalities suspected at another hospital. Routine ultrasound scanning of the fetus demonstrated normal growth and no extra-cardiac congenital structural anomalies, and sequential segmental analysis suggested that situs solitus, concordant atrioventricular and ventriculoarterial connections with balanced chambers. However, moving the transducer to the view of the transverse aspect of the upper mediastinum, a complete vascular ring encircling the fluid-filled trachea and four separate brachiocephalic vessels grouped around the trachea (instead of the normal three vessels) were found by both two-dimensional and color Doppler imaging (**Figure 1**) suggesting an abnormality of the aortic arch, and the longitudi-

nal view of the trachea showed that there was no airway compression at the level of the vascular ring.

The fetus died of severe unexplained intrauterine infection at 27 weeks gestation. Autopsy was performed and confirmed the diagnosis of DAA (**Figures 2-4**), in which the ascending aorta bifurcated directly at the level of the trachea with arches to the right and to the left in a letter "Y" configuration, with both arches passing over the respective bronchi and subsequently joining posterior to the esophagus and fusing into the descending thoracic aorta which had a left-sided course anterior to the spine, an anatomically complete vascular ring formed and each aortic arch gave off the subclavian artery and the common carotid independently. And in this case the dominant arch was the right one and the junction between the distal segment of the left arch was narrow. The autopsy visualization allowed for better understanding of this rare cardiac anomaly and facilitated improving the prenatal diagnostic rate.

Discussion

The embryonic development of DAA is due to persistence of the fourth arches and dorsal vessels which leads to a complete vascular ring [2, 3]. Each arch is usually patent, although the right arch is invariably larger and higher in 75% of cases and there might be narrowing or even

Fetal double aortic arch

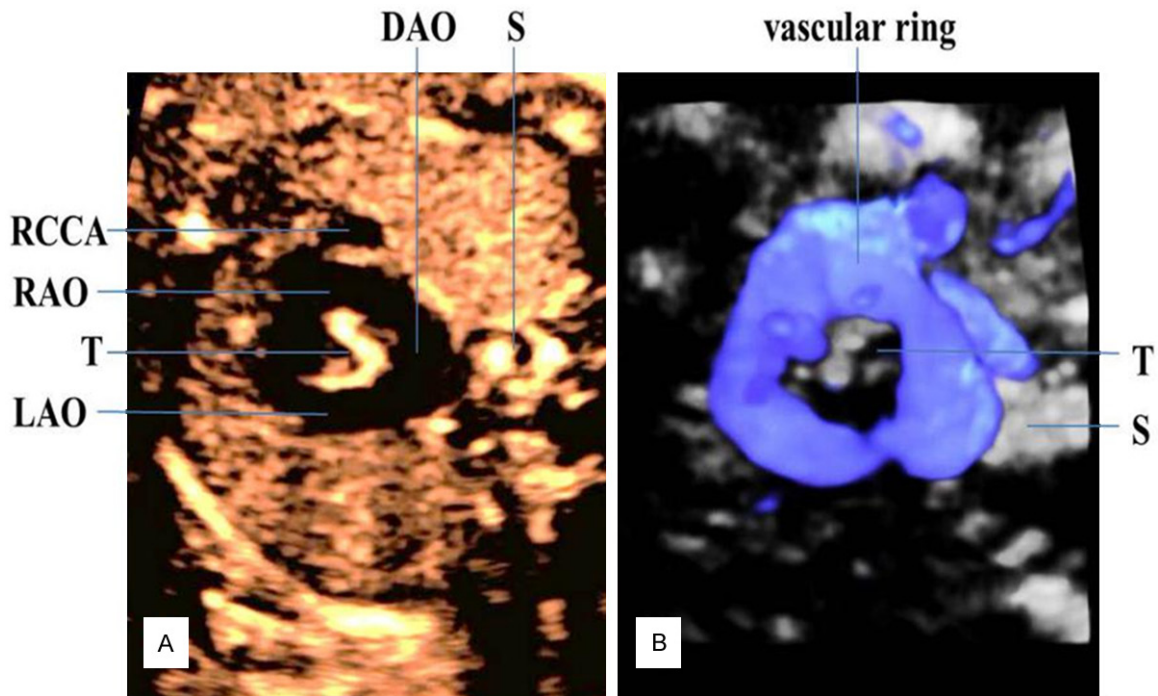


Figure 1. Three vessels and tracheal view showed that both arches form a complete vascular ring which surrounds the trachea and esophagus tightly, and behind esophagus both arches fuse into the descending aorta which is left and anterior to the spine. LAO indicates ascending left aortic arch; RAO, right aortic arch; T, trachea; and S, spine.

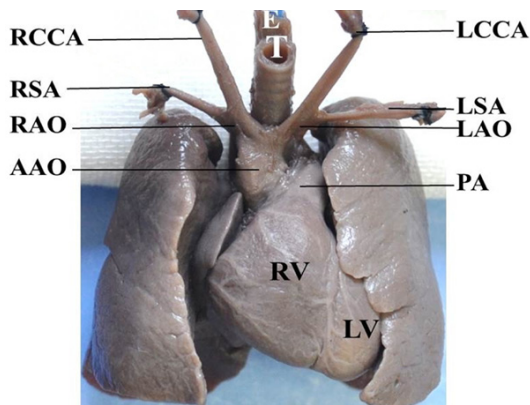


Figure 2. (Anterior view) Autopsy of the fetal heart showed that the heart position is normal with the cardiac apex on the left side. The ascending aorta bifurcates directly at the level of the trachea to have one right arch and one left arch, and each arch gives off one common carotid and subclavian arteries respectively. AAO indicates ascending aorta; PA, pulmonary artery; LAO, left aortic arch; RAO, right aortic arch; LCCA, left common carotid; LSA, left subclavian artery; RCCA, right common carotid artery; RSA, right subclavian artery; LV, left ventricle; RV, right ventricle; T, trachea; and E, esophagus.

atresia of a segment of the left arch. Approximately 20% of cases are associated with other cardiovascular anomalies including tetral-

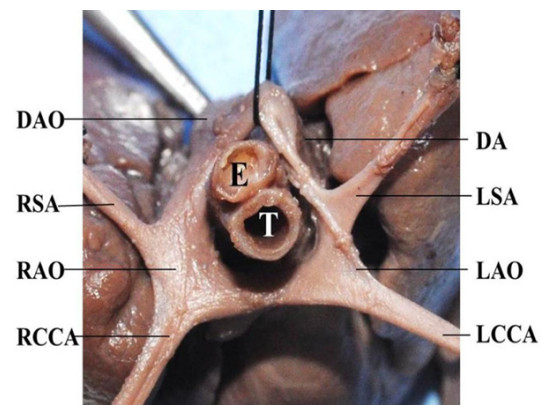


Figure 3. (Superior view) Autopsy of the fetal heart showed that the two aortic arches form a complete vascular ring and embraces the trachea and esophagus, and the left arch is a little narrower than the right one. LAO indicates left aortic arch; RAO, right aortic arch; LCCA, left common carotid; RCCA, right common carotid artery; LSA, left subclavian artery; RSA, right subclavian artery; DAO, descending aorta; T, trachea; and E, esophagus.

ogy of Fallot, transposition of the great arteries, coarctation of the aorta, ventricular septal defect and patent ductus arteriosus. Also, esophageal atresia can be found in some cases [4, 5].

Fetal double aortic arch

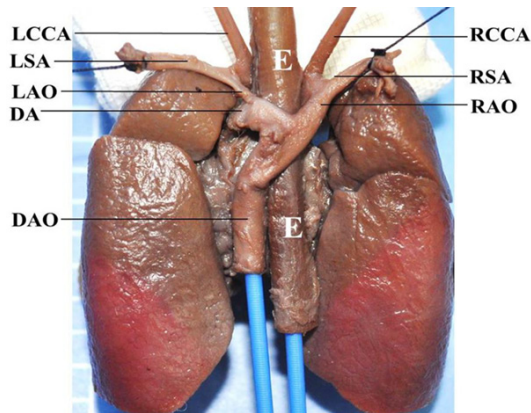


Figure 4. (Posterior view) Autopsy of the fetal heart showed that both arches fuse into the descending aorta behind esophagus. LAO indicates left aortic arch; RAO, right aortic arch; LCCA, left common carotid; LSA, left subclavian artery; RCCA, right common carotid artery; RSA, right subclavian artery; DAO, descending aorta; and E, esophagus.

DAA usually causes respiratory symptoms in 91% and gastrointestinal symptoms in 40% of cases. The respiratory symptoms include stridor, choking, episodes, recurrent respiratory infections, and life-threatening apneic spells [6]. Related to esophageal compression or even atresia, gastrointestinal symptoms manifest as vomiting and feeding intolerance in infants and dysphagia in older children and adults.

Early surgically dividing the arches and freeing the trachea and esophagus from surrounding tissues is imperative to prevent the long-term sequelae of tracheobronchial compression of patients with DAA [7]. Surgical treatment successfully eradicates symptoms in over 70% cases. However, due to residual tracheal stenosis, airflow limitation may persist after operation, patients with that may benefit from resection of a severely malacictracheal segment and tracheoplasty. In general, surgery affords excellent long-term resolution of symptoms, although symptoms may not be relieved immediately, necessitating close long-term follow-up [8, 9].

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Disclosure of conflict of interest

None.

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References

- [1] Lima JA, Rosenblum BN, Reilly JS, Pennington DG, Nouri-Moghaddam S. Airway obstruction in aortic arch anomalies. *Otolaryngol Head Neck Surg* 1983; 91: 605-609.
- [2] Alsenaidi K, Gurofsky R, Karamlou T, Williams WG, McCrindle BW. Management and Outcomes of Double Aortic Arch in 81 Patients. *Pediatrics* 2006; 118: e1336.
- [3] Edwards JE. Anomalies of derivatives of aortic arch system. *Med Clin North Am* 1948; 32: 925-49.
- [4] van Son JA, Julsrud PR, Hagler DJ, Sim EK, Pailorero PC, Puga FJ, Schaff HV, Danielson GK. Surgical treatment of vascular rings: the Mayo Clinic experience. *Mayo Clin Proc* 1993; 68: 1056-1063.
- [5] Bronshtein M, Lorber A, Berant M. Sonographic diagnosis of fetal vascular rings in early pregnancy. *Am J Cardiol* 1998; 81: 101-3.
- [6] Zoumalan R, Maddalozzo J, Holinger L. Etiology of stridor in infants. *Ann Otol Rhinol Laryngol* 2007; 116: 329-334.
- [7] Fleck RJ, Pacharn P, Fricke BL, Ziegler MA, Cotton RT, Donnelly LF. Imaging findings in pediatric patients with persistent airway symptoms after surgery for double aortic arch. *AJR Am J Roentgenol* 2002; 178: 1275-9.
- [8] Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. *J Thorac Cardiovasc Surg* 2005; 129: 1339-47.
- [9] Ullmann N, Menchini L, Salerno T, Tomà P, Cutrera R. Late Diagnosis of Double Aortic Arch: Consequences on Long-Term Follow-Up. *Pediatr Pulmonol* 2013; 9999: 1-3.