

Fetal Vascular Rings: Beyond The Anomalies of The Aortic Arch

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2. Key words

Ductus Arteriosus (DA), Right Ductus Arteriosus (RDA), Aberrant Right Subclavian Artery (ARSA), Left Aortic Arch (LAA), Sd.Di George.

1. Abstract

Different anomalies related to the inappropriate development of the ductus arteriosus or the aortic arch have been described, in some cases accompanied with chromosomal or morphological anomalies, and also being able to form a vascular ring that can compromise the **post-natal** life. For this reason, a complete fetal anatomy examination and cardiovascular study is needed to discard possible other malformations or ultrasound markers for fetal syndromes. A detailed prenatal diagnosis of the type of ductal arch anomaly and possible vascular ring can give us a postnatal prognosis and help pediatricians with the management of symptomatic neonates.

The abnormalities related to the position or branching of the ductal arch (ductus arteriosus) are very uncommon. We describe the first case reported of prenatal diagnosis of right ductus arteriosus, left aortic arch and aberrant right subclavian artery associated with 22q11 deletion.

3. Introduction

The fetal ductus arteriosus (DA) is a vascular structure with functional importance in the fetal circulation. It connects the Main Pulmonary Artery (MPA) or its left branch to the aorta, diverting blood flow apart from the pulmonary vascular bed to the systemic circulation.

In the model of embryonic vascular development, six pairs of aortic arches communicate the ventral and dorsal aortas. The ductus arteriosus arises from the sixth aortic arch. In a normally developed heart, the normal branching of the left aortic arch is, from right to left: brachiocephalic trunk (rising right common carotid and right subclavian), left common carotid and left subclavian artery [1]. In normal cardiac development, both right aortic and ductal arches regress at 4-7 gestational weeks [2] (Figure 1).

Different anomalies related to the inappropriate development of the aortic and ductal arches have been described, with a prevalence of 1-2% [3,4]. Arch anomalies refer to a variety of congenital abnormalities that are related to the position or branching of the aortic and/or ductal arches. Aortic arch anomalies are most common. However, other uncommon abnormalities of position/branching of the ductal arch may occur (right or double DA, unusual, tortu-

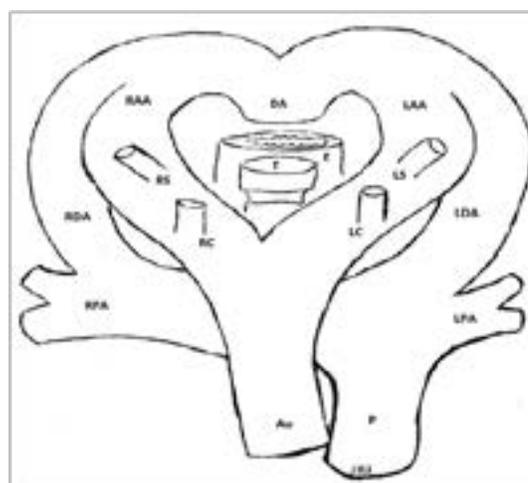


Figure 1: Hypothetical double aortic arch model of Jesse E. Edwards. Aa, ascending (ventral) aorta; DA, descending (dorsal) aorta; T, trachea; E, esophagus; LAA, left aortic arch; LC, left common carotid artery; LPA, left pulmonary artery; LS, left subclavian artery; P, main pulmonary artery; RAA, right aortic arch; RC, right common carotid artery; RPA, right pulmonary artery; RS, right subclavian artery.

ous or S-shaped ductus). Scarce data is published in the literature about right ductus arteriosus (RDA) [2-15]. Most of the papers describe RDA associated to Right Aortic Arch (RAA) or in the context of Congenital Heart Disease (CHD) [2]. The aim of this study is to report a new case of an uncommon type of RDA with Left Aortic Arch (LAA) without a major CHD and to review the literature.

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4. Case Report

Patient of 35 years old, nulliparous, attended our hospital to pregnancy follow-up. First trimester scan was normal and combined test was at low risk of aneuploidy.

At the second trimester scanning (21+2w), an Aberrant Right Subclavian Artery (ARSA) was suspected. In the three-vessel and tracheaview (3VT) a complete vascular ring around the trachea and thymus hypoplasia were detected. Due to the suspicion of congenital heart disease, a detailed echocardiography was performed. The results were as follows: situs solitus, atrioventricular-ventriculoarterial connections and cardiac axis and size were normal. At 3VT view, right-sided ductus arteriosus was detected arising from the right pulmonary artery to the aortic isthmus of the ascending aorta located at the right side of the trachea, with a left-sided aortic arch and trachea in-between, having a U-shaped or ribbon-shaped confluence instead of V-shaped and the normal left aortic arch at the left of the trachea (Figures 2, Figures 3). The thymus was hypoplastic at 3VT view. ARSA was confirmed and RDA formed a complete vascular ring, described by some authors as the cross ribbon sign [12]. The rest of cardiac anatomy was normal.

Due to the diagnosis of ARSA, RDA and thymic hypoplasia, and invasive test was offered. An amniocentesis was performed at 27 weeks. Results revealed a normal karyotype with a 22q11 microdeletion at the microarray (Sd. DiGeorge). A 2940g fetus was delivered vaginally at term. The newborn presented an episode of neonatal hypocalcaemia and mild dysphagia that solved correctly. A postnatal echocardiography was performed confirming the LAA

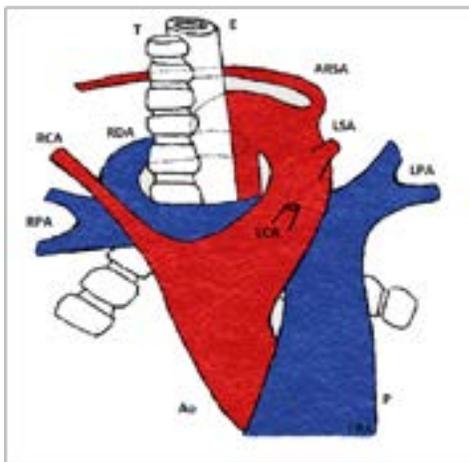


Figure 2: Developmental model of left aortic arch with right ductus arteriosus (RDA) and aberrant right subclavian artery (ARSA). Ao, ascending (ventral) aorta; DA, descending (dorsal) aorta; T, trachea; E, esophagus; LAA, left aortic arch; LC, left common carotid artery; LPA, left pulmonary artery; LS, left subclavian artery; P, main pulmonary artery; RAA, right aortic arch; RC, right common carotid artery; RPA, right pulmonary artery; RS, right subclavian artery; ARSA, Aberrant right subclavian artery.

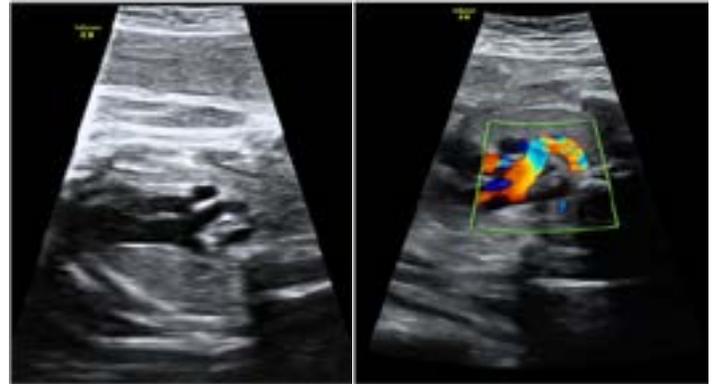


Figure 3: (A) Transversal thoracic 3VT view: Bifurcation of pulmonary artery with the origin of a complete vascular ring formed by the right DA. (B) Same view using colour doppler. R, right; l, left.

and a correct closure of the RDA. The 18-month old child is completely normal with no cardiovascular or respiratory symptoms.

5. Discussion

Anomalies of the DA can be diagnosed prenatally (DA aneurism, absence DA), including abnormalities in DA branching [16,17].

RDA with a LAA could be suspected at the 3VT view, by a complete vascular ring described by some authors [12] as the cross ribbon sign. This vessel is a vascular structure that originates in the bifurcation of the MPA or in the origin of the right pulmonary artery. RDA crosses right to and behind the trachea and joins the descending left aorta. The persistence of RDA with LAA forming this vascular ring is very uncommon and rarely reported. Bronshtein et al. [2] reported a prevalence of 0.035%. The very low detection rate of this anomaly might be due to: (1) unawareness of the existence of this anomaly (2) difficulty of its diagnosis in the second trimester of pregnancy [2].

When a RDA is suspected, we should perform an echocardiography [13, 14] to exclude other congenital heart diseases and vascular rings. The examination must include the 5 transverse planes described by Yagelet al. [18]. Most recently a new sonographic view is suggested to the fetal heart exploration, particularly when a congenital heart disease or arches anomalies are suspected [4]: the subclavian artery view. In case of ductal/aortic arch anomalies, in the 3VT color image, Doppler demonstration of the usual blue or red 'V-sign' is replaced by a variety of patterns, depending on the etiology of the anomaly [19].

Differential diagnosis is important because arches anomalies can present similar patterns in the 3VT image. We should consider the relationship between the aortic and ductal arches with the trachea and the possibility of a vascular ring (complete or incomplete). Dif-

ferent combinations might occur: RAA with left ductus arteriosus (LDA), RAA with RDA, double aortic arch or LAA with RDA, as in our case. In the cases of RDA with LAA, 3VT view is completely abnormal. RDA with RAA, however, needs more attention and it could be easily overlooked: RAA with RDA form a V-shaped configuration like the normal fetal anatomy, but on the right side of the trachea [13].

3D/4D ultrasound can be very useful for a more precise diagnosis in case of vascular rings. In this case, 4D clarified the relationship between the vascular structures (Figures 4, Figures 5).

In addition, a complete fetal anatomy examination is needed to discard other malformations or ultrasound markers for fetal syndromes.

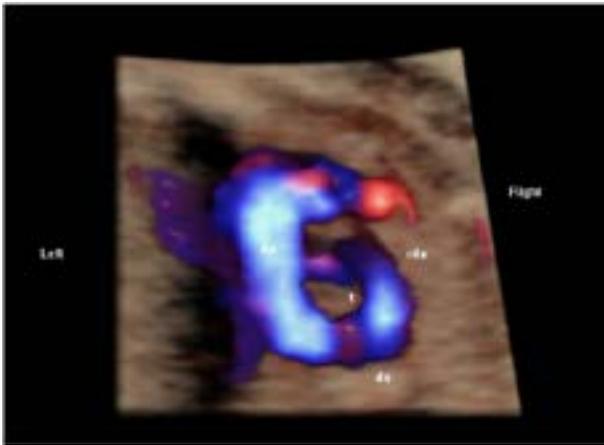


Figure 3: Transversal thoracic view, upper mediastinum using STIC technique: left aortic arch forming a complete vascular ring with the right ductal arch (*cross ribbon sign*). Ao, aorta; t, trachea; da, descending aorta; rda, right ductus arteriosus.

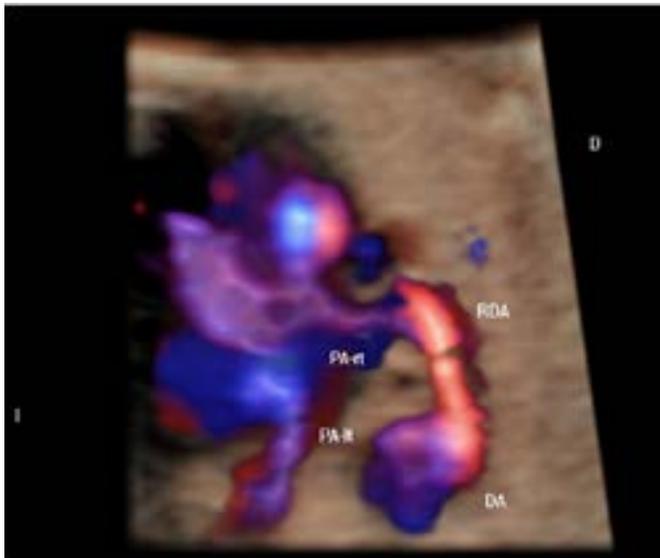


Figure 5: Transversal thoracic view, upper mediastinum: Bifurcation of the pulmonary artery, with the formation of the right DA originating in the right pulmonary artery and ending in the aortic isthmus. DA, descending aorta; RDA, right ductus arteriosus; PA-rt, right pulmonary artery; lt-PA, left pulmonary artery; r, right; l, left.

RDA with LAA is a very uncommon anomaly. We could find no reference to it in the main prenatal diagnosis textbooks and we found only 3 reports about RDA and LAA in the on line literature (Table 1). The lack of literature might be due to the rarity of this anomaly and also to the difficulty of its diagnose. A few more reports describe the association of RDA with RAA [2,13] diagnosed prenatally. All cases of RDA in pediatric literature are associated to other cardiac malformations [5, 8]. Despite of the limited literature, we would suspect that most cases of RDA are isolated, and remain undetected. We believe this because RDA is diagnosed after delivery only when it remains patent, and this occurs mainly in association with CHD or other pathologic states.

To our knowledge this is the first case of prenatal diagnosis of RDA, LAA and ARSA associated with 22q11 deletion. DiGeorge Syndrome is associated with anomalies of the cardiac outflow tracts. In this case, DA was displaced to the right. Classically the anomalies of the aortic arch are associated with congenital heart defects, chromosomal anomalies, especially trisomy 21 or deletion 22q11 [20,21], but DA abnormalities are seldom mentioned.

In our case, the child had mild symptoms of compression after delivery but surgery was not needed. There is no consensus between pediatricians and obstetricians regarding esophagus or trachea compression symptoms. According to prenatal series most of the vascular rings are asymptomatic after delivery, with the exception of DAA and some cases of RAA with ARSA [22,23]. However, pediatricians report that between 30 and 60% of cases of anomalies in aortic arch and ARSA can cause obstructive symptoms due to an extrinsic compression of the esophagus and trachea [24,25]. It is described significant tracheal compression in asymptomatic infants with a RAA and left DA, considering an early airway investigation even in this type of patients [26]. Another sign recently described that should be taken into account is the high incidence of progressive stenosis in aberrant left subclavian artery (ALSA) with RAA in asymptomatic newborns [27]. In our case, the RDA was associated to an ARSA that might worsen the prognosis.

6. Conclusion

In conclusion, ductal abnormalities are uncommon in prenatal life. Prenatal diagnosis of RDA is feasible and its diagnosis provides several benefits. Making a detailed prenatal diagnosis of the type of ductal arch anomaly and possible vascular ring can give us a postnatal prognosis and help pediatricians with the management of symptomatic neonates. Arch anomalies, aortic or ductal, can be either isolated or associated to other anomalies, commonly congenital heart diseases, and chromosomal abnormalities, including

22q11 deletion, as in our case. Based on this association invasive testing should be highly considered prenatally.

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References

- Galindo A., Alteraciones arco aórtico. In: A. Galindo, E. Gratacós, J. Martínez. *Cardiología Fetal Ecografía Obstétrica*. 1a ed. España: Marban. 2015; p. 393-405
- Moshe Bronshtein, Ethan Z. Zimmer, Shraga Blazer, Z. Blumenfeld. Right ductus arteriosus: facts and theory. *Eur J ObstetGynecolReprod Biol*. 2011; 159(2): 282-8.
- Carlos García Guevara, Carlos García Morejón, Felipe Somoza, Jake-line Arencibia Faife, Pablo Marantz. Prenatal Diagnosis of Vascular Ring with Right Aortic Arch. *Rev Argent Cardiol*. 2012; 80(3): 250-3.
- Coral Bravo, Francisco Gámez, Ricardo Pérez, Teresa Álvarez, Juan De León-Luis. Fetal Aortic Arch Anomalies. *JUM*. 2016; 35(2): 237-51.
- Van Hare GF, Townsend SE, Hardy K, Turley K, Silverman NH. Interrupted aortic arch with a right descending aorta and right ductus arteriosus, causing severe right bronchial compression. *PediatrCardiol*. 1988; 9(3): 171-4.
- Lenox CC, Neches WH, Zuberbuhler JR, Park SC, Mathews RA, Fricker FJ. Bilateral ductus arteriosus in d-transposition of the great arteries with right aortic arch. *Chest*. 1978; 74(1): 94-6.
- Yasuda H, Kado H, Imoto Y, Asou T, Shiokawa YI, Yasui H. Staged repair for bilateral ductus arteriosus with pulmonary atresia and non-confluent pulmonary artery a case report. *Nippon KyobuGeka Gakkai Zasshi*. 1996; 44(10): 1907-11.
- Traxer O, de Lagausie P, Kron C, Belarbi N, Aigrain Y. Left aortic arch-right descending aorta-right ductus arteriosus (encircling aortic arch). A rare malformation of the aortic arches. *Arch Pediatr*. 1998; 5(4): 409-13.
- Cazzaniga M, Rico F, Fernández Pineda L, González C, Quero M, Maître M. Contribution of color Doppler in the diagnosis of bilateral ductus arteriosus. *Rev EspCardiol*. 1998; 51(4): 332-5.
- 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. *PediatrCardiol*. 2001; 22(4): 285-91.
- Kikuchi S, Yokozawa M. Double aortic arch associated with common inlet left ventricle. *PediatrCardiol*. 2005; 26(4): 484-5.
- S BoopathyVijayaraghavan, Sathiya Senthil, K Latha. Prenatal diagnosis of a rare aortic arch anomaly with left aortic arch and right ductus arteriosus: cross ribbon sign. *Indian J Radiol Imaging*. 2017; 27(1): 70-2.
- Ekiz A, Gul A, Uludogan M, Bornau H. Prenatal diagnosis and postnatal outcome of persistent right ductus arteriosus: a report of three cases. *J MedUltrasound*. 2015; 42(4): 571-4.
- Gul A, Gungorduk K, Yildirim G, Gedikbasi A, Borna H, Omeroglu RE. Prenatal sonographic features of persistent right arteriosis with a left aortic arch. *J ObstetGynaecol*. 2013; 33(1): 86-7.
- Iliescu DG, Comanescu AC, Tudorache S, Cernea N. Right aortic arch with patent right ductus arteriosus and normal heart. *Ultrasound Obstet Gynecol*. 2012; 40(1): 115-6.
- Puder KS, Sherer DM, Ross RD, Silva ML, King ME, Treadwell MC, et al. Prenatal ultrasonographic diagnosis of ductus arteriosus aneurysm with spontaneous neonatal closure. *Ultrasound Obstet Gynecol*. 1995; 5(5): 342-5.
- Weichert J, Hartge DR, Axt-Flidner R. The fetal ductus arteriosus and its abnormalities: a review. *Congenit Heart Dis*. 2010; 5(5): 398-408
- Yagel S, Cohen SM, Achiron R. Examination of the fetal heart by five short-axis views: a proposed screening method for comprehensive cardiac evaluation. *Ultrasound Obstet Gynecol*. 2001; 17(5): 367-9.
- Helena Gardiner, RabihChaoui. The fetal three-vessel and tracheal view revisited. *Semin Fetal Neonatal Med*. 2013; 18(5): 261-8.
- Chih-PingChen, Shu-ChinChien. Prenatal Sonographic Features of 22q11.2 Microdeletion Syndrome. *J Med Ultrasound*. 2008; 16(2): 123-9.
- Driscoll DA. Prenatal diagnosis of the 22q11.2 deletion syndrome. *Genet Med*. 2001; 3(1): 14-8.
- Razon Y, Berant M, Fogelman R, Amir G, Birk E. Prenatal diagnosis and outcome of right aortic arch without significant intracardiac anomaly. *J Am Soc Echocardiogr*. 2014; 27(12): 1352-8.
- Li S, Luo G, Norwitz ER, Wang C, Ouyang S, Yao Y, et al. Prenatal diagnosis of congenital vascular rings and slings: sonographic features

- and perinatal outcome in 81 consecutive cases. *Prenat Diagn.* 2011; 31(4): 334-46.
24. Kir M, Saylam GS, Karadas U, Yilmaz N, Çakmakçı H, Uzuner N, et al. Vascular rings: presentation, imaging strategies, treatment, and outcome. *PediatrCardiol.* 2012; 33(4): 607-17.
 25. Suh YJ, Kim GB, Kwon BS, Bae EJ, Noh CI, Lim HG, et al. Clinical course of vascular rings and risk factors associated with mortality. *Korean Circ J.* 2012; 42(4): 252-8.
 26. Vlgneswaran T, Kapravelou E, J.Bell A, et al. Correlation of Symptoms with Bronchoscopic Findings in Children with a Prenatal Diagnosis of a Right Aortic Arch and Left Arterial Duct. *PediatrCardiol.* 2018; 39(4): 665-673.
 27. Muraoka M, Nagata H, Hirata Y, et al. High incidence of progressive stenosis in aberrant left subclavian artery with right aortic arch. *Heart Vessels.* 2018; 33(3): 309-315.