

# A novel symptomatic vascular ring in a neonate

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## ABSTRACT

**Introduction:** Vascular rings are congenital anomalies of the aortic arch system characterized by a vascular anomalies of mediastinal vessels that encircle and compress the esophagus and trachea. Aortic arch anomalies may be associated with other congenital cardiac defects or chromosomal anomalies, such as microdeletion of chromosome 22q11. In neonatal/pediatric life, clinical presentation shows stridor and difficulty in swallowing or dysphagia.

**Case Report:** We describe a particular case diagnosed in fetal life with a symptomatic vascular ring in neonatal life: left circumflex retroesophageal aortic arch, right patent ductus arteriosus “PDA”/ligamentum arteriosum, and aberrant right subclavian artery “ARSA.”

**Conclusion:** To the best of our knowledge, we described a novel vascular ring diagnosed in fetal life and symptomatic in neonatal life. In this extremely rare case, a “true” vascular ring, like in case of double aortic arch, it is due by the left circumflex retroesophageal aortic arch, ARSA, and right patent ductus/ligamentum arteriosum.

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## INTRODUCTION

The acronym “vascular ring” was first introduced by Gross [1], who defined the two classic true anatomic vascular rings: double aortic arch and right aortic arch with a left ligamentum. Later, numerous authors have described vessel anomalies responsible for vascular ring. Vascular ring was introduced in the literature a monograph from the Mayo Clinic [2] that classified vascular rings into seven types; however, this classification is not widely used, and this is why roughly 90% of vascular rings fit into four main categories: double aortic arch, right arch/left ligamentum, innominate artery compression and pulmonary artery sling. The aortic arch normally gives rise to three vessels, the brachiocephalic artery, the left common carotid artery, and the left subclavian artery. The brachiocephalic artery branches give origin to the right subclavian artery and the right common carotid artery. In case of ARSA, the aortic arch has four epi-aortic vessels. The aberrant vessel arises on the left side of the thorax and crossed to the right side behind the trachea and esophagus; therefore, it is also described as retroesophageal [3–5].

In the last years, it has been reported numerous variants of vascular ring. However, to the best of our knowledge, neither of the above case series had similar findings to our case.

## CASE REPORT

A 19-year-old lady was referred for suspected double aortic arch at 30+4 weeks' gestation. After performing a detailed sonographic examination of the fetal cardiac anatomy we found an ARSA, right ductus arteriosus, and an abnormal course of the aortic arch (Figure 1). The aortic arch was on the left side of the trachea in the upper mediastinum, while the descending aorta crosses on the right side giving suspicion of a circumflex retroesophageal aortic arch. After a prenatal counselling, the subsequent management of the pregnancy included delivery in hospital with pediatric cardiac intensive care unit and was planned invasive testing for the study of fetal karyotype and micro-array, which was declined by the parents.

The fetal growth was assessed by measuring head circumference, abdominal circumference, femur length, and the overall measurements were on the 5th centile for gestational age.

At 39 weeks gestation a female infant was delivered by emergency caesarean section for intrapartum anomalous cardiocography. Birth weight of the infant was 2630 g.

Postnatal echocardiography confirmed the presence of ARSA and revealed additional findings: PDA with bidirectional shunt. The ventricles showed normal cavity size, normal wall thickness, and normal function. No obstruction of the ventricular outflows was demonstrated.

Computed tomography images (CTIs) of the neonate (Figure 2) showed a normal origin of the aorta from the left ventricle, normal course of the ascendant aorta that in the upper axial plane of the mediastinum was on the left side of the trachea, then the descendant thoracic aorta ran on the right side of the vertebral column. There were also an ARSA and a suspected of Kommerell's diverticulum.

Kommerell's diverticulum is defined as a bulb-like swelling of the proximal portion of an aberrant subclavian

artery, adjacent to its aortic origin. It is a rare condition, which can occur with either the left aortic arch and ARSA (0.5–2% of the population) or with the right aortic arch and aberrant left subclavian artery (ALSA) (0.05–0.1% of the population).

The infant was symptomatic from the first days of life for stridor and regurgitation. Although the stridor improved when PDA closed spontaneously clinical picture was complicated by difficult swallowing, therefore at 31 days of life the infant underwent surgery for resection of the vascular ring. Intraoperative findings were: a circumflex retroesophageal aortic arch associated with ARSA and thick ligamentum arteriosum (Figure 3) that were the determining factors of the vascular ring. Surgeons performed the section of the ligamentum arteriosum, shifted the esophagus in the right side of the chest, ARSA was not sectioned and that is why after the section of the ligamentum the anatomical picture resulting in a "loose vascular ring." Postoperative course was normal with regression of the symptomatology causes by the vascular ring.

## DISCUSSION

Vascular rings are formed when one or more aortic arch abnormalities, with or without a PDA or ligamentum, produce a ring that encircles the trachea and esophagus.

Aortic arch anomalies may be associated with other congenital cardiac defects or chromosomal anomalies, such as microdeletion of chromosome 22q11. In neonatal/pediatric life, clinical presentation shows stridor and difficulty in swallowing or dysphagia. In prenatal life the 3-vessel and trachea view is fundamental for the diagnosis of abnormalities of the aortic arch [6]. The fluid-filled trachea can be easily seen on a fetal sonogram. Absence of a V-shaped confluence of the ductus arteriosus and aortic arch and the presence of any vessel coursing behind the trachea with formation of U-, 6-, or 9-shaped structures around the trachea help in making prenatal diagnosis of a vascular ring.

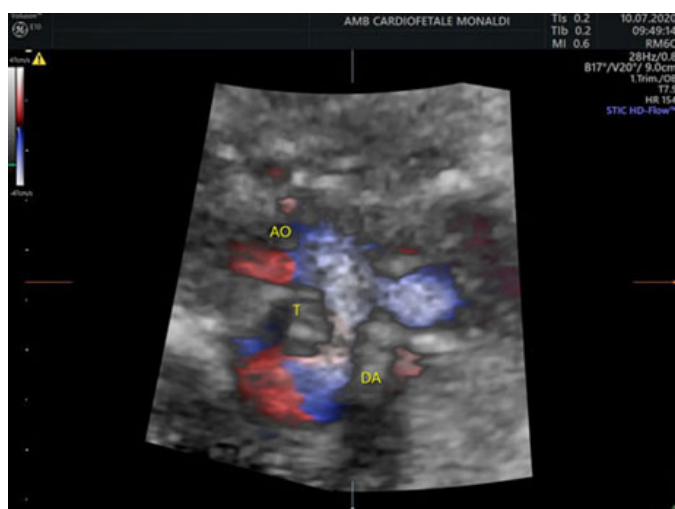


Figure 1: Three-dimensional color Doppler image, course of the aortic arch and ductal arch. Trachea (T), aortic arch (AO), and ductal arch (DA).

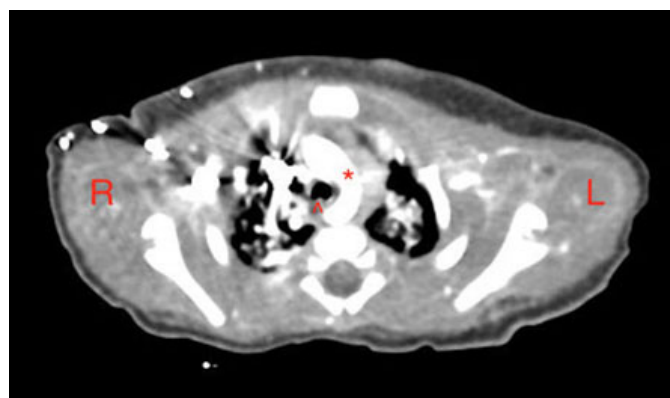


Figure 2: Post-natal contrast-enhanced CTI scan axial slice shows aberrant course of aortic arch (asterisk) with esophageal compression (arrow head), R (right), and L (left).

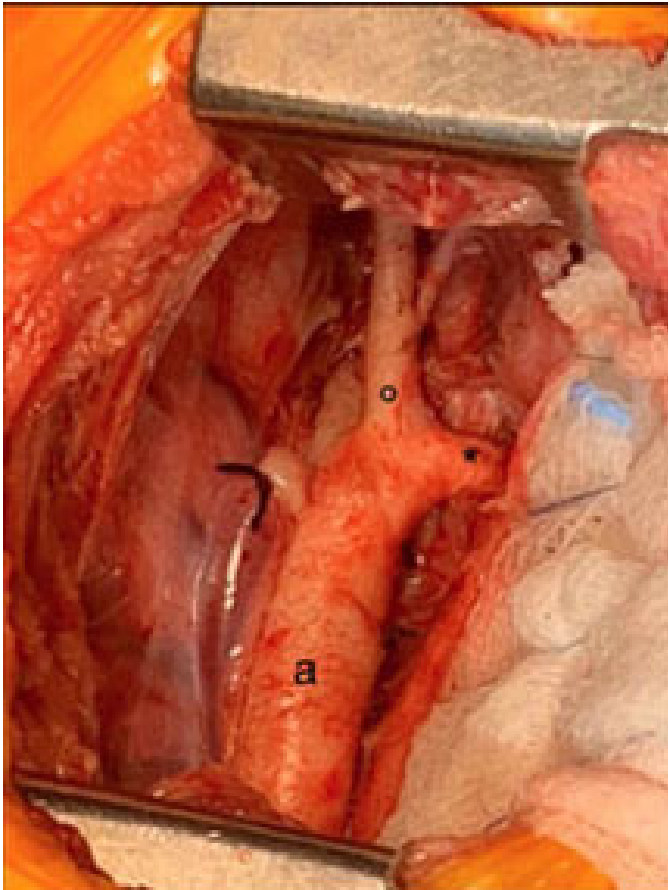


Figure 3: Surgical picture of the aorta with the great arteries and ligamentum arteriosum. The aorta (a), ligamentum arteriosum (asterisk), and left subclavian artery (o).

A right circumflex retroesophageal aortic arch is a very rare form of vascular ring [7, 8]. The aortic arch courses to the right of the trachea, turns to the left behind the trachea and esophagus, and descends on the left side.

In the literature it is reported that 90% of vascular rings fit into four main categories and there is also a classification from the Mayo Clinic that classified vascular rings into seven types.

To the best of our knowledge, we described a novel vascular ring resulting by the association between left circumflex retroesophageal aortic arch, right patent ductus arteriosus/ligamentum arteriosum and ARSA.

## CONCLUSION

We described a novel vascular ring in a neonate. In this extremely rare case, a “true” vascular ring, like in case of double aortic arch, it is due by the left circumflex retroesophageal aortic arch, ARSA, and PDA/ligamentum arteriosum. Interestingly, it is also that the circumflex retroesophageal aortic arch in the CTI was confused for a Kommerell’s diverticulum. We speculate that this is why the abnormal course of the aorta looks like an aneurysmatic dilatation.

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## Author Contributions

Beniamino Tormettino – Conception of the work, Design of the work, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Carmela Morelli – Conception of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Andrea Serrao – Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related

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Guido Oppido – Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maria Giovanna Russo – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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### **Conflict of Interest**

Authors declare no conflict of interest.

### **Data Availability**

All relevant data are within the paper and its Supporting Information files.

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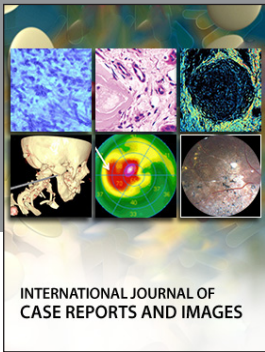
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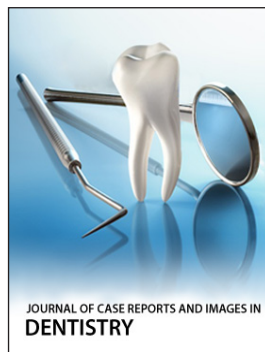
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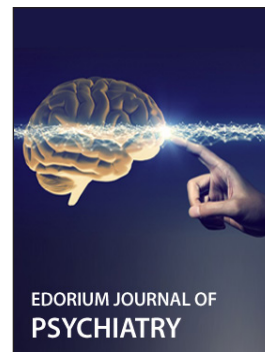
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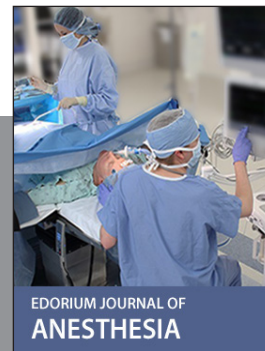
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