Right-sided aortic arch - what the radiologist should know

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Learning objectives

The aim of this educational exhibit is:

- To present a classification of the various types of right-sided aortic arch based on their embryological development.
- To outline their possible presentations and associations.
- To highlight the important radiological features to be recognized by the radiologist and the trainee.
Background

Right-sided aortic arch is a rare congenital abnormality which occurs in approximately 0.1% of the population. It refers to the variant right-sided location and course of the aortic arch in relation to the trachea and may be divided into three main types with respect to the pattern of the branching vessels of the aortic arch, the state of the ductus arteriosus and the association with cardiac anomalies. These include right-sided aortic arch with mirror image branching (59%), right-sided aortic arch with aberrant left subclavian artery (39.5%) and right-sided aortic arch with isolation of the left subclavian artery (1.5%) [1].

The aetiology of right-sided aortic arch is still unknown, although a deletion in chromosome 22q11 has been found to be associated with an increased incidence of anomalies of laterality of branching of the aortic arch [2].

Some types demonstrate an increased association with congenital heart disease which may present earlier on in childhood. Although these variants remain often asymptomatic and are found incidentally in adulthood, they may be the cause of important patient symptomatology. These may be related to compression on adjacent structures causing symptoms such as dysphagia, chest pain and shortness of breath.

The radiologist, therefore, has an important role to offer in appreciating and identifying the radiological features of such anomalies, with the ultimate aim of guiding effective treatment.

What follows is an overview of the main types of right-sided aortic arch, including their embryological development and their presentation and associations. Particular attention will be given to the important radiological findings.
Findings and procedure details

The aorta and its branches develop between the 4\textsuperscript{th} and 7\textsuperscript{th} weeks of gestation from six pairs of symmetrical pharyngeal arch arteries and the subsequent asymmetrical involution and persistence of specific structures. Specifically, the adult left-sided aortic arch develops from the fourth primitive left aortic arch with regression of the right aortic arch. When the opposite process occurs, that is the right arch persists and the left arch disappears, a right-sided aortic arch develops [2].

According to Edwards' hypothesis on the double aortic arch system, during embryonic development there is initially the formation of a double aortic arch with a ductus arteriosus on both sides encircling the trachea and the oesophagus. Subsequently, there is discontinuation of the portion of the right arch between the right subclavian artery and descending aorta and regression of the ductus arteriosus, resulting in the development of the normal left-sided aortic arch. Variations of these normal points of interruptions of the double aortic arch result in a number of anomalies of the aortic arch [3] Fig. 1 on page 7. On the other hand, persistence of the foetal double aortic arch results in the adult double aortic arch Fig. 2 on page 7.

Numerous classifications of right-sided aortic arch have been described. In 1948, Edwards provided the following classification according to the site of interruption of the hypothetical arch. In each case, the ductus arteriosus may be located on the left, on the right or bilaterally [2].

Type I: Right-sided aortic arch with mirror image branching
Type II: Right-sided aortic arch with aberrant left subclavian artery
Type III: Right-sided aortic arch with isolation of the left subclavian artery [4]

Right-sided aortic arch with mirror image branching:

Embryology: Following Edwards' model of the hypothetical double aortic arch, discontinuation of the posterior segment of the left aortic arch between the left subclavian artery and the descending thoracic aorta, together with regression of the right ductus arteriosus, results in a right-sided aortic arch with mirror image branching [3] Fig. 1 on page 7. The left ligamentum arteriosum or ductus arteriosus is often attached
to the left brachiocephalic artery anteriorly. A vascular ring is rarely formed when the ligamentum arteriosum or the ductus arteriosus is attached to the aortic diverticulum posteriorly and to the pulmonary artery anteriorly [5].

**Associations and presentation:** Cyanotic congenital heart diseases, such as tetralogy of Fallot, truncus arteriosus, tricuspid atresia and transposition of the great arteries, are often associated with this variant [3][6]. Although generally asymptomatic, in rare cases, children may present with symptoms of tracheoesophageal compression whilst adults may present with symptoms secondary to atherosclerotic hardening of the aortic arch or compression of mediastinal structures by aneurysmal dilation. In the rare event of vascular ring formation, severe symptoms, secondary to compression of encircled structures, may be present [5].

**Imaging findings:** Since patients usually remain asymptomatic, this variant is often identified incidentally on imaging performed for other reasons. A plain radiograph of the chest may reveal features common to all types of right-sided aortic arch, including the absence of the left aortic contour, bowing of the trachea to the left at the level of the right-sided arch, a right-sided descending thoracic aorta, and a right-sided aortic knuckle which may be taken for a high riding mass in the right paratracheal region [7] **Fig. 3 on page 8.** Cross-sectional imaging demonstrates a right-sided aortic arch with sequential branching of the left brachiocephalic trunk, the right common carotid artery and finally the right subclavian artery. The brachiocephalic trunk continues anterior to the left pulmonary artery after which it divides into the left subclavian and left common carotid arteries [8] **Fig. 4 on page 9.**

**Right-sided aortic arch with aberrant left subclavian artery:**

**Embryology:** This fairly common variant results from the interruption of the posterior portion of the left aortic arch between the left common carotid artery and the left subclavian artery, and regression of the right ductus arteriosus [3] **Fig. 1 on page 7.** A left-sided ductus arteriosus runs from the aortic remnant to the left pulmonary artery with the formation of a complete vascular ring [9].

**Associations and presentation:** Right-sided aortic arch with aberrant left subclavian artery is rarely associated with congenital cardiac anomalies [3]. It is however usually associated with Kommerell's diverticulum, the bulb-like aneuysmal dilatation of the right-sided aortic arch at the origin of the aberrant left subclavian artery [2]. The vascular ring is generally loose and in most cases is asymptomatic, with only a small fraction of patients having a tight enough ring to result in tracheoesophageal compression. Upper respiratory tract symptoms occur most commonly in children since the trachea is more
easily compressible [10]. Adults have a more rigid trachea and are therefore usually asymptomatic or occasionally present with symptoms of oesophageal compression, such as dysphagia lusoria [11].

*Imaging findings:* In addition to the features of right-sided aortic arch on a plain chest radiograph mentioned earlier, a Kommerell’s diverticulum may present as a left para-spinal soft-tissue density. Digital angiography and cross sectional imaging is however necessary to identify the exact anatomy [12] *Fig. 5 on page 10.*

**Right-sided aortic arch with isolation of the left subclavian artery:**

*Embryology:* A right sided-aortic arch with isolation of the left subclavian artery results from interruption of the left arch at two different sites, the first between the left common carotid artery and the left subclavian artery and the second immediately following the attachment of the left ductus arteriosus [3] *Fig. 1 on page 7.* The isolated left-sided subclavian artery does not communicate with the aortic arch, but is instead supplied by retrograde flow from the left vertebral artery [13]. It is also in communication with the pulmonary artery via a left ligamentum arteriosum or, less commonly, a patent ductus arteriosus [13] [14].

*Association and presentation:* Congenital subclavian steal syndrome and vertebrobasilar insufficiency may result secondary to the anatomy of this variant [3]. Moreover, the presence of a patent ductus arteriosus can cause pulmonary steal syndrome. In very rare cases, congenital heart diseases such as tetralogy of Fallot, may be found in association [3][14].

*Imaging findings:* In addition to the role of cross-sectional imaging and angiography in delineating the exact anatomy of this variant, colour-coded duplex ultrasonography may reveal reversal of low within the left vertebral artery as is demonstrated in the provided case *Fig. 6 on page 11 Fig. 7 on page 12.*

One can conclude that the branching pattern and anatomy of the left subclavian artery may guide the radiologist in identifying the variant type of right-sided aortic arch on cross-sectional imaging.
Fig. 1: Edwards' hypothetical double aortic arch system. A: Interruption of the dorsal segment of the left arch between the left subclavian artery and the descending aorta with regression of the right ductus arteriosus leads to the formation of a right-sided aortic arch with mirror image branching. B Interruption of the dorsal segment of the left arch between the left common carotid artery and the left subclavian artery with regression of the right ductus arteriosus leads to the formation of a right-sided aortic arch with an aberrant left subclavian artery. C: Formation of the right-sided aortic arch with an isolated left subclavian artery results from interruption at two levels, between the left common carotid artery and the left subclavian artery and distal to the attachment of the left ductus arterioles. RCA= right common carotid artery, LCA=left common carotid artery, RSA=right subclavian artery, LSA=left subclavian artery, RDA=right ductus arteriosus, LDA=left ductus arteriosus, PA=main pulmonary artery, RPA=right pulmonary artery, LPA=left pulmonary artery, BCT=brachiocephalic trunk, ALSA=aberrant left subclavian artery, ILSA=isolated left subclavian artery.
Fig. 2: A: Plain chest radiograph demonstrates a right paratracheal mass and a left-sided aortic knuckle identified as a double aortic arch causing narrowing of the trachea. B: Coronal MIP confirms a double aortic arch (red arrows) encircling the trachea and oesophagus.

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Fig. 3: Plain chest radiograph demonstrates absence of the left aortic contour, bowing of the trachea to the left at the level of the right-sided arch, a right-sided descending thoracic aorta, and a right-sided aortic knuckle (red arrow) which may be interpreted as a high riding mass in the right paratracheal region.

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Fig. 4: Axial contrast-enhanced MR angiogram in a patient with repaired tetralogy of Fallot: The images show sequential branching of the left brachiocephalic trunk, the right common carotid artery and finally the right subclavian artery from a right-sided aortic arch. The brachiocephalic trunk further divides into the left subclavian and left common carotid arteries. These findings are characteristic of a right-sided aortic arch with mirror image branching.

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**Fig. 5:** Axial CT thorax (porto-venous phase) in a 60 year old gentleman who presented with chest pain, widened mediastinum and tracheal deviation to the left on chest X-ray: The series of images demonstrate a right-sided aortic arch with age-related mural calcification. The first branch arising from the arch is the left common carotid artery followed by the right common carotid artery, the right vertebral artery and the right subclavian artery. A Kommerell diverticulum is seen just posterior to the oesophagus, giving rise to the left subclavian artery which impinges slightly on the oesophagus. These findings in are in keeping with right-sided aortic arch with an aberrant left subclavian artery.

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Fig. 6: A: Coronal MIP shows a right sided aortic arch with an isolated left subclavian artery (red arrow). B: 3D angiographic volume rendered image showing an isolated left subclavian artery (red arrow) supplied by the left vertebral artery and thoracic vessels (green arrows).

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**Fig. 7:** Colour Doppler imaging and spectral Doppler analysis of the left vertebral artery demonstrates reversal of flow within the left vertebral artery towards the isolated left subclavian artery.

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Conclusion

This educational exhibit demonstrates that knowledge of the embryological development of a right-sided aortic arch and its variants leads to proper image interpretation and a better understanding of pathology, with a view to improving patient treatment and care.
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