



# Right Aortic Arch with Bicarotid Trunk and Isolated Left Subclavian Artery: Hitherto Unreported Pattern

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## Abstract

### Keywords

- ▶ right aortic arch
- ▶ isolated left subclavian artery
- ▶ bicarotid trunk
- ▶ arch anomalies
- ▶ tetralogy of Fallot
- ▶ multidetector CT

Variant arch anatomy may be seen associated with many congenital heart diseases. Its accurate preoperative identification is of paramount importance in optimal surgical planning of such cases. This case describes one such variant arch anatomy with two vessel right aortic arch, comprising of bicarotid trunk (giving rise to bilateral common carotid arteries) and right subclavian artery with isolation of the left subclavian artery, in a patient with tetralogy of Fallot. Right aortic arch with isolated left subclavian artery has already been described in association with tetralogy of Fallot. However, to the best of our knowledge, present arch pattern consisting of right aortic arch with bicarotid trunk and isolated left subclavian artery has not been reported in literature so far. Moreover, this case highlights the utility of multidetector computed tomography in accurate identification of variant arch anatomy in addition to delineation of cardiac and extracardiac details.

## Introduction

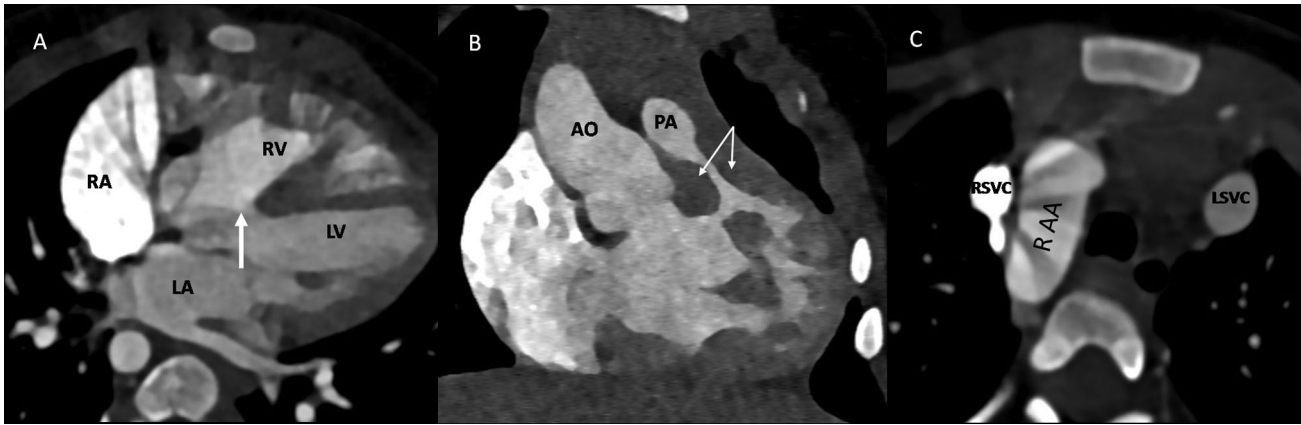
Embryological development of the aorta begins as primitive dorsal and ventral aorta in the third week of life. There are six paired pharyngeal arches between the dorsal and ventral aorta. Aortic arch anomalies transpire from the abnormal persistence or regression of these primitive arches. Right aortic arch (RAA) is formed due to embryological persistence of the right primitive dorsal aorta. Literature commonly describes three different RAA patterns including RAA with mirror image branching pattern (type I), with aberrant left subclavian artery (SCA) (type II), and rarely with isolated left SCA (type III). In these cases, the first branch is generally the

left innominate artery or the left common carotid artery (CCA) followed by the right CCA depending on the type of the RAA. The presence of bicarotid trunk (BCT) is more frequently described with left aortic arch and associated aberrant right SCA.<sup>1–3</sup> A very recent report highlights its association with RAA and aberrant left SCA.<sup>4</sup> We, hereby, present a complex and unfamiliar arch anomaly in a 12-month-old infant with tetralogy of Fallot (TOF), where RAA was seen associated with presence of BCT and isolated left SCA.

## Case Details

A 12-month-old cyanotic infant with echocardiographic diagnosis of TOF underwent computed tomographic (CT) angiography. It revealed characteristic imaging features of TOF including large subaortic ventricular septal defect (VSD) with aortic

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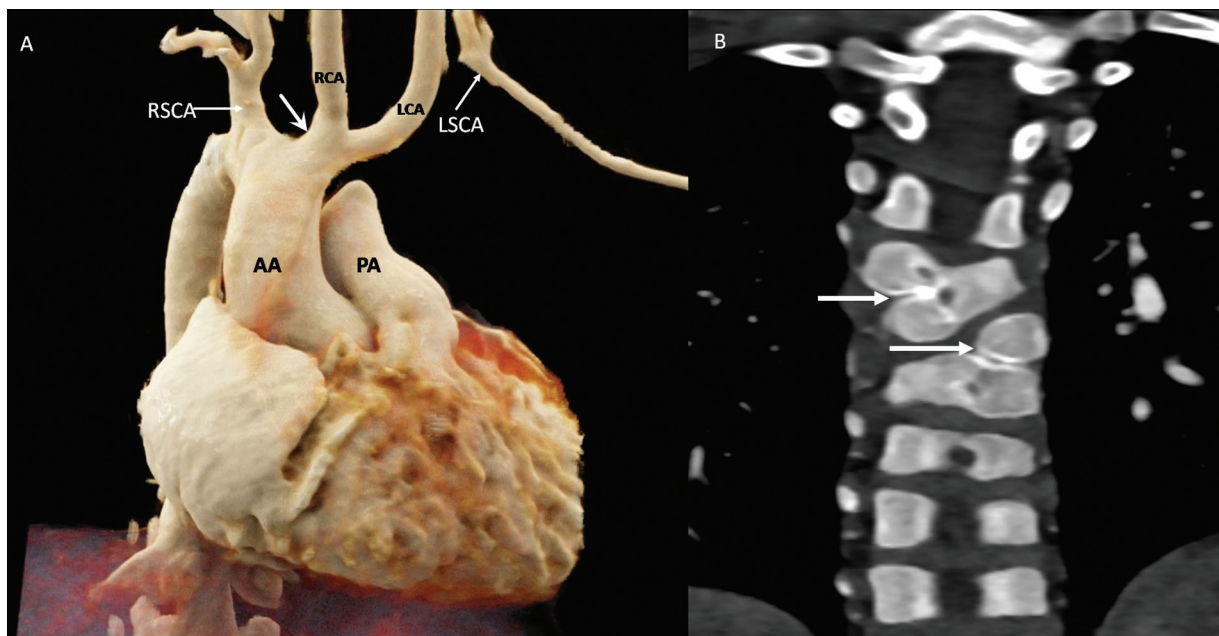


**Fig. 1** (A–C) Computed tomography angiography images showing ventricular septal defect (arrow in A) with aortic override, infundibular pulmonary stenosis (grouped arrows in B), right aortic arch (RAA), and double superior vena cava. AO, ascending aorta; LA, left atrium; LSVC, left superior vena cava; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RSVC, right superior vena cava; RV, right ventricle.

override, and infundibular pulmonary stenosis (→Fig. 1A, B) with confluent good sized pulmonary arteries. Interestingly, an unusual aortic arch pattern was seen in the form of RAA (→Fig. 1C) with origin of two arch vessels from the aortic arch. BCT arises as the first branch of the RAA (giving rise to the left and the right CCA), followed by the right SCA. Left SCA origin from the RAA was not seen. Instead, it was seen reforming via ipsilateral vertebral artery with occluded arterial duct (→Fig. 2A). Additionally, associated double superior vena cava (→Fig. 1C) and thoracic vertebral segmentation anomalies (→Fig. 2B) were also seen. Pulmonary venous drainage was normal. No airway compression, significant aortopulmonary collaterals, coarctation, or anomalous coronary artery were seen.

**Discussion**

RAA with isolated left SCA is a rare arch anomaly that represents the least common type of RAA.<sup>5</sup> Here, the left SCA is disconnected from the aortic arch and is instead attached to the pulmonary artery via the ductus arteriosus. On spontaneous closure of the arterial duct, SCA draws blood via the left vertebral artery or other thoracic collaterals. Majority of the cases are seen associated with other congenital heart diseases, most commonly TOF and other conotruncal abnormalities.<sup>6</sup> Moreover, such patients usually present early with symptoms related to the underlying congenital heart disease (cyanosis in cases of TOF or failure to thrive in cases of large patent arterial duct). Isolated cases are mostly



**Fig. 2** (A–B) Volume rendered computed tomography angiography image (A) showing right aortic arch with bicarotid trunk (arrow) as the first branch, giving rise to the left (LCA) and the right (RCA) common carotid arteries followed by the origin of the right subclavian artery (RSCA). Left subclavian artery (LSCA) was not seen arising from the arch and was seen reforming through left vertebral artery. Coronal reformatted image (B) showing associated vertebral segmentation anomalies (arrows). AA, ascending aorta; PA, pulmonary artery.

asymptomatic initially, but later they can present with features of left limb ischemia and small limb due to poor perfusion. It can also be associated with the subclavian steal phenomenon leading to vertebrobasilar insufficiency and posterior circulation infarcts.

BCT is a variant branching pattern that is more commonly being reported with left aortic arch and associated aberrant right SCA.<sup>1-3</sup> Rarely, it has been reported with double aortic arch or RAA with aberrant left SCA, in association with congenital heart diseases, including large aortopulmonary window or VSD, respectively.<sup>4,7</sup> However, its association with RAA and isolated left SCA has not been reported in literature previously. Persistence at an early stage of development with a common trunk origin of both carotid arteries from the third arch might explain the occurrence of BCT.

Anomalies of the aortic arch and its branches must be evaluated in detail because of their implications for endovascular interventions and cardiovascular surgical planning. Preoperative identification may allow simultaneous correction along with intracardiac repair. Moreover, it may help in planning the cannulation strategy. In patients requiring preoperative embolization, it may help in choosing the optimal hardware with reduction in fluoroscopy time and contrast usage. Multidetector CT allows optimal evaluation of the arch anatomy and provides additional important information regarding pulmonary artery size, anomalous coronaries, aortopulmonary collaterals, airway and lung parenchymal abnormality in addition to delineation of cardiac anatomy.<sup>8,9</sup>

#### Funding

None.

#### Conflict of Interest

None declared.

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