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**Dr. Surbhi Singh**  
Gurunanak Dev  
Superspeciality Hospital,  
Tarn Taran, India

**Dr. Akhil Tanwar**  
Government Medical College,  
Amritsar, Punjab, India

## A unique constellation of multiple aortic and caval anomalies-Truncus bicaroticus with right aberrant subclavian artery, and persistent left sided SVC with left sided azygous vein-imaging and clinical implications

**Dr. Surbhi Singh and Dr. Akhil Tanwar**

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### Abstract

**Introduction:** A case investigated for mild dysphagia, turned out to be an interesting amalgam of multiple vascular anomalies involving rare caval and aortic arch variants including-persistent left SVC, left azygous vein, aberrant right subclavian artery and truncus bicaroticus. A literature review has revealed a number of case reports on these entities in isolation but no case with such a constellation of vascular variants coexisting in the same individual has been reported till date according to our knowledge and research.

**Case Report:** Barium swallow study showed posterior indentation on esophagus. On MDCT, an abnormal course of RSA was seen behind the esophagus. Bilateral carotids were seen to be arising from a common midline trunk- often called truncus bicaroticus double SVC was seen with persistent left SVC draining into the coronary sinus. A prominent retroaortic azygous vein was seen draining into the left SVC with absence of right azygous.

**Discussion:** Anatomic and embryological basis of these anomalies are discussed with review of respective literature that highlights the imaging characteristics of aortic and caval anomalies and their clinical significance. It aims at better understanding of such vascular anomalies for radiologists based on CT findings, in order to identify and subsequently report them elaborately for a clearer surgical roadmap.

**Keywords:** Aortic anomalies, caval anomalies, aberrant RSA, left sided SVC, truncus bicaroticus

### Introduction

Aberrant right subclavian artery is the most common congenital thoracic aortic arch anomaly; however, its coexistence with truncus bicaroticus, which is a common origin for both common carotids is rare. Even rarer is the coexistence of these aortic anomalies with a venous caval anomaly, in this case a persistent left SVC with left sided azygous vein.

A 45 year old female patient presented to our facility with complaints of dysphagia since past year. On performing the basic barium swallow examination, mild posterior indentation on the esophageal wall was seen with slight hold up of contrast proximal to it. She was further evaluated with contrast enhanced MDCT to confirm the diagnosis which revealed various other rare vascular anomalies in the same patient including-right aberrant subclavian artery, Truncus bicaroticus, persistent Left sided SVC draining into coronary sinus and Left sided azygous vein.

This article provides insight into the embryonic basis of these anomalies and review the practical implications of their presence and the key role of imaging in their identification and thus avoiding clinical mishaps.

### Case findings

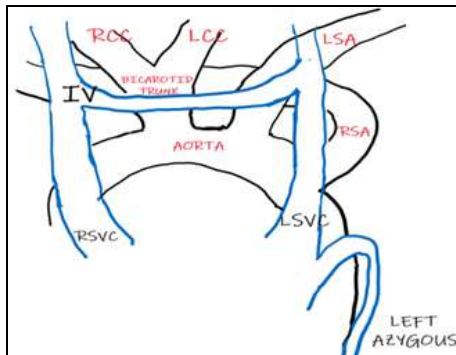
Initial investigation for dysphagia with barium study was done which showed posterior indentation on esophageal column. This was followed by further evaluation by CECT. Upon review of CECT, the patient had three branches of the aortic arch, proximal most being a common trunk which gave rise to right and left common carotids consistent with truncus

**Corresponding Author:**  
**Dr. Surbhi Singh**  
Gurunanak Dev  
Superspeciality Hospital,  
Tarn Taran, India

bicaroticus. The second branch was the left subclavian artery. The brachiocephalic trunk was absent. The third and most distal origin artery passed posterior to the oesophagus as it crossed the midline to supply the right arm suggestive of an aberrant right subclavian artery (ARSA) or *arteria lusoria*. Compression of the esophagus was noted posteriorly causing its luminal narrowing. The right vertebral artery was seen to be arising from the right carotid artery.

On evaluation of the venous system in the thorax, persistent left SVC was found which was draining into the coronary sinus. The left superior vena cava (LSVC) began from the confluence of the left internal jugular and left subclavian veins, and extended downwards medially into the left edge of the coronary sinus. The right superior vena cava was formed by the union of the right internal jugular and right subclavian veins, and drained into the right atrium. A narrowed innominate trunk is seen connecting the two SVCs.

The Azygous vein was formed by the union of the right and left ascending lumbar veins at the level of the tenth thoracic vertebra. It ascended along the left margin of the thoracic vertebra following a retroaortic approach, receiving bilateral posterior intercostal veins and then opened into the LSVC via the azygos arch. Thus forming a left sided azygous vein. Azygous or hemiazygos vein on the right side could not be identified.



**Fig 1:** 1. Schematic diagram of all the aortic and venous anomalies encountered in the case, which are as follows- Right aberrant subclavian, 2. Truncus bicaroticus, Left svc draining into coronary sinus, Left sided azygous vein draining into LSVC.

(RCC-Right common carotid, LCC-left common carotid, LSA-left subclavian artery, RSA-right subclavian artery, RSVC-right superior vena cava, LSVC- left sided SVC, IV-innominate vein)

## Discussion

Arteria lusoria/Aberrant right subclavian artery with truncus bicaroticus.

**Anatomy:** Aberrant right subclavian artery is the most common congenital aortic arch anomaly with a prevalence of 0.4-2% [1]. It is an aortic arch anomaly where the right subclavian artery arises as the distal most branch of the arch, from the left side and courses behind the esophagus and trachea to reach the right arm. The artery crosses the midline between the oesophagus and spine. Often, ARSA arises from a diverticulum at the proximal descending aorta, called the Kommerell's pouch [2].

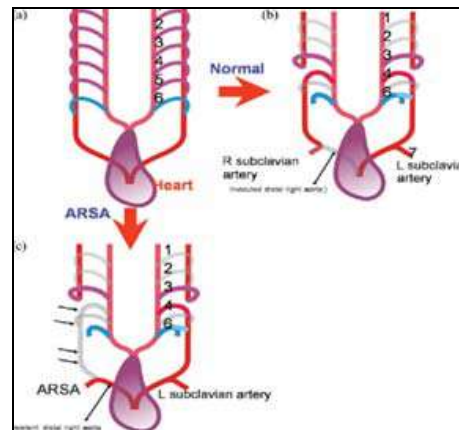
Another variation observed in this case was the Right and

left CCA originating from a shared common trunk, known in the literature as Truncus bicaroticus. This trunk was the first branch of the aortic arch with absence of the brachiocephalic trunk and two subclavian arteries arising as separate aortic arch branches. The coexistence of ARSA and truncus bicaroticus is rare. The existence of both a TB and a ARSA is reported in up to 4.0% in the general population [3].

A truncus bicaroticus is also associated with other vascular anomalies such as VSD, ASD, valvular pulmonary stenosis, and genetic anomalies like Down's, Williams, and Charge. [4]

## Development

The proximal part of the right subclavian artery originates from the fourth right aortic arch and distal part from the involution of the right dorsal aorta and right seventh intersegmental right arteries, which originate from the descending aorta. Anomalous development of ARSA occurs when there is involution of the fourth right aortic arch, with persistence of the right dorsal aorta and the seventh intersegmental artery, which remains fixed to the aortic arch in the descending part. When the dorsal aorta rotates the right seventh intersegmental artery becomes the ARSA and arises as a last aortic branch [1, 5].



**Fig 2:** a) Shows primitive embryological aortic arches, b- shows normal development of RSA where fourth aortic arch forms its proximal part and involution of right distal aorta and 7<sup>th</sup> right intersegmental artery forms its distal part. c- shows anomalous development of ARSA due to involution of the right fourth vascular arch with the persistence of the proximal right dorsal aorta and of the right seventh intersegmental artery.

The bicarotid trunk is an extremely rare anomaly of the aortic arch with a reported prevalence of <0.1% and develops because the third pair of primitive aortic arch persists [4].

Normally, the proximal part of the third aortic arch gets absorbed into the left horn of the aortic sac. Rarely, it does not get absorbed, with extension and absorption of the left third (III) aortic arch artery into the right horn of the aortic sac, which normally forms the brachiocephalic trunk. Since in this case the right subclavian is aberrant and arising as the distal most branch, the brachiocephalic trunk does not exist and thus the common trunk gives rise to the left and right common carotid arteries and it consequences into the "Truncus bicaroticus" [6].

**Clinical Aspects:** Arteria lusoria is usually asymptomatic

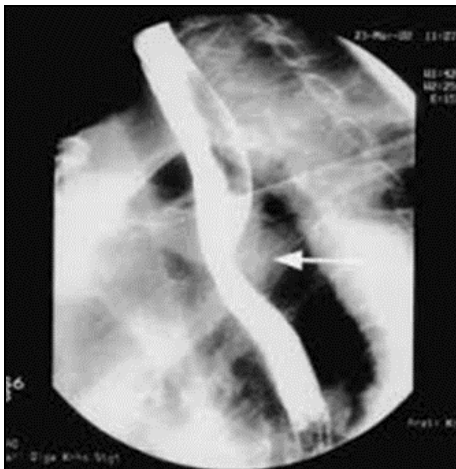
and is most often discovered incidentally during evaluation of other mediastinal anomalies, such as in our case [5, 6]. But it has been reported that when it coexists with truncus bicaroticus, both these anomalies together, may contribute and add to “dysphagia lusoria,” that is difficult swallowing due to an aberrant artery [7]. It is suggested that the ARSA puts pressure on the esophagus, compressing it anteriorly against the trachea; and the truncus bicaroticus does not allow forward translation of the trachea and esophagus, leading to dysphagia [4, 7]. In our case, the patient had a history of mild and intermittent dysphagia caused by solid foods, the condition was elaborately explained to her and she chose to be under follow up for the same.

ARSA can sometimes get complicated with conditions like arteritis, aneurysmal dilatation and rarely arterial dissection, thus posing a clinical threat. Mulligan *et al* suggested that an ARSA shows susceptibility to atherosclerosis and its associated complications, including aneurysms, dissections and stenosis, however no such complications were seen in our observation. Aneurysmal dilatation of the proximal segment of an ARSA called kommerals diverticulum occurs in 8% of cases [7].

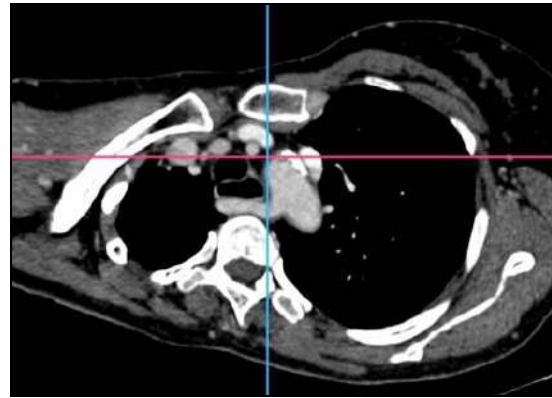
Presence of ARSA can also pose clinical dilemmas in various thoracic surgeries, like the transradial approach (TRA) used for acute coronary disease. The TRA coronary procedures with the presence of ARSA may result in problems; like arterial injury. According to the literature, only 60% success rate of TRA was seen in patients with ARSA [8].

**Imaging:** On barium studies, Arteria lusoria shows an oblique pulsating defect on the posterior aspect of the esophagus, passing from left to right just above the aortic arch. In the anterior and lateral views, a wedge-shaped impression on the dorsal aspect of the esophagus, as seen in the present case [9]. The actual vascular anatomy and the relationship with the surrounding structures can be most precisely demonstrated with CT or MR angiography [11].

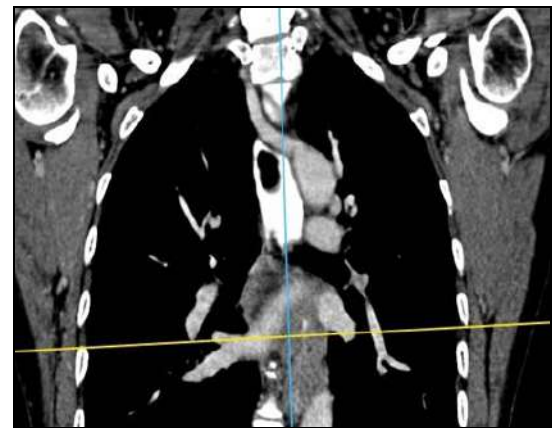
In this case barium study was done for mild intermittent dysphagia which showed posterior indentation on esophageal column and prompted to investigate further with MDCT. CECT angiography was done which proved to be diagnostic in identifying the vascular anomalies in this case, observing a bicarotid trunk arising first from the aortic arch, the second branch was the left subclavian artery, and the third and last was the ARSA, passing the right behind the esophagus.



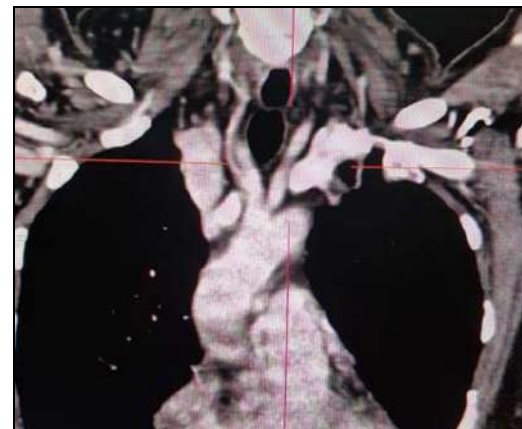
**Fig 3:** barium swallow lateral view image showing posterior indentation on esophagus by the ARSA



**Fig 4:** CECT axial shows the aberrant course of the RSA, behind the esophagus compressing it anteriorly against the trachea



**Fig 5:** CECT coronal section shows ARSA arising from the left side of aortic arch from a small diverticulum (kommerals)



**Fig 6:** CECT coronal section shows origin of both common carotids from a common trunk-truncus bicaroticus.

### Persistent Left Superior Vena Cava with Left Sided Azygous Vein

**Anatomy:** Persistent left superior vena cava (PLSVC) is an uncommon vascular anomaly; however, it is the most common anomalous systemic vein-to-cardiac connection. PLSVC is found in approximately 0.3-0.5% of the general population, and its occurrence along with absent right-sided SVC is reported in 0.1% of the general population. Up to 90% of the people with persistent left SVC present with the right SVC as seen in the present case. About 65% of the people with persistent left SVC have no left brachiocephalic



vein or atrophic one [12].

In present case, both right and left SVC was seen with a narrow bridging innominate vein connecting them, which appears to be lying just anterior to and being compressed by the truncus bicaroticus in right paramedian location.

Rarely, the caudal right superior cardinal vein regresses leading to an absent right SVC with PLSVC, where the left SVC returns all the blood from cranial aspect of the body. In maximum cases, PLSVC drains into the right atrium via the coronary sinus and does not cause any hemodynamic consequence, as seen in this case. In few cases, it may drain in left atrium resulting in a right to left sided shunt [13].

**Left Azygous Vein:** Absence of azygous vein and arch or any other venous structure in the right paravertebral location on the right side suggests an aberrant azygous system. A prominent venous system was seen on the left side, into which intercostal veins from both sides and bilateral lumbar veins drained. It opens on the left side of the left SVC, suggestive of a left sided azygous system [14].

### Development

**PLSVC:** In a normal case, in the 4-week-old embryo, the common cardinal veins system consists of the paired anterior and posterior cardinal veins. The common cardinal veins extend into the sinus venosus of the primitive heart. In the 7-week-old embryo, the anastomosis vein appears between the right and left anterior cardinal veins. In the 12-week-old fetus, the proximal part of the left common cardinal vein becomes the coronary sinus, and extended into the right atrium. The remainder distal part of left common cardinal vein disappears. The anastomosis vein becomes the left brachiocephalic vein. The right common cardinal vein becomes the superior vena cava [14, 15]. In anomalous cases when the left common cardinal vein fails to be obliterated, it becomes the persistent left SVC and opens into the coronary sinus, as seen in this case.

**Left Azygous Vein:** In the 6-week-old embryo, the bilateral supracardinal veins appears, and extends into the posterior cardinal veins, the bilateral supracardinal veins develop inferiorly. At 9 weeks, the anastomosis vein appears between the right and left supracardinal veins. The proximal part of the right posterior cardinal vein remains and forms the root of the azygos vein. Azygos vein is formed by the right supracardinal vein. The connection between right and left supracardinal veins forms at the level of the sixth thoracic vertebra. The left supracardinal vein forms the hemiazygos which extends into the azygos vein via the anastomosis vein. The left supracardinal vein undergoes obliteration cranial to the anastomotic site, or it may persist as the accessory hemiazygos vein. The azygos vein extends into the superior vena cava [16].

In the present case, the possible developmental mechanism of the left azygos vein could be- In the 12-week-old fetus, the proximal part of the left posterior cardinal vein remained, and the root of the left azygos vein was formed. The right supracardinal vein was separated to the right hemiazygos and the accessory hemiazygos veins. The right hemiazygos vein extended into the left azygos vein via the anastomosis vein. The left azygos vein extended into the left superior vena cava

**Clinical Aspects:** PLSVC is usually asymptomatic and is

detected when cardiovascular imaging is performed for other reasons. It is most commonly observed in isolation but can be associated with other cardiovascular abnormalities including atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum.

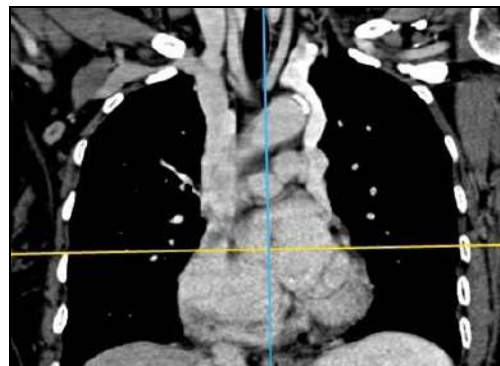
There are many clinical implications to consider when variant venous vasculature is discovered. The presence of PLSVC can render access to the right side of heart challenging via the left subclavian approach, which is a common site of access utilized when placing pacemakers, Swan-Ganz catheters and implantable cardioverter-defibrillators. Cannulation of PLSVC may be required during surgery which depends on several factors: the presence or absence of the innominate vein (between the two venae cava), the absence of the right superior vena cava, its caliber, blood flow through the left superior vena cava [16]. Hence it is important for the radiological report to cover all the necessary data in the preoperative planning.

PLSVCs can also complicate cardiac surgery. It is an absolute contraindication for retrograde cardioplegia as perfusion up the PLSVC can lead to compromised myocardial protection. PLSVC together with other cardiac malformations may increase postoperative mortality after cardiac surgery, making its identification crucial in preventing intra and postoperative complications [17].

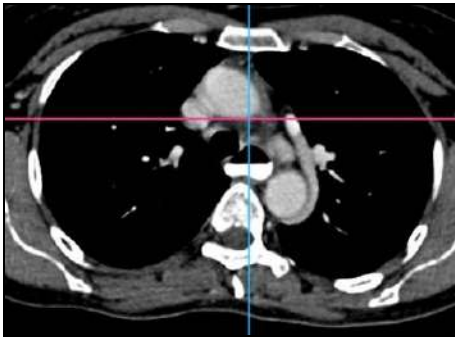
Other important surgical implications of PLSVC can be during cardiac transplantation, when cardiopulmonary bypass is required. Few authors postulated that PLSVC may be associated with several cardiac arrhythmias, present in 36% of cases (23), which has been explained by several mechanisms such as possible histological modifications in the atrioventricular sinus, existence of multiple electric nodes between PLSVC, coronary sinus and right atrium [17].

It is important to identify variations of the azygos system when mediastinum CT is performed, as they can get confused with an aneurysm, lymphadenopathy, or other pathologies.

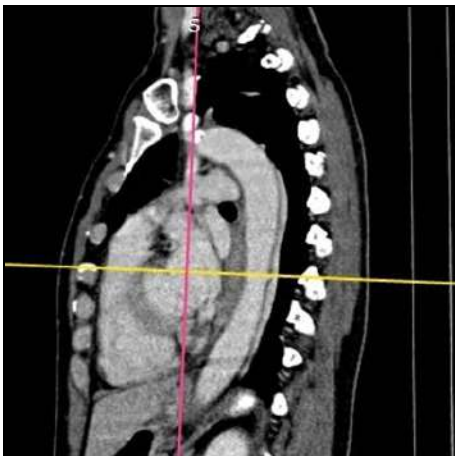
**Imaging:** Diagnosis of left SVC and azygous anomalies is usually made as an incidental finding during cardiovascular imaging or surgery. A contrast enhanced MDCT scan done in the early venous phase, at about 45-60 seconds post contrast injection, gives an elaborate picture of the venous system. For reducing radiation exposure, a single phase venous study can be done to visualize both aortic and caval anomalies.



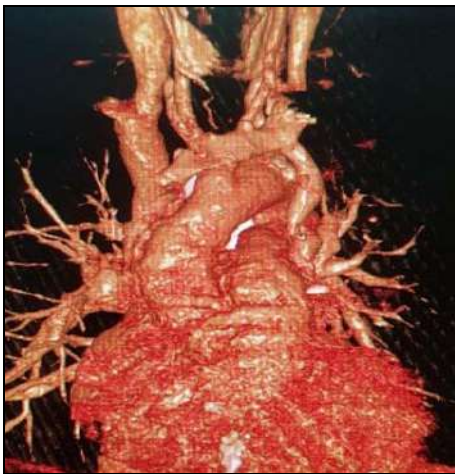
**Fig 5:** CECT coronal shows the persistent left sided SVC draining into the right atrium through the coronary sinus, along with the right SVC.



**Fig 6:** Axial CECT shows a prominent azygous vein on the left side, coursing on the lateral side of the aorta to drain into the left SVC



**Fig 7:** CECT sagittal shows the retroaortic course of the left sided azygous vein



**Fig 8:** Volume rendered CT image showing the left and right SVC, connected by an innominate vein. Common trunk for origin of bilateral carotids is also seen just being the innominate vein.

Recent studies have suggested that cross sectional imaging (CT/MRI) is the best means of investigation, being significantly superior to both transthoracic and transesophageal echocardiography. Economic constraints and limited availability of MRI makes CT the mostly used investigation for identifying caval anomalies in developing countries. However, radiation exposure, and iodinated contrast use has its own limitations.

### Conclusion

Review of embryology indicates that occurrence of aortic arch and venous caval anomalies, is more likely a coincidence and the various anomalies coexisting in this case are not related. Knowledge of their embryological development and anatomy aids in better understanding of the same and planning further steps to alleviate any clinical outcomes of such anomalies.

By making use of three dimensional capabilities of both CTA and MRA, anatomical relationship of the aortic arch with trachea and esophagus can be defined properly thus avoiding invasive techniques like bronchoscopy and conventional angiography. The radiology report should describe all the anatomical and functional details, high risk vascular characteristics, and differential considerations in order to best aid preoperative surgical planning and prevent any unnecessary obstacles in diagnosis and treatment of these conditions.

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