

## CASE REPORT

# Double superior vena cavae with bilaterally symmetrical azygos veins and incomplete left circumflex artery

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## SUMMARY

Vascular variations should be considered seriously, since the majority of these are incidental findings during surgeries or catheterization. A prior knowledge of the possible existence of variations in the veins, especially in the vena cava, is necessary for surgeons, radiologists, or anaesthesiologists, since central catheterization procedures have been increased over the years. We are presenting double superior vena cavae, bilaterally symmetrical azygos veins, and an incomplete left circumflex coronary artery, which were noted during routine dissection of a 65-year-old male cadaver. Knowledge of the combination of these variations makes this case significant during cardiothoracic surgeries. The embryological basis and clinical significance of the above-mentioned vascular aberrations have been discussed.

**Key words:** Double superior vena cavae – Left superior vena cava – Left azygos vein – Left circumflex artery – Variations – Embryological significance

## INTRODUCTION

Awareness in the variations of veins should be given paramount importance during surgical and radiological interventions, since the existence of such variants may lead to fatal complications during catheterization. Persistent LSVC is a silent venous anomaly, since it will be revealed normally during venous catheterization in the majority of the cases (Josloff and Kukora, 1995), and it is usually asymptomatic, that is, it does not require treatment unless accompanied by other cardiac variations. Developmentally, the superior vena cava derives from the right anterior cardinal vein and the right common cardinal vein (Moore et al., 2008). The left anterior cardinal vein usually regresses. However, if the left anterior cardinal vein fails to regress it will result in a persistent LSVC; hence there will be two superior vena cavae. The abnormal LSVC, derived from the left anterior cardinal and common cardinal veins, opens into the right atrium through the coronary sinus (Bergman et al., 1988; Cormier et al., 1989). The present case reports the existence of LSVC in association with an incomplete circumflex

artery. Its developmental basis along with clinical significances is discussed in the report.

## CASE REPORT

During dissection of a 65-year-old male cadaver in the department of Anatomy, Kasturba Medical College, Mangalore, affiliated to Manipal University, multiple vascular variations were observed in the thoracic cavity. The most striking variation was the presence of the double superior vena cava as shown in Fig. 1. The left brachiocephalic vein (LBCV) divided into two unequal divisions. A much wider lateral division continued as the left superior vena cava(\*) and a thin, elongated medial division (2) crossed the midline to communicate with the right brachiocephalic vein. This communicating vessel received the left internal thoracic vein (blue arrow) and the inferior thyroid veins (white arrow). Right brachiocephalic vein (RBCV) and the thin elongated medial division of the left brachiocephalic vein (2) joined to form the right superior vena cava (1). The left SVC descended vertically to the left of the arch of aorta (3) and the pulmonary trunk (PT). Then it further descended to run behind the left ventricle, but

anterior to the pulmonary veins. Finally, it pierced the pericardium to reach the posterior atrioventricular groove and continued as the coronary sinus. Both the RSVC and the LSVC were equal in calibre. The coronary sinus was enlarged with its diameter equal to that of LSVC entering it. It opened into the right atrium at its usual site. The size of the opening was equal to the diameter of the coronary sinus. It received the middle cardiac vein, posterior vein of the left ventricle, and 2-3 veins accompanying the diagonal artery.

The right azygos vein (AV) entered the RSVC after arching over the root of the right lung (Fig. 2). It received the right superior intercostal vein (white arrow) before joining the RSVC. A left azygos vein (LAV) replaced the accessory azygos and the hemiazygos veins. The left azygos vein arched over the root of the left lung to drain into the LSVC. It received the left superior intercostal vein (yellow arrow) before draining into the LSVC. The azygos and the left azygos veins did not communicate with each other.

The following arterial variations were also observed in the same cadaver. The *thyroidea ima* artery (red arrow in Fig. 1) arose from the left side of the brachiocephalic trunk to supply

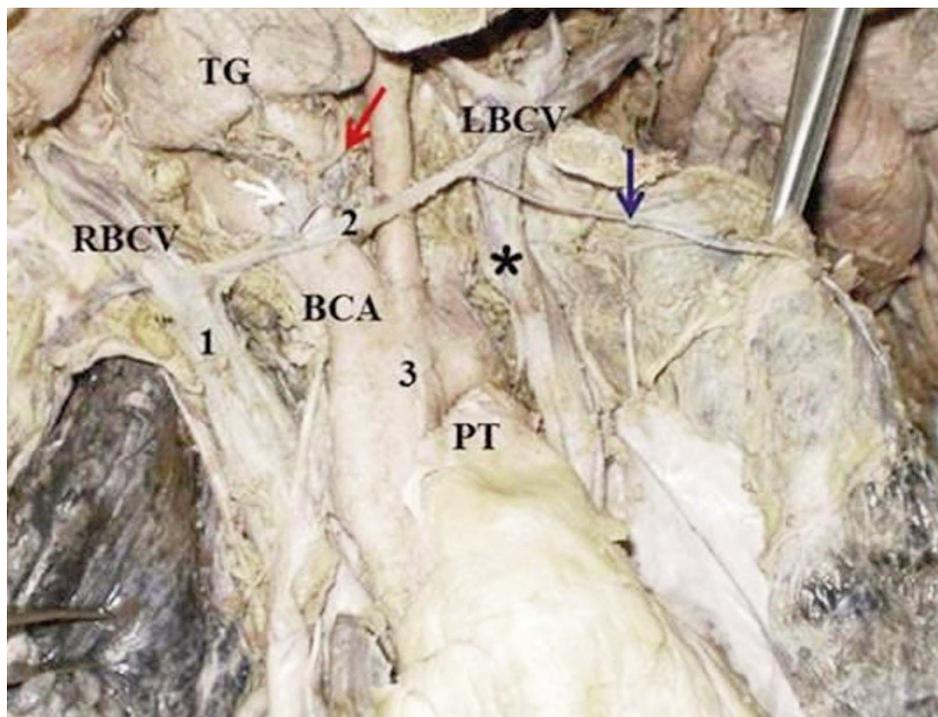


Fig. 1. Dissected thoracic cavity showing double venae cavae. TG – thyroid gland; LBCV – left brachiocephalic vein; RBCV – right brachiocephalic vein; BCA – brachiocephalic trunk; PT – pulmonary trunk; 1 – right superior vena cava; 2 – thin, elongated medial division of LBCV; 3 – arch of aorta; White arrow – inferior thyroid vein; Red arrow – *thyroidea ima* artery; Blue arrow – internal thoracic vein; \* - left superior vena cava.

the thyroid gland. Examination of the coronary system (Fig. 3) revealed that the left circumflex artery (LCX) did not continue into the posterior atrioventricular groove and was incomplete. The right coronary artery (RCA) was unusually thicker than the left coronary artery (LCA). The RCA had a normal course and crossed the crux of the heart but did not anastomose with LCX as the LCX stopped short of the posterior atrioventricular groove (Fig. 3). In addition to the marginal artery and the posterior interventricular artery, the RCA also gave three ventricular branches (1 & 2) to the left ventricle after it crossed the crux. The left coronary artery (LCA) originated from the left posterior sinus and immediately bifurcated into anterior interventricular artery and the LCX. A little beyond the left margin of the heart the LCX continued as the diagonal artery (DA). The anterior interventricular artery provided three ventricular branches (4) and the incomplete circumflex artery gave two ventricular branches to the left ventricle. A part of the posterior atrioventricular groove was devoid of any artery (yellow arrow) and that part of the left ventricle was appearing to be supplied by those additional ventricular branches from both anterior interventricular and incomplete LCX. The LCA also gave a

*vasa vasorum* to the left superior vena cava (white arrow).

## DISCUSSION

Persistent LSVC is asymptomatic and compatible with life unless the LSVC opens into the left atrium or is accompanied by other congenital heart defects like ASD, VSD, and tetralogy of Fallot (Cormier et al., 1989). The presence of LSVC assumes great importance during CT scan evaluation of the mediastinum, because LSVC can be mistaken for a lymph node and result in false staging of lung cancer (Tzilias et al., 2010). Procedures like transvenous pacemaker electrode placement, central venous catheterisation, pulmonary artery catheterisation, cardiopulmonary bypass and left sided thoracotomy can become complicated or even life-threatening with the persistent LSVC (Edwin et al., 2009). In the present case the persistent LSVC is accompanied by dilated coronary sinus and recent reports state that enlarged coronary sinus may result in obstruction to the ventricular outflow tract since this enlarged coronary sinus lies superior to the mitral valve partially obstructing it (Dibardino et al., 2004).

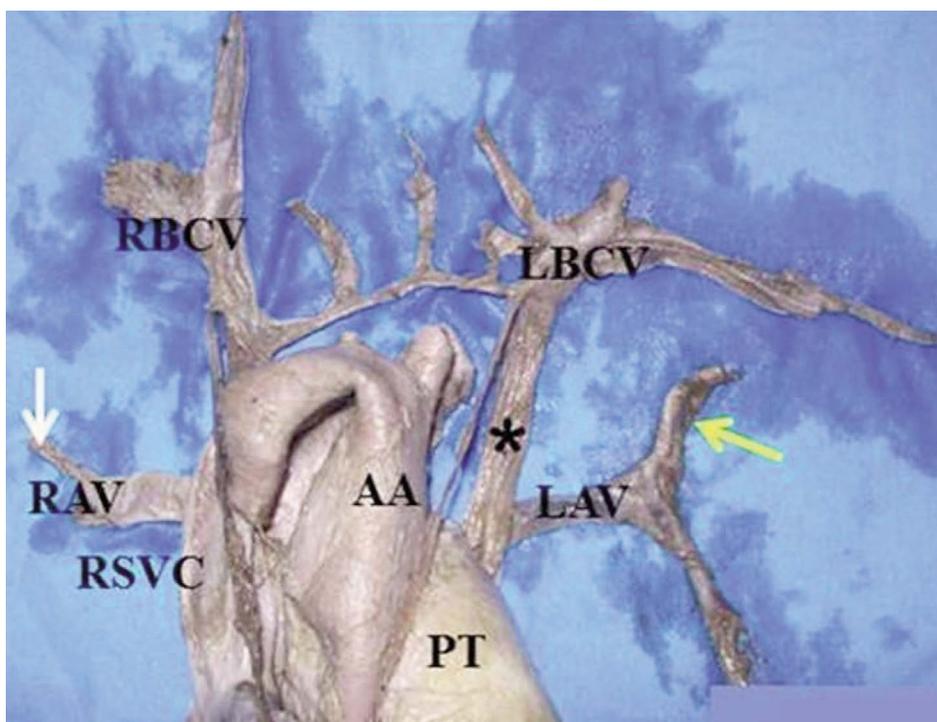


Fig. 2. Dissected venous channels showing double azygos veins. LBCV – left brachiocephalic vein; RBCV – right brachiocephalic vein; AA – arch of aorta; PT – pulmonary trunk; RAV –right azygos vein; LAV – left azygos vein; RSVC – right superior vena cava; White arrow – right superior intercostal vein; Yellow arrow – left superior intercostal vein; \* - left superior vena cava

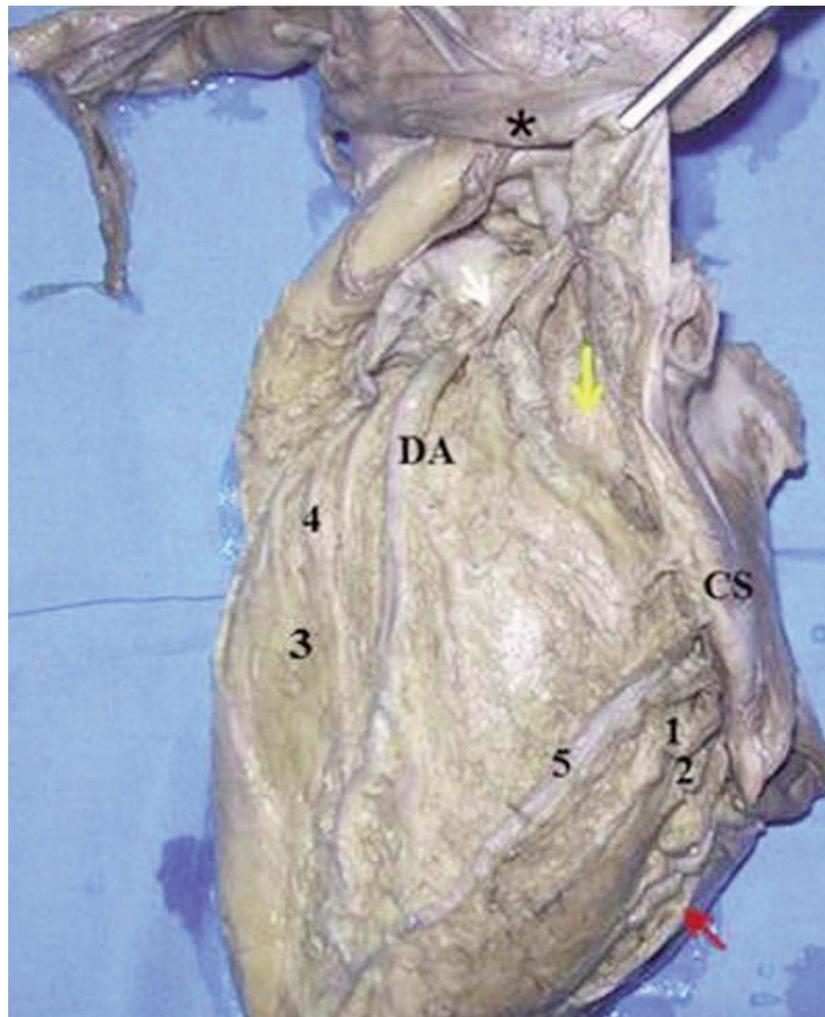


Fig. 3. Left lateral surface of heart showing incomplete circumflex artery. CS – coronary sinus; DA – diagonal artery; 1 & 2 – ventricular branches of posterior interventricular artery; 5 – posterior vein of left ventricle; Red arrow – posterior interventricular artery; Yellow arrow – atrioventricular groove without artery; White arrow – *vasa vasorum* for left SVC; \* - left SVC.

Double superior vena cavae with bilaterally symmetrical azygos veins have been reported earlier (Nandy and Blair, 1965) which is similar to the present case. The left azygos vein appears to be developed from the persistent proximal portion of the left posterior cardinal vein and the left supra cardinal vein, as suggested by Nandy and Blair (1965).

The persistence of LSVC along with coronary artery anomaly appears to be a rare variation, if we rely on the existent literature. The prevalence of absent LCX has been shown to be 0.003% (Hashimoto et al., 2004). The absence of LCX is usually compensated with the large super dominant right coronary artery, crossing the crux and perfusing the LCX territories (Kardos et al., 1997). The present case agrees with the abovementioned reports, since the LCX is incomplete and is compensated by unusually thick right coronary artery which crosses the crux and pro-

vides additional branches to the left ventricle through its posterior interventricular branch.

In conclusion, since the existence of PLSVC is an accidental finding during cardiothoracic surgeries or echocardiograms, prior knowledge of such an anomaly in the heart should be considered seriously by the surgeons or interventional radiologists. The presence of associated coronary artery anomaly may make it more vulnerable to complications during the surgical procedures. Therefore the present report is an attempt to alert the physicians or surgeons on the possible existence of such a variation, in order to minimise diagnostic errors.

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