

## Double superior vena cava and anomaly of cardiovascular system with a review of the literature

By

Akira IIMURA<sup>1</sup>, Takeshi OGUCHI<sup>1</sup>, Masakazu SHIBATA<sup>2</sup>, Masato MATSUO<sup>1</sup>,  
Tsuneo TAKAHASHI<sup>1</sup>

<sup>1</sup>Department of Anatomy, Kanagawa Dental College 82 Inaoka, Yokosuka, Kanagawa, 238-8580, Japan

<sup>2</sup>Kanagawa University of Human Services, Faculty of Health & Social Work, 1-10-1 Heiseicho,  
Yokosuka, Kanagawa, 238-8522, Japan

– Received for Publication, February 28, 2011 –

**Key Words:** double superior vena cava, persistent left superior vena cava, interatrial shunt, cardiac anomaly

**Summary:** In a student course of gross anatomy dissection at Kanagawa Dental College in 2008, we found an extremely rare case of the double superior vena cava that has a shunt between the right and left atria of a 81-year-old Japanese male cadaver. The left superior vena cava passed through the space between the left cardiac auricle and the left pulmonary vein and entered the coronary sulcus. Then it opened near the opening of the inferior vena cava as the coronary venous sinus to the right atrium. The upper edge of the interatrial septum was located at the site where the right superior vena cava opened to the right atrium. Accordingly, the right atrium connected with left atrium through this site. We discuss the anatomy and etiology of these anomalous structures with a brief review of the literature.

### Key to Abbreviation

ACV: anterior cardinal vein  
CCV: common cardinal vein  
CS: coronary sinus  
IVC: inferior vena cava  
LA: left atrium  
LAZV: left azygos vein  
LBV: left brachiocephalic vein  
LHSV: left horn of sinus venous  
LPV: left pulmonary vein  
LSVC: left superior vena cava  
LV: left ventricle  
OCS: orifice to coronary sinus  
OIVC: orifice of inferior vena cava  
OLPA: orifice of left pulmonary artery  
OLSVC: orifice of left superior vena cava  
ORA: orifice to right atrium  
ORSVC: orifice of right superior vena cava  
OV: oblique vein  
PA: pulmonary artery  
PCV: posterior cardinal vein  
PIV: posterior intercostals vein

RA: right atrium  
RAZV: right azygos vein  
RBV: right brachiocephalic vein  
RoAZV: root of azygos vein  
RPV: right pulmonary vein  
RSVC: right superior vena cava  
SVC: superior vena cava  
UEAS: upper edge of atrial septum

### Introduction

There have been no small number of morphological reports discussing the double superior vena cava<sup>1–19</sup>. The development of procedures for cardiovascular catheterization and clinical imaging examinations involving computed tomography (CT) and magnetic resonance imaging (MRI) has increased the number of these report<sup>20–24</sup>.

However, there have been only the limited number of reports which have discussed the double superior vena cava by dissecting cadavers. The reason is that the double superior vena cava often involves other cardiovascular anomalies, allowing on the limited number of adult

patients to survive. In the course of a routine anatomical dissection, we encountered the double superior vena cava that had a shunt between the right and left atria. Based on our observation findings of this anomaly, we discuss this anomaly from anatomical and embryological aspects in the present study.

## Subject and Methods

This article describes the dissection of a cadaver of an 81-year-old Japanese male in the laboratory of anatomy of Kanagawa Dental College during a routine educational dissection in 2008. The cause of the death was senility. The length and the diameter of the vessel were measured with calipers. Gross dissection was performed according to the standard procedure, and the morphology of the superior vena cava was observed. We found the superior vena cava on the right and left sides of the heart respectively.

## Results

### 1. Left superior vena cava

The left internal jugular, left external jugular and left subclavian veins joined to form the left superior vena cava in front of the left anterior scalene muscle. The left superior vena cava descended vertically in front of the arch of the aorta. The left superior vena cava passed through the space between the left cardiac auricle and the left pulmonary vein and entered the coronary sulcus. Then it opened near the opening of the inferior vena cava as the coronary venous sinus to the right atrium.

The left azygos vein connected with the left superior vena cava. And the connection point was inferiorly located 55.0 mm away from the origin of the left superior vena cava. On the right side 20.0 mm from the junction of the left superior vena cava and the coronary venous sinus, the width of the coronary venous sinus was 29.0 mm. On the right side 60.0 mm from the junction, the width of the coronary sinus was 35.0 mm. The coronary sinus ultimately opened to the right atrium in the area adjacent to the left side of the opening of the inferior vena cava (Fig. 1, 2 and 3).

### 2. Right superior vena cava and interatrial septum

The same as on the left side, the right internal jugular, right external jugular and right subclavian veins joined to form the right superior vena cava in front of the right anterior scalene muscle. It descended vertically and opened to the right atrium. At 57.3 mm from the origin of the right superior vena cava, the right azygos vein connected with the right superior vena cava. The right superior pulmonary vein opened to the junction of the right superior vena cava and the right atrium. The upper edge of the interatrial sep-

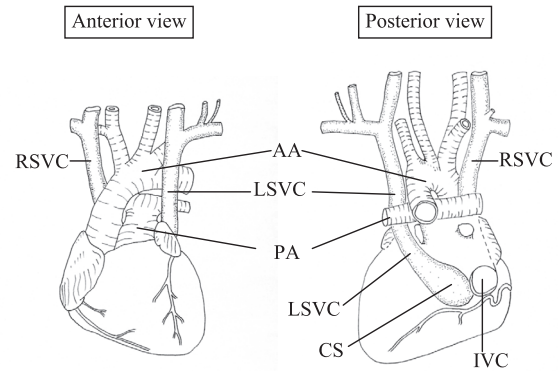


Fig. 1. Diagrams of the double superior vena cava in the present case. Anterior view

The right superior vena cava is formed as usual by the joint of the right internal jugular and the subclavian veins, and passes downward to reach the right atrium. The left superior vena cava is formed by the joint of the left internal jugular and the subclavian veins, descends, crosses the part in front of the aortic arch, and reaches the posterior region of the heart through the space between the left auricle and the left upper pulmonary vein. Then it obliquely flows on the surface of the left atrium into the coronary sinus.

#### Posterior view

The left superior vena cava flows into the coronary sinus. The coronary sinus opens to the right atrium. The coronary sinus is dilated in the shape of a wide spindle.

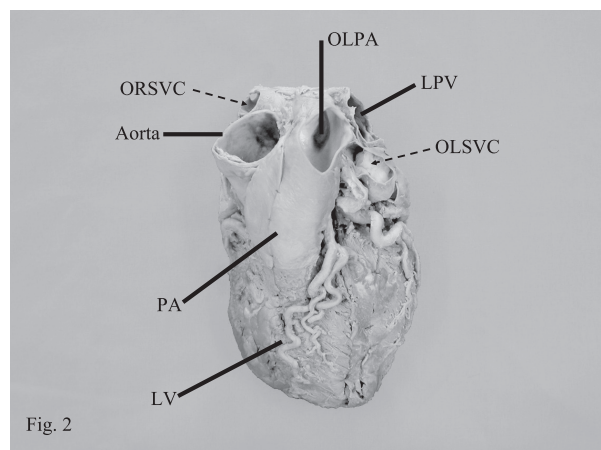


Fig. 2. A left lateral view of the heart.

The left side of the heart shows the orifice of the coronary sinus that receives the left superior vane cava.

tum was located at the site where the right superior vena cava opened to the right atrium. Accordingly, the right atrium connected with the left atrium through the site. There was no communicating branch between the right and left superior vena cava (Fig. 4).

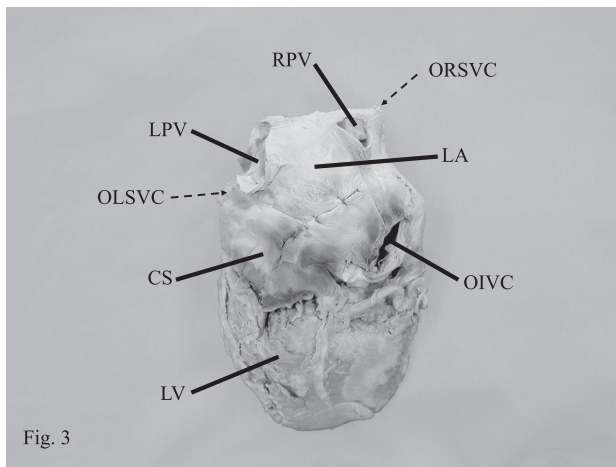


Fig. 3. Posterior view of the heart.

The coronary sinus lead to the left superior vena cava. It is dilated in the shape of a wide spindle (ca. 35 mm in width, ca 80 mm in length) The coronary sinus ultimately opens to the right atrium.

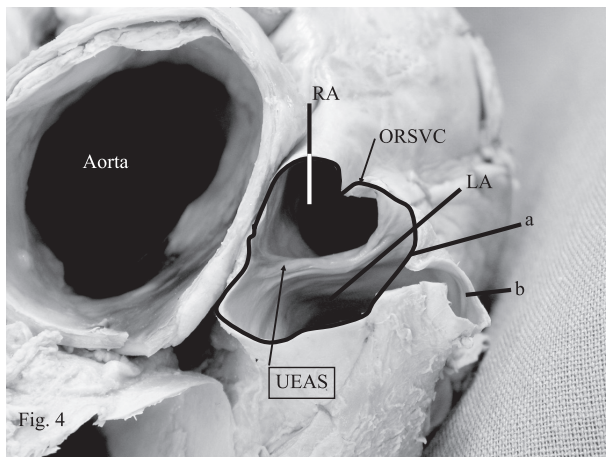


Fig. 4. The orifice of the right superior vena cava of the right atrium.

The large part (a) of the orifice was communicated with the right superior vena cava. The small part (b) was communicated with the right upper pulmonary vein.

The upper edge of the interatrial septum is located in the center of the large part (a). Accordingly, there is a shunt between the right and left atria.

#### 4. The azygos veins

The azygos venous system showed the symmetries and the right and left azygos vein opened to the superior vena cava on each side (at a height of the Th3). However, the left azygos vein (ca. 4 mm in width) was thinner than the right one (ca. 6.4 mm). There were two anastomotic veins (at heights of the Th7 and Th9, respectively) between the

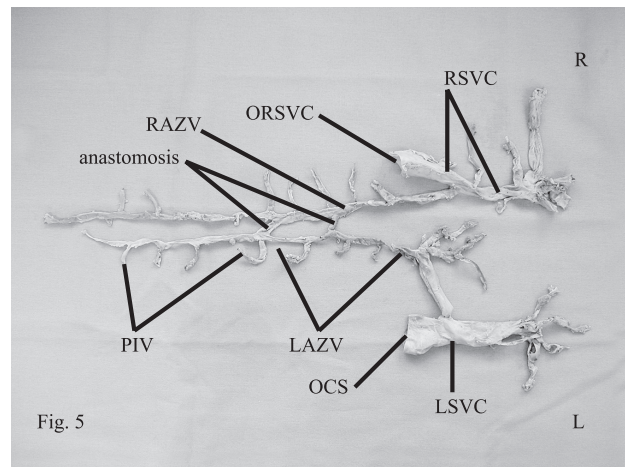


Fig. 5. The superior vena cava and the azygos veins.

Each superior vena cava received the azygos vein, and ultimately reached the heart. The right azygos vein had communicated with the left azygos vein at heights of the Th7 and Th9.

right and the left azygos veins. On the left side, the second and third posterior intercostals veins were drained by a common stem, then flew into the upper end of the azygos vein (Fig. 5).

## Discussion

No conclusion has been drawn from evaluation regarding the true incidence of the double superior vena cava (B Singh 2005)<sup>16</sup>. The incidence of the double superior vena cava has been documented by many authors as follows: 0.5% by Adachi (1933)<sup>1</sup>, 0.16% by Bergman RA (1988)<sup>12</sup>, and 0.3% by M. Uemura (2009)<sup>19</sup>.

A persistent left superior vena cava is the most common anomaly of abnormal veins flowing into the heart, with an incidence of 0.3–0.5% in healthy individuals, and 2–4% in patients with congenital heart diseases (B. Singh 2005<sup>16</sup>, H. Chen 2006<sup>18</sup>, J. Peltier 2006<sup>17</sup>).

Furthermore, AP. Gesase (2008)<sup>25</sup> have shown the high incidence (10–11%) of the double superior vena cava in congenital heart disease patients.

## Embryology and Classification

In the process of formation of the normal superior vena cava the right and left anterior cardinal veins and the posterior cardinal veins open to the venous sinus via the common cardinal veins at 4 gestational weeks. An anastomotic branch communicating the right and left anterior cardinal veins appears at 7 gestational weeks. The left anterior cardinal vein is ultimately replaced by this anastomotic

branch, which leads to the left brachiocephalic vein and joins the right brachiocephalic vein derived from the right anterior cardinal vein to form the superior vena cava.

In this process the left anterior cardinal vein and the left common cardinal vein degenerate and remain as the left atrial oblique vein. During the time of this phenomenon the left posterior cardinal vein also almost degenerates and disappears. On the other hand, the left supracardinal vein shows the increased connection with the right supracardinal vein along with degeneration of the left common cardinal vein and forms a hemiazygos vein, while the right supracardinal vein of the supracardinal veins, which appear at 7 gestational weeks, develops to become an azygos vein. Thus, the normal morphology in an adult body is formed by the rightward predominant development of the paired gestational venous system (Moor, K.L.1988)<sup>26</sup>.

There was no communicating branch between the right and left superior vena cava in the present case, probably because no anastomotic branch was formed between the anterior cardinal veins for some reason or an anastomotic branch, which has been once formed, did not develop. Therefore, the left common cardinal vein, which should essentially become the left atrial oblique vein, developed to form the dilated coronary sinus.

According to bibliographies and references in the literature regarding the azygos venous system, the system develops from a pair of the right and left supracardinal veins in the early stage of development. The veins communicate with the posterior cardinal veins to form the primordium of a pair of right and left azygos veins. Subsequently, only the anastomosed site between the common cardinal vein of the posterior cardinal vein and the supracardinal vein remains on the right side, and the root of the azygos vein is formed. The distal part of the posterior cardinal vein degenerates and disappears.

On the left side of normal cases an anastomosis between the anterior cardinal veins leads to disappearance of the anterior cardinal vein beneath the anastomosed site and the common cardinal vein. The anastomosed site of the anterior cardinal veins frequently receives only the limited number of the superior intercostal blood vessels. Consequently, the left supracardinal vein comes to flow into the right supracardinal vein through the transverse anastomosis to the right supracardinal vein, leading to formation of a hemiazygos vein.

However, these bibliographies have shown that communication of each of the right and left supracardinal veins with the respective common cardinal veins via the part of the posterior cardinal vein is maintained and azygos veins are formed on the right and left sides respectively, when the left superior vena cava remains. With regard to the veins corresponding to azygos veins in the present case, transverse anastomoses were observed at two sites. They independently opened to the right and left superior vena cava, indicating that they are regarded as paired azygos veins.

Posterior view of the developing heart (citation from Moor)

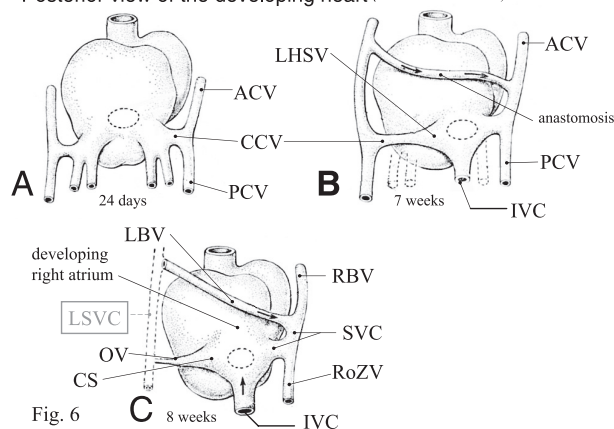


Fig. 6. Diagrams of embryology of the superior vena cava. (Posterior view)

During the 8th gestational week the right anterior cardinal vein is obliquely anastomosed with the left anterior cardinal vein. Normally, the right anterior cardinal vein and the right common cardinal vein form the superior vena cava. In the double superior vena cava the anastomosis, which usually forms the left brachiocephalic vein, is small or absent. The residual left superior vena cava, which is derived from the left anterior cardinal and the common cardinal veins, opens to the right atrium via the coronary sinus.

It has been believed that the persistent left superior vena cava is a factor for formation of the paired azygos veins in the double superior vena cava, whereas paired azygos vein is not observed in all the cases of the double superior vena cava. From these observations, it cannot be said that the feature of right-left predominance of the cardinal venous system does not have direct influence on the supracardinal veins. Further studies are needed in this regard in the future (Fig. 5, 6).

McCotter (1916)<sup>27</sup>, Donadio (1925)<sup>28</sup>, Nandy and Blair (1965)<sup>9</sup>, and Takenoshita (1986)<sup>11</sup> have reported classification of cases of the persistent left superior vena cava. Takenoshita has classified the cases on the basis of the presence/absence of an anastomosis between the right and left superior vena cava and direction of the inclination. On the other hand, Nandy and Blair<sup>9</sup> have classified the cases according to the presence/absence of pairing of azygos veins, as well as the presence/absence of anastomotic branch, into the following 4 types: I) The pattern with an anastomotic branch between the right and left superior vena cava; II) the pattern without any anastomotic branch; III) the pattern showing the presence of the left superior vena cava alone and degeneration and disappearance of the right superior vena cava; and IV) the pattern of the double superior vena cava with paired azygos veins. The present case is considered to correspond to Type IV, which was associated with paired azygos veins.



## The Presence/Absence of a Shunt between the Left and Right Atrial Systems and Its Classification

From a clinical viewpoint, any anastomosis between systemic vein has no functional problem, but some reports have shown that one of the right and left superior vena cava opens to the left atrium and leads to formation of a shunt between the right and left atrial systems. On this occasion, oxygenation of the arterial blood becomes an issue. Many of these reports have been based on CT, MRI, and ultrasonic scanning findings in clinical cases, rather than the anatomical assessment findings. It has been indicated that differences in the formation and degree of a shunt have clinically different meanings. When patterns of the anomaly are classified according to the presence/absence of a shunt between the right and left atrial systems on the basis of these previous reports, they may be classified into the following 5 types:

- (1) The type without a shunt in the outside and the inside of the heart; in the double superior vena cava both right and left superior vena cava open to the right atrium, and there is no shunt between the right and left veins in the inside of the heart<sup>7,29,30,17,25</sup>.
- (2) The type with a shunt in the inside of the heart; in the double superior vena cava both right and left superior vena cava open to the right atrium, and there is a shunt between the right and left veins in the inside of the heart.
- (3) The type with a shunt in the communication with an external blood vessel in the absence of a shunt in the inside of the heart; in the double superior vena cava the right superior vena cava opens to the left atrium, but there is no shunt in the inside of the heart<sup>20,31-33</sup>.
- (4) The type with a shunt in the communication with an external blood vessel in the absence of a shunt in the inside of the heart; in the double superior vena cava the left superior vena cava opens to the left atrium, but there is no shunt in the inside of the heart<sup>32,34-37</sup>.
- (5) The type with a defect of the right superior vena cava; only the left superior vena cava is present and communicates with the right atrium, but the right superior vena cava is partially lost<sup>15,19,30,38</sup>.

Of these 5 types, Types (1), (3), (4), and (5) have respectively been reported, but there have been no reports on Type (2).

In the present case, the right atrium connected with the left atrium because of the partial defect of the upper edge of the interatrial septum at the site where the right superior vena cava opened to the heart, and the case may be classified into Type (2) because of the presence of a shunt in the inside of the heart. There have been no reports on this type. This case is consistent with the previous reports, which have shown that the incidence of coexistence of a shunt between the right atrial system and the left atrial system with the double superior vena cava is high. Clinical problems including hypoxia and brain abscess have been offered to cases of a right-left shunt (Edgar CS. 1985)<sup>39</sup>,

while in the present case it is estimated that there was no marked clinical problem with the function due to a shunt between the right atrial system and the left atrial system, when the case age (81 years) at the time of death is considered.

## The Defect of the Upper Edge of the Interatrial Septum

Moore (1988)<sup>40</sup> has classified patterns of the defect of the upper edge of the interatrial septum into 6 types; when the venous sinus is abnormally adsorbed by the right atrium and when the secondary septum does not yet develop or abnormally develops, high-level septum defect may be present in the upper part of the normal oval foramen in some cases. According to this author, this type of case is routinely associated with partial abnormal connection with the pulmonary vein, and is one of the uncommon patterns of interatrial septum defect. The present case is similar to this type in the point of that the defect was present at a high level of the septum, and is consistent with the indication by Moore in the point of that the right superior pulmonary vein connected with the right superior vena cava. In the venous system usually showing the rightward-predominant development, in the formation of the systemic venous system suggests that it becomes a factor for formation of the double superior vena cava and formation of this type of septum defect.

## Clinical Significance

Recent reports have shown clinical problems with the double superior vena cava, as follows. B. Singh (2005)<sup>16</sup> has described that the dilated coronary venous sinus makes contrast radiographic findings of the interatrial septum and the space between the ventricles obscure on cardiac angiography. The author has added that possibilities of other congenital anomalies in the heart and an associated shunt between the right cardiac system and the left cardiac system occurring should be considered from the presence of the persistent left superior vena cava.

J. Peltier (2006)<sup>17</sup> has described that the presence/absence of an anastomosis between the right and left superior vena cava and a defect of the right superior vena cava also have influence on catheterization and results of examinations and that it is important to recognize the presence of the anastomosis and the defect.

S.K. Goyal (2008)<sup>22</sup> has shown that the presence of the persistent left superior vena cava makes an approach from the left side to the right atrium with a catheter difficult. The author has added that the persistent left superior vena cava is present when a catheter or a guide-wire passes the course on the left side of the superior mediastinum, not the usual position, through the left subclavian vein.

As these authors have indicated, we consider it desirable to comprehend the basic morphology of the vascular

system by non-invasive methods including CT, MRI, ultrasonic scanner, etc. before implementation of invasive diagnostic techniques such as catheterization. When the persistent left superior vena cava is ascertained by these diagnostic techniques, the presence/absence of other congenital anomalies should be confirmed, and comprehension of risks related to catheterization and selection of a safe route should be considered.

## References

- Adachi B. Anatomie der Yapaner. II Das Venensystem der Japaner. I Lieferung. 1933; 68–76.
- Ymada S. Über zwei Fälle von Verdoppelung der Vena cava superior Acta Anat. Nippon 1934; **7**:82–87. (in Japanese)
- Otsuka N. Ein Fall mit Doppelter Vena cava superior bei einer Japanischen Frau. Acta Anat Nippon 1961; **36**:298–304. (in Japanese)
- Abe K. An abnormal case of the coronary sinus with no opening into the right atrium (A case of the so-called left superior vena cava). Acta Anat Nippon 1964; **32**:249–251. (in Japanese)
- Kalidas Nandy, Charles B. Blair. Double superior venae cavae with completely paired azygos veins. Anat Rec 1965; **151**:1–10.
- Yamadori T, Takashima S. A case of the double superior vena cava. Acta Anat Nippon 1966; **41**:213–221. (in Japanese)
- Fujimoto Y, Okuda H and M. Yamamoto. A case of the bilateral superior vena cava with some other anomalous veins. Okajima Fol. Anat. Jap 1972; **48**:413–426.
- Ohkubo M, Yoshida Y. A case of bilateral superior caval veins. Acta Anat Nippon 1976; **51**:251–256. (in Japanese)
- Yano M, Sato T. A case of the double inferior vena cava with the persistent left superior vena cava. Acta Anat Nippon 1980; **55**:224–240. (in Japanese)
- Masuko S., Inoue K. A case of the double superior vena cava and anomalous left hepatic vein opening directly into the right atrium. Acta Anat Nippon 1982; **57**:169–174. (in Japanese)
- Takenoshita H. Persistent left superior vena cava with paired azygos. – Report of a case and attempt at new classification – Acta Anat Nippon 1986; **61**:669–682. (in Japanese)
- Bergman RA, Thompson SA, Afifi AK, Saadeh FA. Compendium of human anatomic variation. Urban & Schwarzenberg, Baltimore. 1988: 32–35, 88–89.
- Konishi M, Kikuchi M. A case of persistent left superior vena cava with a Horseshoe kidney. Acta Anat Nippon 1991; **66**:524–536. (in Japanese)
- Sakamoto H, Akita K, Sato T. Left superior vena continuing to the accessory hemiazygos without anastomosis with the coronary sinus. Surg. Radiol Anat 1993; **15**:151–154.
- Sahinoglu K, Cassell MD, Miyauchi R, Bergman RA. Human persistent left superior vena cava with double coronary sinus. Ann Anat 1994; **176**:451–454.
- B. Singh, L. Ramsaroop, J. Maharaj, A. Reddi. Case of double superior vena cava. Clin Anat 2005; **18**:366–369.
- J. Peltier, C. Destrieux, J. Desme, C. Renard, A. Remond, S. Velut. The persistent left superior vena cava: anatomical study, pathogenesis and clinical considerations. Surg Radiol Anat 2006; **28**:206–210.
- H. Chen, S. Shoumura and S. Emura. Bilateral thoracic ducts with coexistent persistent left superior vena cava. Clin Anat 2006; **19**:350–353.
- Uemura M, Takemura A, Ehara D, Yasumitsu H, Ohnishi Y, Suwa F. Left superior vena cava with left azygos vein. Okajima Fol. Anat 2009; **86**(2):55–60.
- Pieter M Pretorius, Fergus V. Gleeson. Case 74: Right sided superior vena cava draining into left atrium in a patient with persistent left sided superior vena cava. Radiology 2004; **232**:730–734.
- N.K. Shyamkumar, R. Brown. Double superior vena cava with a persistent left superior vena cava: an incidental finding during peripherally inserted central catheter placement. The Authors J. compilation 2007; **51**:B257–B259.
- Sandeep K. Goyal, Sujeeth R. Punnam, Gita Verma, Frederick L. Ruberg. Persistent left superior vena cava: a case report and review of literature. Cardiovascular Ultrasound 2008; **131**:e78–e80.
- J.P. Kaski, J. Wolfenden, A. Magee. Obliteration of left superior caval vein draining to the atrium during spontaneous closure of ventricular defect. European J. of Echocardiography 2009; **10**:160–162.
- Demirkan B, Gungor O, Turkvatan A, Guray Y, Guray U. Images of persistent left superior vena cava draining directly into left atrium and secundum-type atrial septal defect. J. Cardiovasc Compt Tomogr 2010; **4**(1):70–72.
- Ainory P. Gesase, Flora M. Fabian, David N. Ngassapa. Double superior vena cava presenting with anomalous jugular and azygos venous system. Indin Heart 2008; **60**:352–358.
- Moor K.L. The Developing Human. 4th W.B. Saunders Co., 1988: 286–291.
- McCotter R.E. Three cases of persistence of the left superior vena cava. Anat Rec 1916; **10**:371–383.
- Danadio N. Ein Fall von Verdoppelung der Vena cava superior. Anat Anz 1925; **59**:321–327.
- C. Mori, H. Hashimoto, K. Hoshino. Two cases of double superior vena cava. Jpn. Heart J 1990; **31**(6):881–888.
- Gonzalez JC, Testa A, Vidan J, Izquierdo R, Garcia-Castelo A, Daniel C, Armesto V. Persistent left superior vena cava draining coronary sinus: 10 cases and literature view. Clin Cardiol 2004; **27**(9):515–518.
- Mattias Gorenflo, Christian Sebening, Herbert E. Ulmer. Anomalous connection of right superior caval vein to the morphologically left atrium. Cardiol Young 2006; **16**:184–186.
- Fabio Miraldi, Iacopo Carbone, Adriano Ascarelli, Antonio Barretta, Ilaria D. Angeli. Double superior vena cava: Right connected to left atrium and left to coronary sinus. International J. of Cardiology 2009; **131**:e78–e80.
- Charles Baggett, Shawn J. Skeen, D. Scott Gantt, Bradley R. Trotter, Krista L. Birkemeier. Isolated right superior vena cava drainage into the left atrium. Texas Heart Institute Journal 2009; **36**(6):611–614.
- Henry B. Wiles. Two cases of left superior vena cava draining directly to a left atrium with a normal coronary sinus. Br. Heart J 1991; **65**:158–160.
- Kato Y, Miyamoto S, Minamimura H, Ishikawa T, Ohue K, Shimizu Y. Persistent left superior vena cava draining to left atrium with normal coronary sinus. The Jap. Thoracic and Cardiovasc Surgery 2005; **53**(1):46–48.
- Dehghani P, Benson LN, Horlick EM. Transcatheter closure of persistent left sided superior vena cava draining into atrium importance of balloon test occlusion. J. Invasive Cardiol 2009; **21**(7):122–125.
- Ugaki S, Kasahara S, Fujii Y, Sano S. Anatomical repair of a persistent left superior vena cava into the left atrium. Interactive Cardiovascular and Thoracic Surgery 2010; **111**:199–201.
- Özgül Ucar, Lale Pasaoglu, Hülya Cicekcioğlu, Murat Vural, Ibarhim Kocaoglu, Sinan Aydogdu. L. Persistent left superior vena cava with absent right superior vena cava: a case report and review of the literature. Cardiovasc J. Afr 2010; **21**(3):164–166.
- Edgar C. Schick. Persistent left superior vena cava and right superior vena cava drainage into the left atrium without arterial hypoxemia. American College of Cardiology 1985; **5**(2):374–378.
- Moor K.L. The Developing Human. 4th W.B. Saunders Co., 1988: 308–312.