

Anomalies of the subclavian artery and superior vena cava : Aberrant right subclavian artery and double superior vena cavae.

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Anomalies of the great vessels were recorded from 450 cadavers during the period 1980 to 1992. Four cases (0.89%) of aberrant right subclavian artery (ARSCA) and one case (0.02 %) of double superior vena cavae (DSVC) were found. One of the ARSCA was consisted of a dilated sac at its origin. There was one case of ARBSCA in which the thoracic duct terminated at the junction of the right internal jugular vein and right subclavian vein instead of the left side. In the case of the left superior vena cava, it terminated into the coronary sinus.

Key words: *Aberrant right subclavian artery (ARSCA), Double superior vena cavae (DSVC).*

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ได้ทำการศึกษาจากศพในชั้นเรียน 450 ศพ ตั้งแต่ พ.ศ. 2523-2535 พบว่ามี *Aberrant right subclavian artery (ARSCA)* อยู่ 4 ราย (คิดเป็น 0.89 %) และ *Double superior vena cavae* 1 ราย (คิดเป็น 0.02 %) หนึ่งในสี่รายของ *ARSCA* มี 1 รายที่มีลักษณะโป่งพองตรงตำแหน่งที่เส้นเลือดนี้แยกออกมา และอีก 1 รายของ *ARSCA* มี *Thoracic duct* มาเปิดเข้าสู่ตรงบริเวณที่ *Right internal jugular vein* บรรจบกับ *right subclavian vein* แทนที่จะทะลุเข้าสู่ทางด้านซ้ายตามปกติ ส่วน *Left superior vena cava* นั้นทะลุเข้า *coronary sinus*.

Because many changes are involved in the transformation of the embryonic aortic arch system into the adult arterial pattern. It is understandable that the variations may occur. Most anomalies result from the persistent part of aortic arches that normally disappear or vice versa. Examples for anomalies of the arch of aorta are double aortic arch, i.e., right arch of the aorta, and abnormal origin of the right subclavian artery.⁽¹⁾ The aberrant right subclavian artery (ARSCA) is a well known vascular anomaly. It always forms a vascular ring and is rarely clinically significant because the ring is usually not tight enough to constrict the esophagus and the trachea. Some patients may be presented with recurrent dysphagia, hemoptysis, chest pain, dysphagia lusorum and a mediastinal mass.⁽²⁾ The possible complications of ARSCA is hematemesis which is due to esophageal and ARSCA fistula. Rupture of the aneurysm of ARSCA is also a common complication.⁽³⁻⁵⁾

The congenital anomalies of the systemic venous connection to the heart are rather wide and vary in physiological or pathological signs and symptoms. The malformations could be the left superior vena cava connected to the coronary sinus, interrupted inferior vena cava and absence of right superior vena cava. Left superior vena cava connected to the left atrium is due to incorporation of the coronary sinus into left atrial cavity. It resulted in a right to left shunt.⁽⁶⁾ The purpose of this study was to learn more about finding out how many percentages of the anomaly of the great vessels occur in Thai populations.

Materials and Methods

The hearts and great vessels from 450 cadavers were examined routinely in the labo-

ratory of gross anatomy in the Faculty of Medicine, Chulalongkorn University during the period 1980 - to 1992. The ascending aorta, arch of aorta and its branches were recorded. The pulmonary trunk, pulmonary artery, superior vena cava, inferior vena cava and all possible anomalies of the great vessels and the hearts were carefully observed.

Results

By routine dissection, we found four cases of aberrant right subclavian artery (ARSCA = 0.89%), (Figure 1B-D) and one case of double superior vena cavae (DSVC=0.02 %),(Figure 3). There were no anomaly of the heart associated with these findings.

In all cases of ARSCA, the arteries were the fourth branch of the aorta. They arised from the proximal part of the descending aorta (Figure 1B-D) and crossed the midline behind the esophagus to reach the right arm (Figure 4A). In of these cases, dilated sac at the origin of the artery called the Diverticulum of Kommerell was observed (Figure 4B). In another case, there was an abnormal termination of the thoracic duct drained to the junction between the right internal jugular vein and the right subclavian vein.

In the case of double superior vena cavae, the left superior vena cava drained into the coronary sinus which was dilated more than normal (Figure 3D). The left superior vena cava received the venous drainage from the left brachiocephalic vein. The right superior vena cava received venous drainage from the right brachiocephalic vein and drained directly into the right atrium (Figure 3C). There was no connection between these two superior vena cavae.

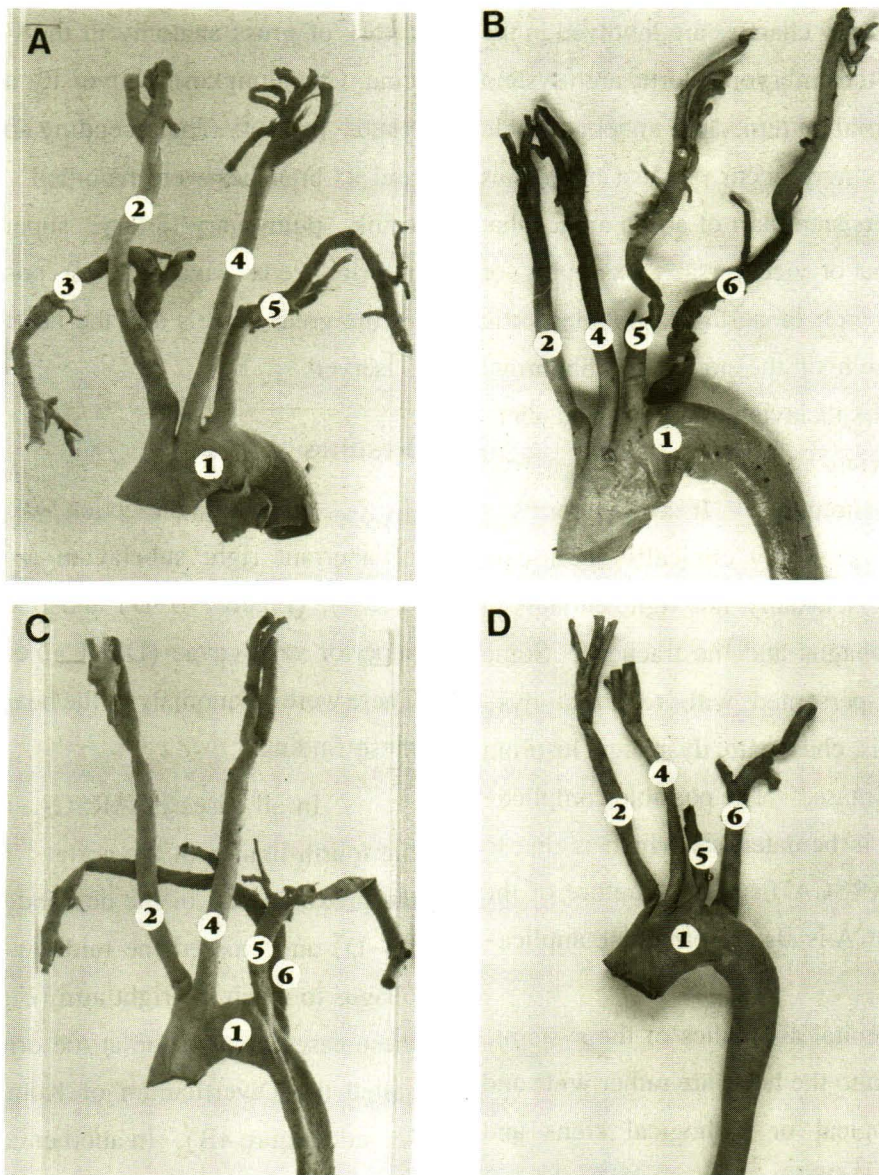


Figure 1. Arch of aorta and its branches.

A normal branches

B-D Aberrant right subclavian artery

1 = arch of aorta,

2 = right common carotid artery

3 = right subclavian artery

4 = left common carotid artery

5 = left subclavian artery

6 = aberrant right subclavian artery

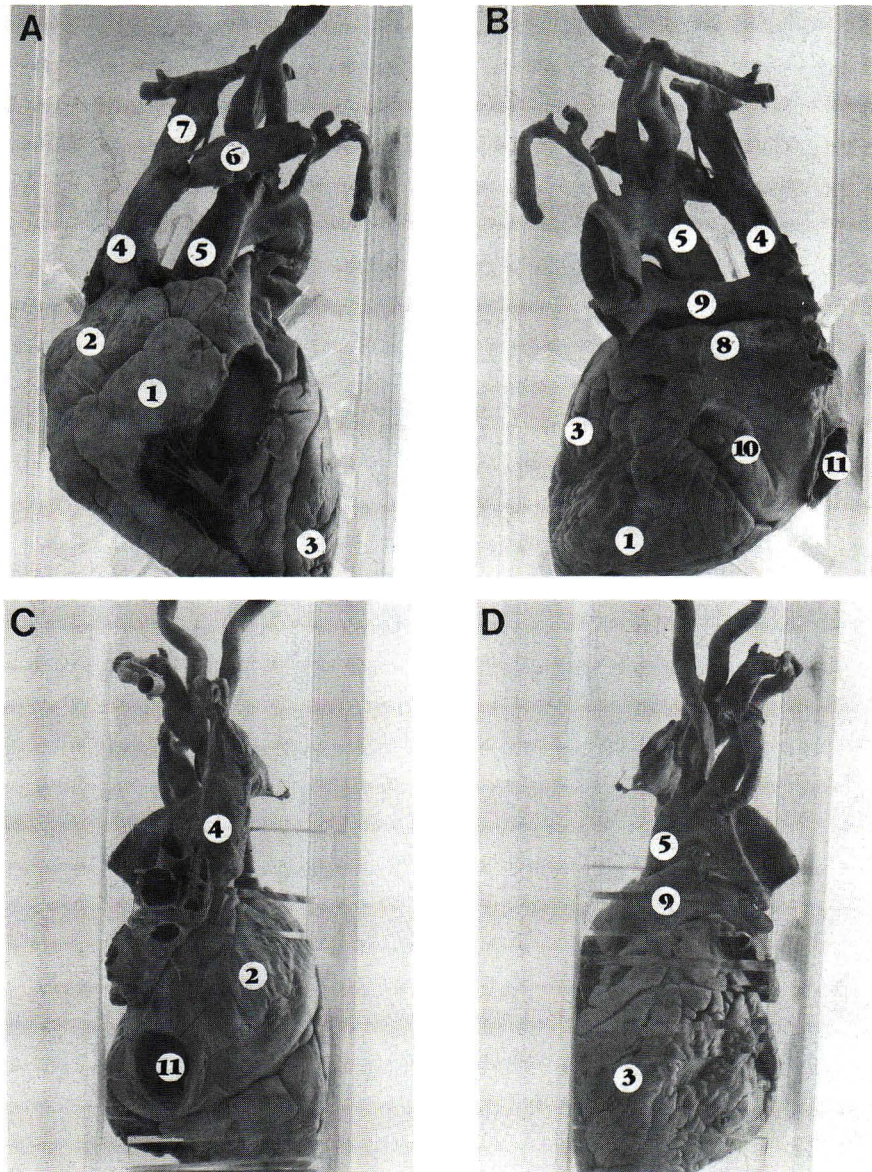


Figure 2. Normal heart and great vessels.

A Anterior view

B Posterior view

C Right lateral view

D Left lateral view

1 = right ventricle

2 = right atrium

3 = left ventricle

4 = right superior vena cava

5 = ascending aorta

6 = left brachiocephalic vein

7 = right brachiocephalic vein

8 = left atrium

9 = pulmonary trunk

10 = coronary sinus

11 = inferior vena cava

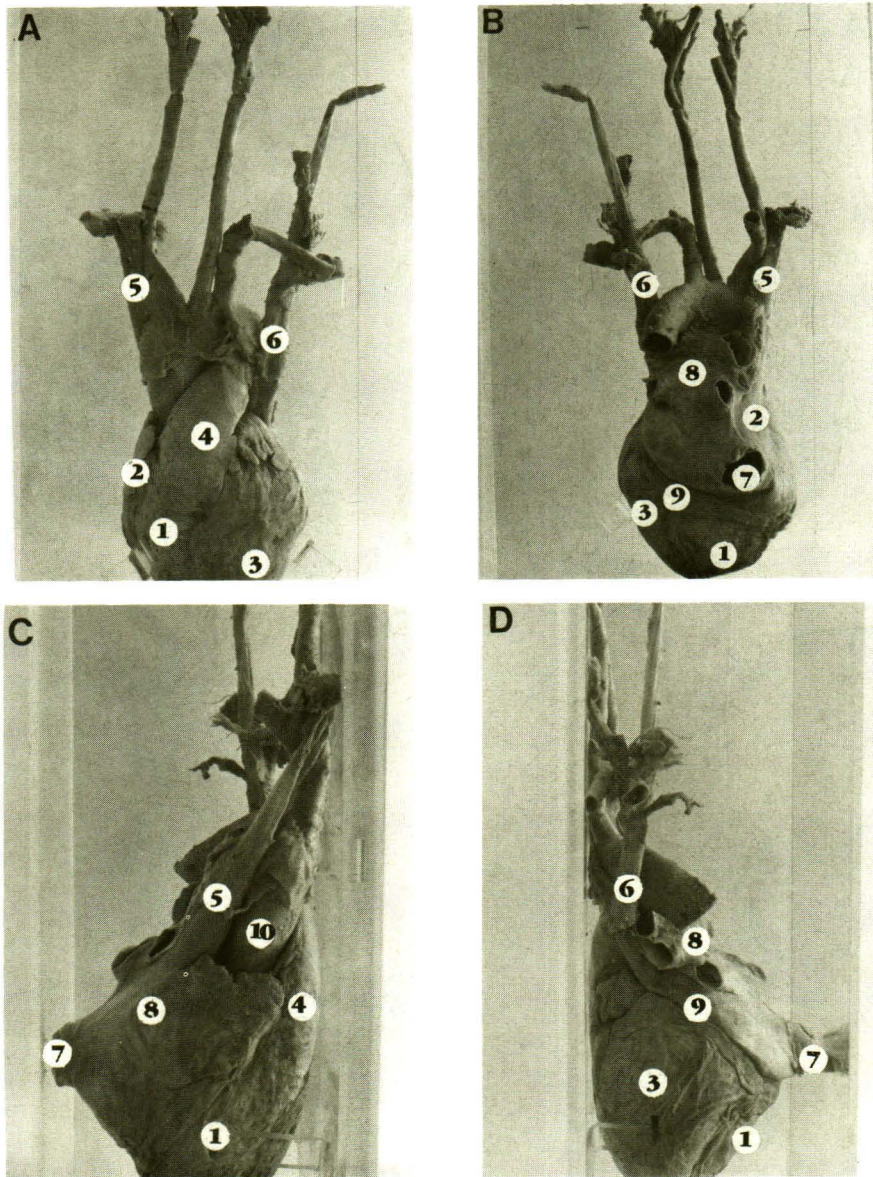


Figure 3. Double superior vena cavae.

A Anterior view

B Posterior view

C Right lateral view

D Left lateral view

1 = right ventricle

2 = right atrium

3 = left ventricle

4 = pulmonary trunk

5 = right superior vena cava

6 = left superior vena cava

7 = inferior vena cava

8 = left atrium

9 = coronary sinus

10 = ascending aorta

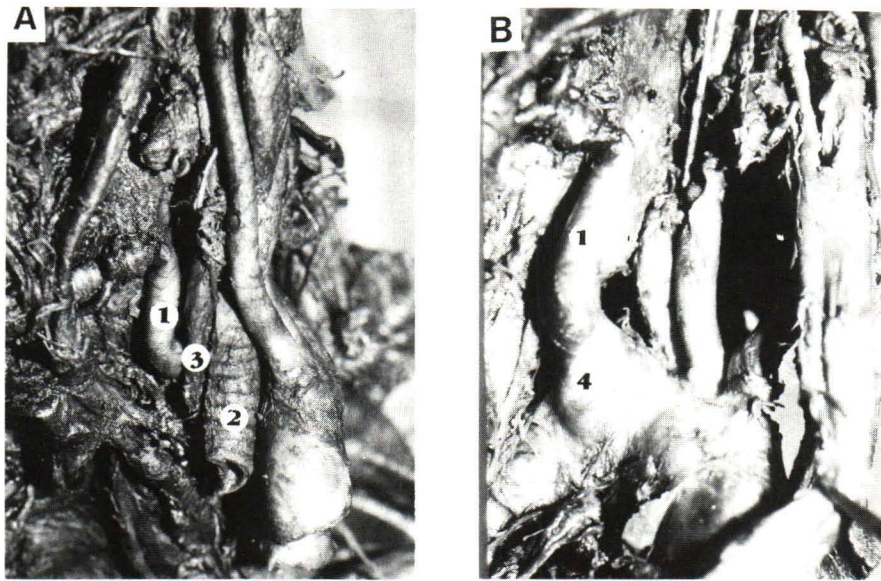


Figure 4. A The course of aberrant right subclavian artery (1) passing behind the trachea(2) and esophagus (3)
B Diverticulum of Komorell (4) at the origin of aberrant right subclavian artery (1)

Discussion

The aberrant right subclavian artery occurs when the right fourth aortic arch and the right dorsal aorta disappear cranial to the seventh intersegmental artery. As a result, the right subclavian artery forms from the right seventh intersegmental artery and the distal part of the right

dorsal aorta (Figure 5A). As development proceeds, differential growth shifts the origin of the right subclavian artery cranially until it comes to lie close to the origin of the left subclavian artery (Figure 5B). It passes posterior to the trachea and esophagus to supply the right upper limb. This anomaly is common and always forms a vascular ring (Figure 5C).

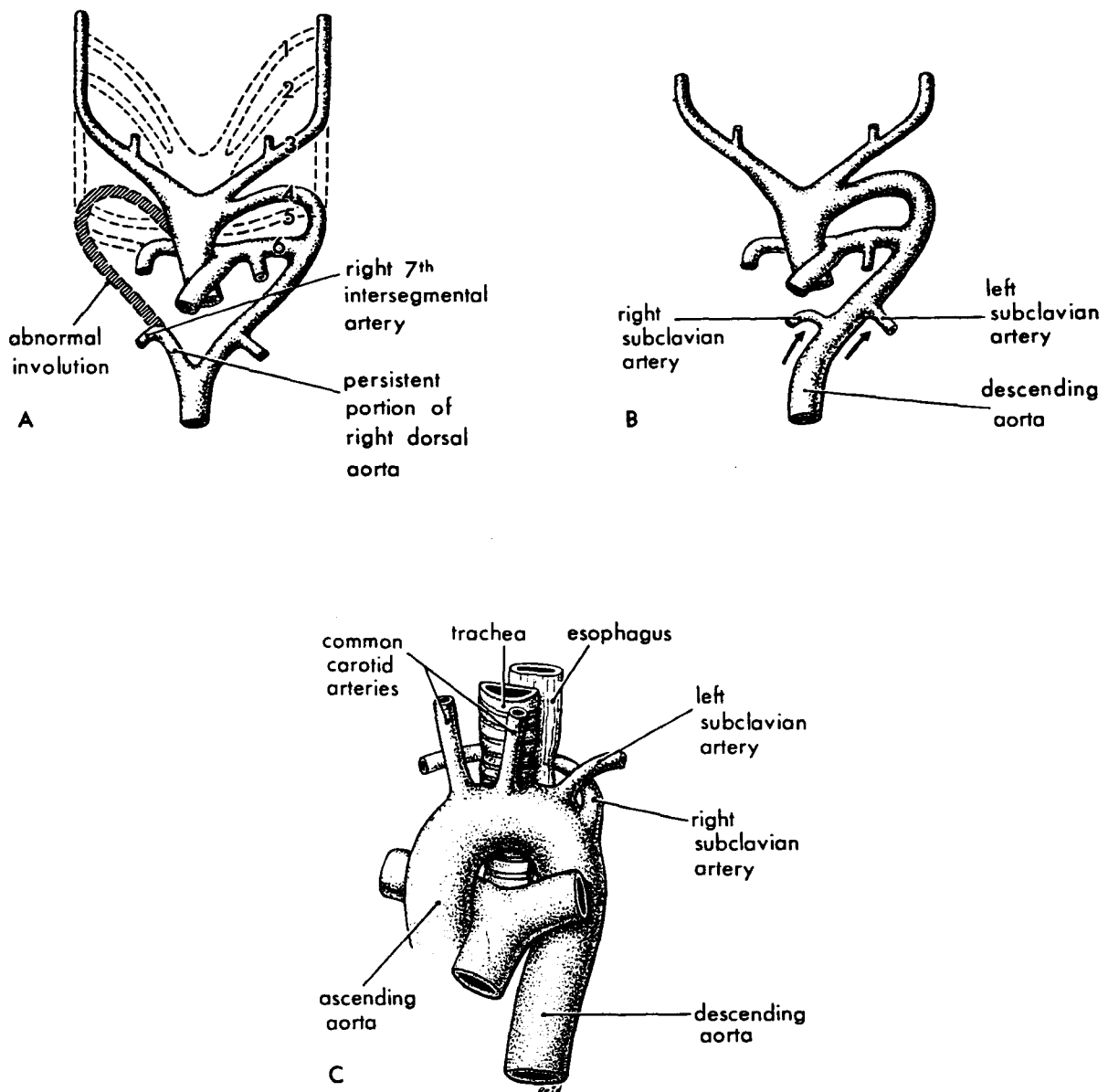


Figure 5. Sketches illustrating the probable embryological basis of abnormal origin of the right subclavian artery.

- A The right fourth aortic arch and the cranial portion of the right dorsal aorta have involuted. As a result, the right subclavian artery forms from the right seventh intersegmental artery and the distal segment of the right dorsal aorta.
- B As the arch of the aorta forms, the right subclavian artery is carried cranially (arrows) with the left subclavian artery.
- C The abnormal right subclavian artery arises from the aorta and passes posterior to the trachea and esophagus.

(From Moore and Persuud. The cardiovascular system. 5th Ed.)

ARSCA is said to occur in 0.4% to 3% of patients. The Diverticulum of Kommerell is a ramnant of the right dorsal aortic arch.⁽⁷⁾ In our study we found one out of four cases. In report

from 278 cases of routine dissection, there are two cases of retroesophageal right subclavian artery (0.27%).⁽⁸⁾ All cases in our study were retroesophageal right subclavian artery. (Table 1)

Table 1. Comparison in number of cases of ARSCA with other sources.

Sources	Total cases	Number of cases	%
Sere-sturm M.,et al., in 1985	278	2	0.72
Urai and Vilai (present report 1995)	450	4	0.89

The ARSCA could arise from the proximal portion of the descending thoracic aorta,⁽⁹⁾ or from the common carotid trunk in which carotid artery associated with ARSCA,⁽¹⁰⁾ or from the right

pulmonary via a right ductus arteriosus,⁽¹¹⁾ or from thoracic aneurysm at the fourth branch of the aortic arch.⁽⁴⁾ In our study, the ARSCA arises from the proximal portion of the descending aorta. (Table 2.)

Table 2. Comparison in the origins of ARSCA with Various sources.

Sources	Origin of ARSCA
Gross C., et al.,in 1990	Thoracic aneurysm (at the fourth branch of the aortic arch)
Stone W.M.,et al.,in 1990	Descending thoracic aorta
Hofbeck M.et al., in 1991	Pulmonary artery (Via a right dustus arteriosus)
Calleja F.,et al, in 1990	Common carotid trunk
Urai and Vilai (present report 1995)	Proximal portion of the Descending aorta

Double superior vena cavae are characterized by the persistence of the left anterior cardinal vein and the failure of the left brachiocephalic vein to form.⁽¹²⁾ The abnormal left superior vena cava which is derived from the left anterior cardinal vein opens into the right atrium via the

coronary sinus⁽¹³⁾ (Figure 6). In our study, we found this type of left superior vana cava. In a report, the two cases of 300 Japanese cadavers dissected at Kyoto University from 1980-1989, were the double superior vena cavae. In both cases, they drained into the coronary sinuses.⁽¹⁴⁾

Two cases were reported on persistency of left superior vena cava (PLSVC) draining into left atrium with a normal coronary sinus.⁽¹⁵⁾ In 1991, Briggs C.A., et.al., reported two cases of PLSVC

with atrial displacement and inversion of the venous drainage from the body wall. (azygos system of vein)⁽¹⁶⁾

(dorsal view).

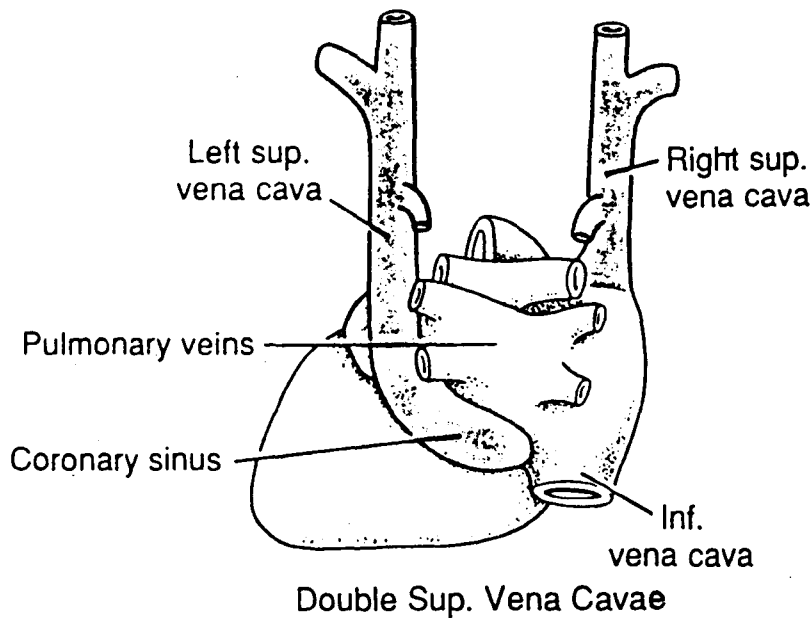


Figure 6. Sketch of the dorsal view of the heart showing double superior vena cavae. The communicating (brachiocephalic vein) between the two anterior cardinals has failed to develop. (From Langman's Medical Embryology. The cardiovascular system.

In 1991, Hayase et.al., reported a case of ASD associated with absence of coronary sinus (completely unroofed coronary sinus). The PLSVC was connected to the hemiazygos vein before it drained into the left atrium. The left innominate vein was also absent.⁽¹⁷⁾

In the pathological collection of the John Hopkins Hospital, there was a report on 1,208 specimens of hearts with congenital abnormality of CVS, only 104 cases (9%) had a PLSVC

with a coronary sinus connective. The association between PLSVC and A-V canal of defects, i.e. cor-triatriatum, and mitral atresia are found more frequently. It is also statistically significant. However it was rarely observed between PLSVC and atrial septal defect or patent foramen ovale as a primary defect.⁽¹⁸⁾ An analysis of superior vena cava abnormalities from 510 patients with congenital heart diseases found that 11% of cases had complete bilateral superior vena cavae and 72% in

patient associated with situs abnormalities.⁽¹⁹⁾ In 1986, Rey C., et al reported a case of double superior vena cavae. The left superior vena cava drained into the coronary sinus and right superior vena cava drained into the left atrium. These two vena cavae were intercommunicated by anastomoses.⁽²⁰⁾ In 1986, Yoshida K. reported a

case of double superior vena cavae which was communicated by left brachiocephalic vein.⁽²¹⁾ This abnormality is usually found association with other abnormalities of heart and great vessels. The surgical correction will be excellent if the diagnosis is early and no other severe cardiac anomalies involved.⁽²²⁻²⁴⁾ (Table 3)

Table 3. Comparison in various sources of Double Superior Vena cavae associated with congenital Heart diseases.

Sources	Total cases	Number of Double Superior Vena Cavae	%	Congenital Heart diseases
Nsah E.N.,et al., in 1991	1,208	104	9	A-V defects i.e., cortriatriatum, Mitral atresia etc.
Buirski.,et al.,	510	56	11	Situs abnormality in 1986 of Heart (72 %)
Moric, et al.,	300	2	0.67	No record of congenital heart diseases
Urai & Vilai (present report 1995)	450	1	0.02	No congenital heart

Chest X-ray is a presumptive diagnosis of this vascular anomaly. Confirmation of the anomaly is achieved by CT, echocardiography or by contrast transesophageal.⁽²⁵⁻²⁸⁾

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