# A CASE OF DOUBLE SUPERIOR VENAE CAVAE WITH PAIRED AZYGOS VEINS

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Abstract: A case of double superior venae cavae with completely paired azygos veins was found in a 76-year-old Japanese man during an ordinary dissection by medical students at Nara Medical University in 2000. The left superior vena cava was formed by the union of the left internal jugular vein and the left subclavian vein, coursed vertically downward along the thoracic aorta, and entered the coronary sulcus. The vein ran horizontally toward the right in the coronary sulcus and opened into the right atrium. A typical left brachiocephalic vein was not found. The left superior vena cava was the same size as the right one, without communication between them. Paired azygos veins were present.

Key words: double superior venae cavae, azygos vein, venous anomaly, gross anatomy

#### INTRODUCTION

Recently, cases of double superior venae cavae have been found clinically by the use of heart catheter and angiography<sup>1,2)</sup>. However, this anomaly is rarely encountered in an ordinary dissection room<sup>3)</sup>. During an ordinary dissection by medical students at Nara Medical University, a rare case of double superior venae cavae with bilateral azygos veins was found in a Japanese man. The finding of double superior venae cavae with paired azygos veins is described in the present article.

#### **FINDINGS**

During an ordinary dissection by medical students at Nara Medical University in 2000, a case of double superior venae cavae was found in a 76-year-old Japanese man who died of chronic renal failure, hypertension and diabetes mellitus.

### Left Superior Vena Cava

The left superior vena cava was formed by the union of the left internal jugular and left subclavian veins just posterior to the left sternoclavicular joint (Fig. 1). The caliber of the left superior vena cava was about 16 mm at the origin. The left superior vena cava coursed vertically downward along the thoracic aorta, sat posterior to the left pulmonary artery, and finally entered the coronary sulcus (Fig. 2). Its length was about 104 mm from the origin to the point of the entry into the coronary sulcus.

During the course of running horizontally toward the right into the coronary sulcus, the left superior vena cava became wider, reaching a diameter of about 20 mm (Fig. 3). The left superior vena cava continued the coronary sinus and the coronary sinus opened below an

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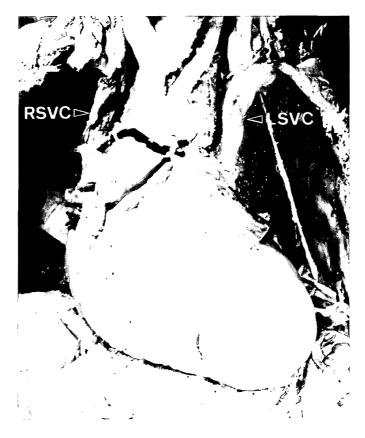


Fig. 1. The heart in situ. LSVC and RSVC indicate the left superior vena cava and the right superior vena cava, respectively.

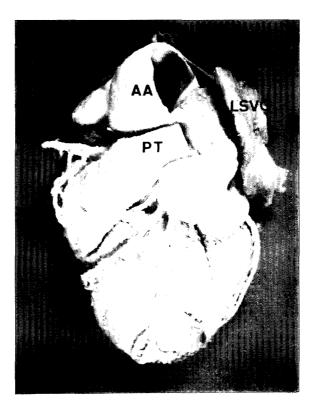


Fig. 2. Anterior view of the resected heart. AA = aortic arch; LSVC = left superior vena cava; and PT = pulmonary trunk. The arrowhead indicates the anterior interventricular sulcus.

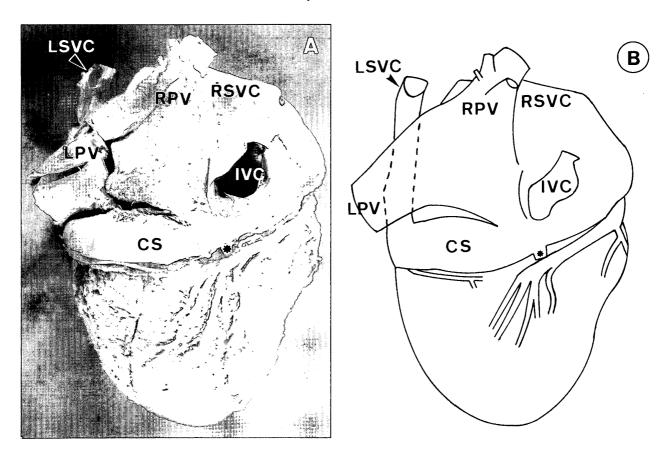


Fig. 3. Posterior view of the resected heart (A) and the tracing (B). CS = coronary sinus; IVC = inferior vena cava; LPV = left pulmonary vein; LSVC = left superior vena cava; RPV = right pulmonary vein; and RSVC = right superior vena cava. The asterisk indicates the middle cardiac vein.

orifice of the inferior vena cava. In this course, the coronary sinus received the great cardiac vein, the posterior veins of the left ventricle, and the middle cardiac vein.

The left superior vena cava received the hemiazygos vein and two supreme intercostal veins (Fig. 4). Communication between the left and right superior venae cavae was absent.

## Right Superior Vena Cava

The right superior vena cava was formed in the usual way by the union of the right internal jugular and right subclavian veins. The right superior vena cava coursed downward in front of the root of the right lung, sat right and posterior to the ascending aorta, and terminated in the superior portion of the right atrium. At about 35 mm below the formation, the right superior vena cava received the azygos vein on the right posterior wall. The openings of the right superior vena cava and inferior vena cava into the right atrium had calibers of about 17 mm and 18 mm, respectively.

Calibers of the left and right superior venae cavae were similar to each other.

## Azygos and Hemiazygos Veins

The azygos venous system was made up of bilaterally symmetric veins (Fig. 4). The hemiazygos vein ran upward along the left side of the vertebral column and joined to the

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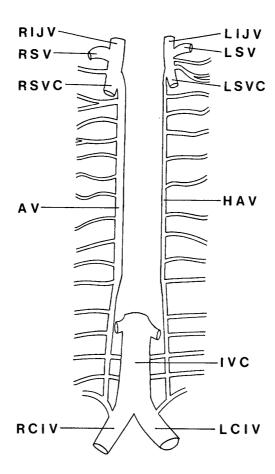


Fig. 4. Schematic representation of the azygos system. AV = azygos vein; HAV = hemiazygos vein; IVC = inferior vena cava; LCIV = left common iliac vein; LIJV = left internal jugular vein; LSV = left subclavian vein; LSVC = left superior vena cava; RCIV = right common iliac vein; RIJV = right internal jugular vein; RSV = right subclavian vein; and RSVC = right superior vena cava.

posterior wall of the left superior vena cava at the height of the 3rd thoracic vertebra. The hemiazygos vein received the intercostal veins and the left superior vena cava received two supreme intercostal veins.

The azygos vein ran upward along the right side of the vertebral column and joined to the posterior wall of the right superior vena cava at the height of the 3rd thoracic vertebra.

The azygos vein received the intercostal veins and the right superior vena cava received a supreme intercostal vein. There was no anastomosis between the azygos and hemiazygos veins. The azygos and hemiazygos veins had calibers of 5 mm and 3 mm, respectively. The azygos vein was larger than the hemiazygos vein.

#### Heart

The chamber of the right ventricle was very small, whereas the chamber of the left ventricle was large. No other anomalous finding was found in the heart.

## **DISCUSSION**

The incidence of double superior venae cavae ranged from 0.13% to 0.49% in adult subjects<sup>3-6)</sup> and was 1.6% in fetus and newborn<sup>7,8)</sup>. Most cases of fetus or newborn were associated with some congenital cardiac anomalies. The authors found the double superior venae cavae in one out of 280 adult subjects at our Department. The incidence of this anomaly is 0.36% at our Department.

Anomalies of the superior vena cava were classified by many investigators<sup>9-12)</sup>. According to the condition of anastomosis between the left and right superior venae cavae, McCotter<sup>9)</sup> classified double superior venae cavae into the following five types: (1) Double superior venae cavae without anastomosis, (2) double superior venae cavae with small anastomosis, (3) double superior venae cavae with normal anastomosis, (4) left superior vena cava without right, and (5) persistent left superior vena cava unclassified. He reported that more than half the cases of this anomaly belonged to the 1st and 2nd types. The present case belonged to the 1st type of McCotter<sup>9)</sup>.

Cases of double superior venae cavae with completely paired azygos veins are rare. To our knowledge, six cases were reported by Yamamoto<sup>13</sup>, Fujimoto et al.<sup>10</sup>, Otsuka<sup>14</sup>, Inoue and Masuko<sup>15</sup>, Mori et al.<sup>16</sup>, and Yoshida et al.<sup>17</sup> in Japan.

It is well known that in the human fetus, the veins develop symmetrically in the early stage<sup>6</sup>. The heart in the embryo of six weeks has paired common cardinal veins which are formed by the union of anterior and posterior cardinal veins. Thereafter, an anastomosis takes place between right and left anterior cardinal veins (formation of the left brachiocephalic vein), whereas the left anterior cardinal vein becomes the supreme intercostal vein and the left common cardinal vein becomes the oblique vein of the left atrium. The proximal portions of the left posterior and superior cardinal veins are transformed into a hemiazygos system which anastomoses to the right azygos system. Finally, a symmetric system of the veins changes into a right dominant system.

It is considered that the development of a symmetric venous system is characterized by two remarkable events of both the anastomoses between the right and left anterior cardinal veins and between the right and left azygos veins. If these two events do not take place for some reasons, a symmetric venous system will remain.

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