

A persistent left superior vena cava

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ABSTRACT

Persistent left superior vena cava is the most common form of anomalous venous drainage involving the superior vena cava, and represents persistence of the left horn of the embryonic sinus venosus, which normally involutes during normal development to become the coronary sinus. Almost always, a persistent left superior vena cava enters the right atrium through the orifice of an enlarged coronary sinus. In this case report of a 60-year-old male cadaver, we describe a persistent left superior vena cava and discuss its embryology and clinical significance.

Keywords: anomalous venous drainage, coronary sinus, persistent left superior vena cava, sinus venosus, superior vena cava

Singapore Med J 2007; 48(3):e90–e93

INTRODUCTION

Incidence of a persistent left superior vena cava (PLSVC) is uncommon. PLSVC is seen in 0.3%–0.5% of the normal population^(1,2) and 1.5%–10% of patients with other congenital heart abnormalities.^(2,3) It is a persistent remnant of a vessel that is present as an embryological counterpart of the normal right-sided superior vena cava. Presence of a PLSVC was first reported in 1787, and the first account of foetal structures comprising PLSVC was published by Marshall in 1850.⁽⁴⁾ In 1975, de Leval et al reported that PLSVC occurred in 2.1%–4.3% of patients with congenital heart disease, making PLSVC the most common venous anomaly associated with congenital heart deformities.⁽⁵⁾

CASE REPORT

During a routine dissection for the medical students, we found a PLSVC in a 60-year-old male cadaver. The PLSVC was connected to the right superior vena cava by a narrow communicating vein (Fig. 1). Both the vena cavae were formed as a continuation of brachiocephalic vein (innominate vein) of the corresponding side. The PLSVC continued as the coronary sinus (Fig. 2). The coronary sinus was enlarged and drained into the right atrium between the opening of inferior vena cava and the right atrio-ventricular opening (Fig. 2).

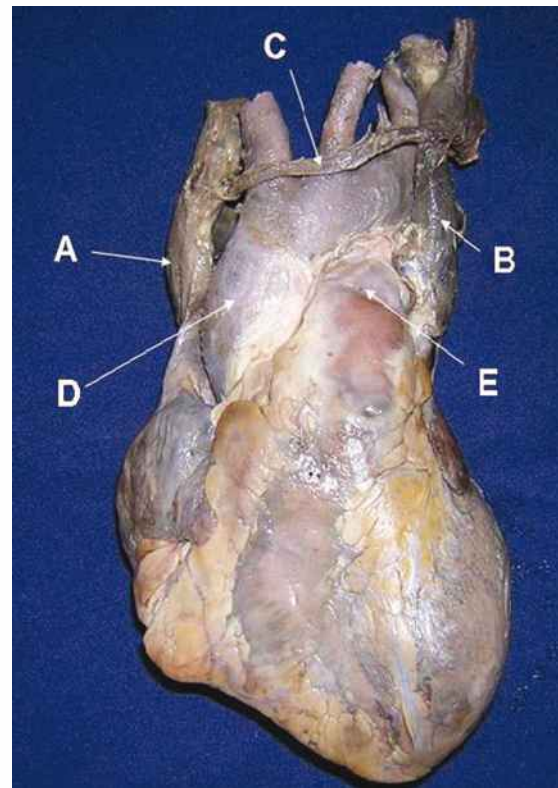


Fig. 1 Photograph of the anterior view of the heart. A: superior vena cava (right side); B: superior vena cava (left side); C: communicating vein; D: aorta; E: pulmonary trunk.

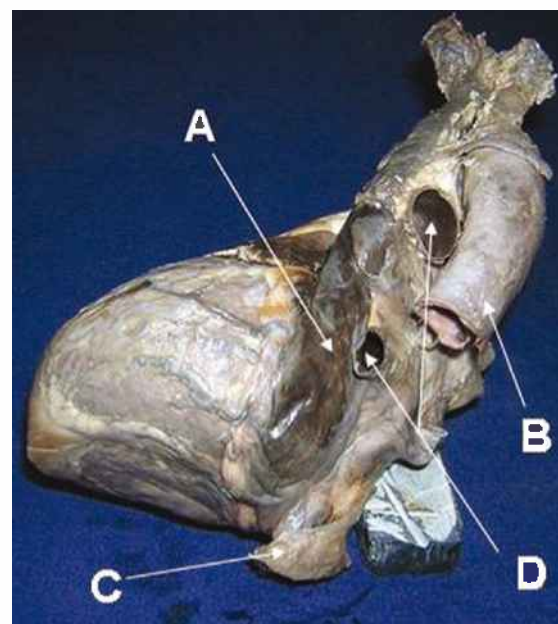


Fig. 2 Photograph of the lateral view of the heart. A: coronary sinus (enlarged); B: aorta; C: inferior vena cava; D: left pulmonary veins.

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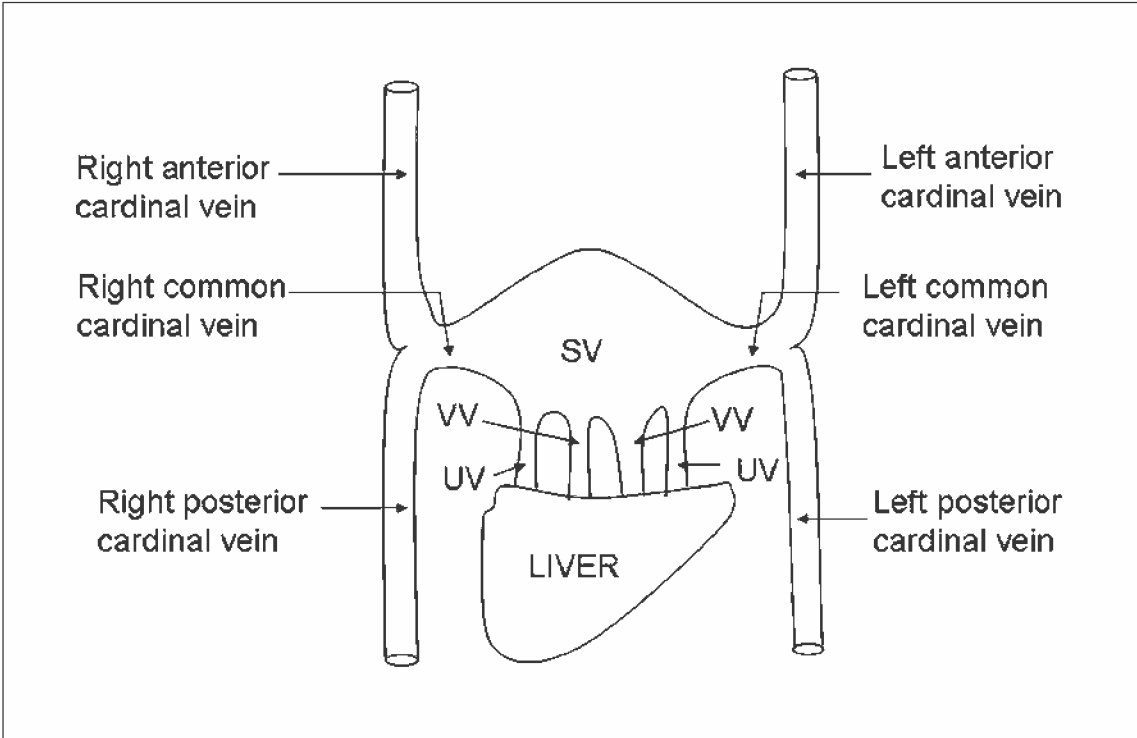


Fig. 3 Diagram shows the cardinal veins. SV: sinus venosus; UV: umbilical vein;VV: vitelline vein.

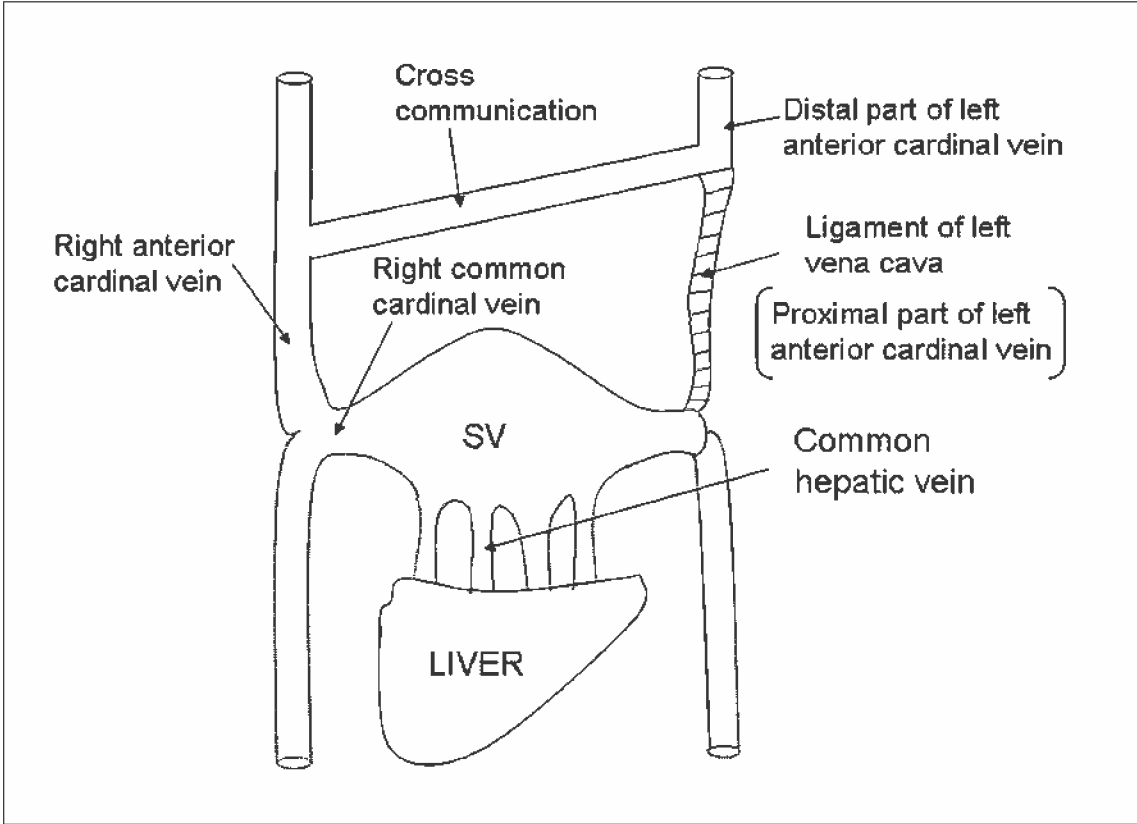


Fig. 4 Diagram shows the changes in the left anterior cardinal vein. SV: Sinus venosus.

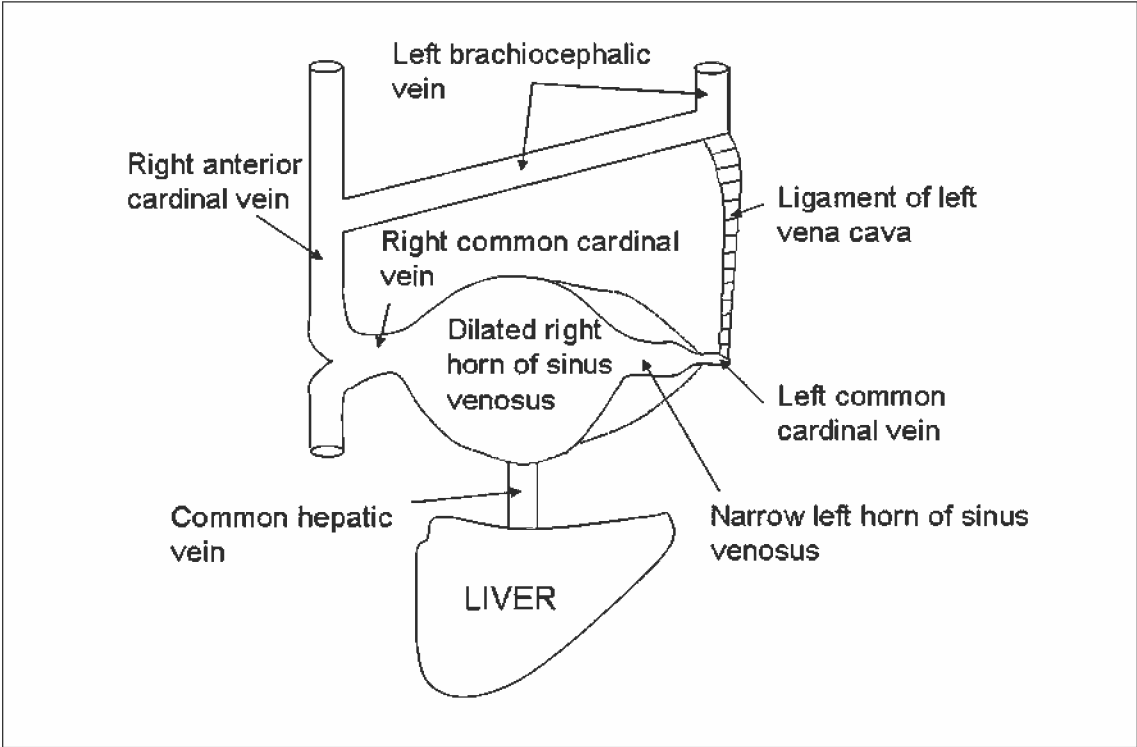


Fig. 5 Diagram shows the formation of the left brachiocephalic vein.

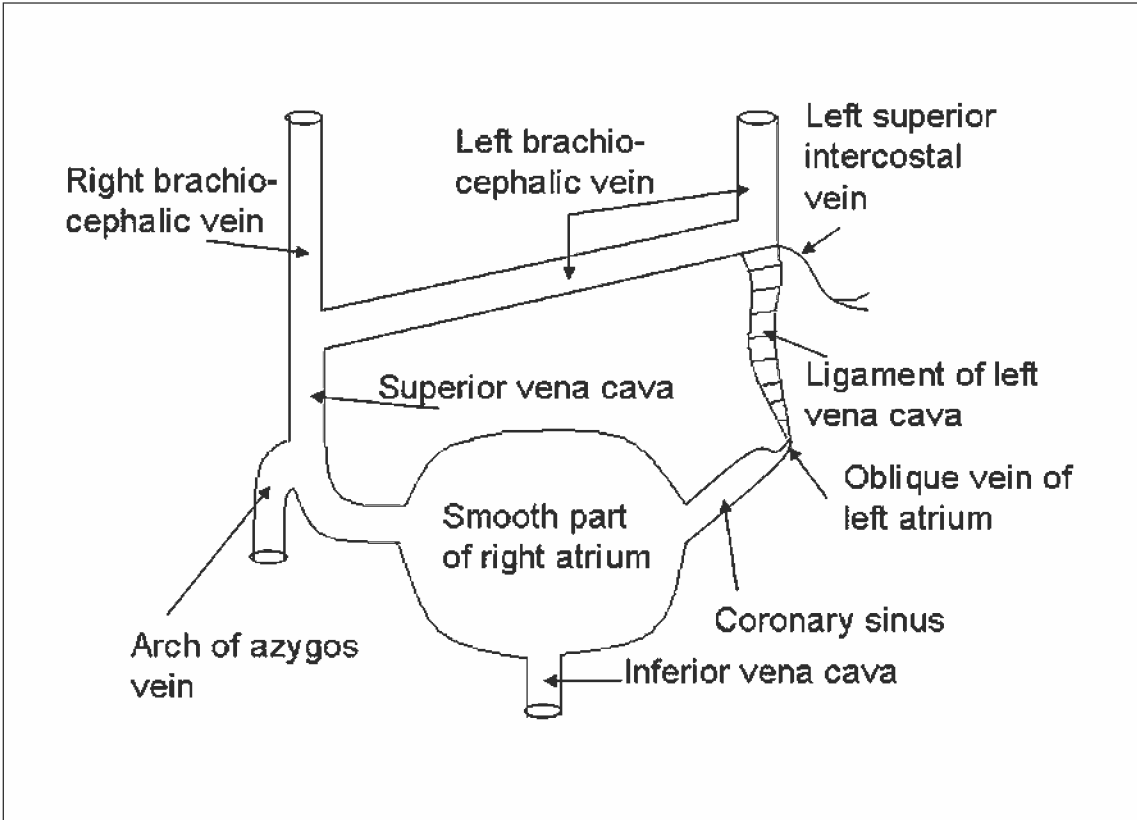


Fig. 6 Diagram shows the formation of the superior vena cava.

DISCUSSION

There are two types of PLSVC reported in the literature. PLSVC connecting to the right atrium via coronary sinus forms 90% of the anomalies of the superior vena cava. In the other 10%, PLSVC connects to the left atrium.^(1,2) PLSVC connecting to the roof of the left atrium is very rare, and in this case, the anomaly is termed complete unroofing of the coronary sinus. The orifice of the coronary sinus then persists as an interatrial communication.⁽¹⁾ In most of the cases, including this case report, the PLSVC enters the right atrium through the orifice of an enlarged coronary sinus. In this case, the defect in the development is considered to be an anomaly of the coronary sinus.

We review the development of the superior vena cava in detail:⁽⁶⁾ in the human foetus, in the fifth week of intrauterine life, three pairs of major veins can be distinguished: (a) the vitelline, carrying blood from the yolk sac to the sinus venosus; (b) the umbilical veins, originating in the chorionic villi and carrying oxygenated blood to the embryo; and (c) the cardinal veins, draining the body of the embryo proper (Fig 3). Initially, the cardinal veins form the main venous drainage system of the embryo. This system consists of the anterior cardinal veins, which drain the cephalic part of the embryo, and the posterior cardinal veins, which drain the remaining part of the body of the embryo. The anterior and posterior veins join before entering the sinus horn and form the short common cardinal veins. During the fourth week, the cardinal veins form a symmetrical system (Fig. 3).

Formation of the vena cava system is characterised by the appearance of anastomoses between the left and right sides in such a manner that the blood from the left side is channelled to the right side (Fig. 4). The anastomosis between the anterior cardinal veins develops into the left brachiocephalic vein (Figs. 4 & 5). Most of the blood from the left side of the head and the left upper extremity is then channelled to the right. The terminal portion of the left anterior cardinal vein entering into the left brachiocephalic vein is retained as a small vessel, the left superior intercostal vein (Fig. 6). This vessel receives blood from the second and third intercostal spaces. The superior vena cava is formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein.

A left-sided superior vena cava is an abnormality caused by the persistence of the left anterior cardinal vein and obliteration of the common cardinal and proximal part of the anterior cardinal veins on the right. In such a case, blood from the right is channelled

toward the left by way of the right brachiocephalic vein. The left superior vena cava drains into the right atrium by way of the left sinus horn, i.e., the coronary sinus.

The clinical implications of a coronary sinus dilatation (Fig. 3) are:⁽²⁾

1. Cardiac arrhythmia due to stretching of the atrioventricular node and bundle of His.
2. Obstruction of the left atrioventricular flow because of partial occlusion of the mitral valve. The symptoms may increase due to left to right shunt across the atrial septum.

In cases of the PLSVC opening into the coronary sinus, the ostium of the coronary sinus opens directly into the right atrium in close proximity to the insertion of the atrioventricular valves. If the coronary sinus is dilated, it can create the appearance of an atrioventricular canal defect. Park et al presented three cases in which the correct diagnosis avoided an unnecessary termination of pregnancy.⁽⁷⁾ A PLSVC has important clinical implications in certain situations. It may complicate placement of cardiac catheters or pacemaker leads. Awareness of this anomaly may reduce confusion about the position of a catheter/lead that appears to have strayed.⁽⁴⁾

During cardiac surgery, a PLSVC is a relative contraindication to retrograde administration of cardioplegia.⁽³⁾ The coronary sinus catheter balloon may not be able to occlude the dilated coronary sinus, resulting in the failure to ensure retrograde flow of cardioplegia to the myocardium. Also, cardioplegia delivered would largely be distributed to the left internal jugular and left subclavian veins, rather than myocardium. The coronary sinus would have to be carefully dissected during heart transplant so that the PLSVC can be re-anastomosed to the right atrium.⁽³⁾

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