Congenital Anomalies of the Superior Vena Cava: A CT Study

By Mary G. Cormier, Joseph W. Yedlicka, Richard J. Gray, and Rogelio Moncada

The precise and elegant rendering of the human anatomy achieved by CT has, for over a decade, provided a novel perspective to the medical diagnostic armamentarium. CT has had a significant impact on almost every avenue in the daily practice of medicine and has gained broad acceptance by our clinical colleagues.

A wide variety of congenital vascular anomalies of the upper mediastinum exist. Since most of these are clinically silent, they are often unsuspected and discovered incidentally on radiographic studies done for other reasons. Familiarity with the CT appearance of such anomalies may aid in the interpretation of otherwise potentially confusing CT images.

Innovative techniques for solving clinical problems have been designed. Some have improved our diagnostic capabilities, while others have introduced interventional techniques that offer therapeutic and palliative alternatives to surgery. CT has had a significant impact on just about every avenue in the daily practice of medicine. Its broad acceptance by our clinical colleagues and the documented direct benefit to the patient crown a historical decade of medical imaging progress.

ANATOMIC CONSIDERATIONS

The normal SVC is devoid of venous valves; it drains the returning blood flow of the head and upper half of the trunk. The vessel measures approximately 7 cm in length. Its cross-sectional shape varies in proportion to its intraluminal flow or pressure, and generally exhibits an oblong, flat, or rounded contour; the latter more often seen when the patient is examined during a Valsalva maneuver. The caliber of the SVC varies, but generally is 1.5 cm and rarely as much as 2.0 cm at its greatest diameter.

The short and vertically oriented right brachiocephalic vein and its longer oblique counterpart on the left form the origin of the SVC at the level of the first right costal cartilage. From this point, the SVC descends vertically along the right side of the mediastinum to end in the upper part of the right atrium at the level of the right third costal cartilage. In its descent it describes a minimal convexity to the right. Its lower half is covered by the parietal pericardium and its wall is partially composed of visceral pericardium. The major tributary of the SVC, the azygos vein, enters the dorsal aspect of the mid-SVC.

Ventrally, the SVC is related to the mediastinal fat, pleura, thymus, and adjacent right lung. Caudally, it is overlapped at the left margin of its intrapericardial portion by the ascending aorta. The SVC is related posteriorly to the innominate artery, trachea, right main bronchus, right main pulmonary artery, right superior pulmonary vein, and vagus nerve. In addition, the SVC is flanked by a large number of adjacent lymph nodes; most of these occur along the dorsal aspect.

EMBRYOLOGY

The embryo has a closed blood circuit by the end of the second week of fetal development. By the third or fourth week, two paired venous trunks are formed: the anterior cardinal veins, arising from the cranial part of the body, and the posterior cardinal veins, arising from the caudal part of the body. On each side of the heart the ipsilateral anterior and posterior cardinal veins unite into a short main trunk called the common cardinal vein or duct of Cuvier. Each duct of Cuvier in turn empties into the sinus venosus.

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<th>ABBREVIATIONS</th>
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<tr>
<td>ASD, atrial septal defect</td>
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<td>IVC, inferior vena cava</td>
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<td>PDA, patent ductus arteriosus</td>
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<td>RA, right atrium</td>
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<td>SVC, superior vena cava</td>
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<td>VSD, ventricular septal defect</td>
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Normally, as the heart descends caudally, a wide transverse anastomosis forms between the two anterior cardinal veins; this channel becomes the left innominate vein, presumably derived from the union of smaller thyroid or thymic branches of the primitive jugular veins. The segment of the right anterior cardinal vein that lies caudal to this anastomosis, together with the right common cardinal vein (right duct of Cuvier), forms the primitive SVC.

At this stage a left SVC also exists, similarly formed by the corresponding segment of the left anterior cardinal vein that lies caudal to the primitive left innominate vein, and the left common cardinal vein (left duct of Cuvier). A major portion of this left SVC undergoes atrophy during the following few weeks. Its cranial remnant forms the trunk of the first left intercostal vein, and its caudal remnant becomes the oblique vein of the left atrium. In some cases the involuted vessel remains as a few fibrous strands or as a fibrous cord that attaches to the posterolateral aspect of the pericardium and coronary sinus, referred to as the oblique ligament of Marshall.

The sinus venosus is oriented transversely, lies dorsal to the undivided atrial part of the heart, and communicates with the single common atrial chamber through an oval opening. Its lateral portions, known as horns, become directed upward and posteriorly as the heart descends into the thorax. By approximately 4 to 5 weeks the symmetric arrangement is lost. The sinoatrial opening migrates to the right, and an atrial septum is formed in such a manner that the sinus venosus communicates only with the right atrial chamber. Eventually the left horn of the sinus venosus becomes the coronary sinus. The right horn expands, receiving blood from the SVC and future IVC, and ultimately is totally absorbed into the wall of the right atrium.

Many anomalies of the SVC are known, each representing some form of developmental arrest. The basic abnormality is failure of the left anterior cardinal vein to involute normally, resulting in persistence of a left SVC.

**LEFT SUPERIOR VENA CAVA**

The sign of a left SVC, either alone or in conjunction with a right SVC, depends upon resultant hemodynamic effects. Approximately 75% of both double SVC and single left SVC are asymptomatic and have no circulatory disturbance. However, in the remainder significant dysfunction may exist, possibly severe enough to be incompatible with life.

**Incidence**

The true overall incidence of this condition is difficult to establish. Estimates ranging from 1 per 330 to 1 per 750 normal individuals, and 1 per 25 patients with congenital heart disease are reported. Among these, a coexisting right SVC is present in about 85%.

**Anatomic Relations**

When a double SVC exists, the left SVC descends vertically, anterior and to the left of the aortic arch and main pulmonary artery, slightly lateral to the left vagus nerve. It has been said by some to course between the left pulmonary artery and pulmonary veins, and by others to lie anterior to all of the left pulmonary vessels. The latter position corresponds to that of the right SVC relative to the right pulmonary vessels.

The left SVC then swings medially, pierces the pericardium to run in the posterior atrioventricular groove, and enters the coronary sinus. In this case, the coronary sinus is often abnormally large, perhaps due in part to an increased volume flowing through it, and partially to regurgitant enlargement during atrial contraction. In approximately 25% of 174 cases of left SVC, Winter found the left hemiazygos vein looping over the left main bronchus to enter the left SVC posteriorly, at the same level that the azygos vein enters the right SVC.

When a double SVC exists, the caliber of the right vessel is often normal; however, it may be smaller than usual because of reduced blood volume. The diameter of the left cava rarely exceeds that of the right; it is frequently equal to it or smaller.

**Specific Entities**

Figure 1 schematically depicts the major patterns and types of developmental anomalies of the SVC. The most frequent is a double vena cava, with normal right SVC anatomy and a left SVC entering the coronary sinus, and subsequently draining into the right atrium. There may or may not be a transverse anastomosis between the paired cavae. When there is none, the right and left halves of the head and upper truck drain back to the heart independently, and
the coronary sinus is large, approaching the caliber of the cava entering it (Fig 2). When an anastomosis does exist between the two cavae, it may be oriented inferiorly toward the right, inferiorly toward the left, or directed transversely. The relative size of the two cavae and of the coronary sinus depends on the amount of blood each carries as routed above. In some cases, additional cross-communication exists in the form of several small, slender, irregularly branching veins, a plexiform arrangement that Papez suggests probably represents the persistence of vessels that normally contribute to the formation of the left innominate vein.

The most frequent cardiac anomaly associated with a left SVC that drains into the RA is an ASD. Gensini et al noted that a left-to-right shunt at the atrial level is often present in conjunction with anomalous pulmonary venous drainage into either a left or right SVC. The anomalous drainage in the majority of cases is total. Septal defects always coexist. Infrequently the left SVC enters the left atrium or the left side of a common atrium. In this case other cardiac anomalies are almost always associated.

The true incidence of a persistent left SVC with absence of a right SVC is not known, but it is rare. Functionally, this arrangement results in no circulatory disturbance if the left SVC drains directly or indirectly into the right atrium (Fig 3).

A left SVC may be associated with a wide variety of cardiac defects, including single atrium, VSD, PDA, tetralogy of Fallot, truncus arteriosus, pulmonary stenosis or atresia, Ebstein malformation, atroventricular canal defects, Taussig-Bing complex, mitral atresia, tricuspid atresia, transposition, anomalous pulmonary venous return, and coarctation of the aorta.

There is a higher incidence of left SVC in cases of transposition of the viscera. Campbell and Deuchar found a left SVC in 40% of 46 cases of congenital heart disease in which there was also some degree of visceral transposition. A double SVC is more common when visceral transposition is partial than when it is complete. Finally, left SVC is frequently described with asplenia.

**VERTICAL VEIN**

Careful distinction should be made between a persistent left SVC and an anomalous left vertical vein. The left vertical pulmonary vein that receives partial or total left pulmonary venous drainage, ascends superiorly, usually anterior to
Fig 2. Double SVC with normal right SVC anatomy and the left SVC draining into an enlarged coronary sinus. This patient also had an unrelated incidental finding: hemiazygos continuation of a left inferior vena cava. SVC, superior vena cava; Ao, aorta; MPA, main pulmonary artery; LPA, left pulmonary artery; CS, coronary sinus; R, right; L, left; h, hemiazygos vein.
Fig 3. Left SVC in the absence of a right SVC. The right superior pulmonary vein should not be mistaken for a right SVC. R In V, right innominate vein; L In V, left innominate vein; SVC, superior vena cava; Ao, aorta; LPA, left pulmonary artery; RPA, right pulmonary artery; PA, main pulmonary artery; R, right; L, left.
the aorta, and drains into the left innominate vein. It is now generally accepted that a true left SVC drains into the heart.\(^{13,14}\)

**ANOMALIES OF THE RIGHT SUPÉRIOR VENA CAVA**

Relatively few intrinsic right SVC anomalies occur. The right SVC rarely drains into the left atrium without any coexisting defects. This creates a right-to-left shunt with cyanosis. Only a handful of cases have been reported.\(^{11,15,16}\)

Freedom et al\(^{17}\) reported a case in which the right SVC drained into the RA via a low insertion associated with complex congenital heart defects.\(^{17}\)

Congenital aneurysm of the SVC is rare. It is nonetheless important in the differential diagnosis of a superior mediastinal mass. Two distinct types are recognized: saccular and fusiform. Direct opacification of the vascular lesion, either angiographically or on contrast-enhanced CT, provides the diagnosis (Fig 4). It is generally accepted that because this is a low-pressure system, the aneurysm does not thrombose, enlarge, or rupture, making conservative management appropriate.\(^{18,19}\)

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