Congenital anomalies of the vena cava: CT and MR imaging findings

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Learning objectives

1. To illustrate the embryologic development of the vena cava and describe the variations in vena cava anatomy.

2. To present CT and magnetic resonance (MR) imaging appearances of the more frequently encountered anomalies and some unusual variants.
Background

Congenital anomalies of the vena cava and its tributaries have become more commonly recognized in asymptomatic patients since the development of cross-sectional imaging. In addition, these anomalies are sometimes detected during surgical or diagnostic procedures. Vascular structures are usually readily identified on CT scans of the chest or abdomen obtained with intravenously administered contrast material. However, with the use of multidetector CT (MDCT), the venous structures may be imaged during the arterial phase, when little or no contrast material is present in the veins. This fact should be borne in mind to avoid pitfalls, and in case of a possible filling defect, delayed imaging should be performed. The isotropic data sets that are possible with the MDCT scanners allow superior-quality multiplanar reformation, which is useful in delineating vena cava anomalies. On the other hand, magnetic resonance (MR) imaging is the most reliable technique for depicting the presence of anomalies with no use of contrast materials. Familiarity with these variations of anomalies is essential for correct interpretation of cross-sectional images such as MDCT or MR, to avoid erroneous diagnosis of retroperitoneal and mediastinal masses or adenopathy, to alert the surgeon and angiographer of potential sources of complications preoperatively.
Imaging findings OR Procedure details

1) Embryogenesis of the IVC (Fig 1)

The embryologic development of systemic veins is complex and subject to considerable variation. However, a basic knowledge of the embryogenesis of the vena cava is essential for understanding the anomalies of the vena cava. We review the embryogenesis of the vena cava which is a composite structure formed from the continuous appearance and regression of three paired embryonic veins.

Fig.: Drawing illustrates embryologic development of the vena cava
References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

A. Anterior cardial vein and posterior cardial vein

In the early embryonic stage, paired anterior cardinal veins drain the cranial portion of the embryo, and posterior cardinal veins drain the caudal portion of the embryo. Blood return from the viscera is conveyed by the vitelline veins which drain the yolk sac.
During normal development, the anterior cardinal veins, which drain the head, neck, and arm, unite with the posterior cardinal vein to form the common cardial vein and enter the heart as the right and left horns of the sinus venosus. Most of the left-sided cardinal system disappears, leaving only the coronary sinus and a remnant known as the ligament of Marshall.

The distal ends of the anterior cardinals give rise to the internal jugular veins. The proximal end of the left anterior cardinal vein regresses but the proximal end of the right anterior cardinal becomes the superior vena cava.

Posterior cardinal veins are progressively replaced, first by the subcardinal veins and later by the supracardinal veins, which together form the subhepatic IVC, but persist in the pelvis as the common iliac veins.

**B. Subcardinal vein**

The subcardinal veins develop ventromedial to the posterior cardinal veins and ventrolateral to the aorta. The right subcardinal vein forms the suprarenal segment of the IVC. The intersubcardinal anastomosis forms between the paired subcardinal. Anastomoses between the posterior cardinal and subcardinal veins also develop on each side. At the same time, union occurs between the right subcardinal vein and the hepatic segment of the IVC, which forms from the vitelline vein.

**C. Supracardial vein**

Subsequently, the next major development is the appearance of the paired supracardinal veins which lie dorsomedial to the posterior cardinal veins and dorsolateral to the aorta. In the thoracic region, the supracardinal veins give rise to the azygos and hemiazygos veins. Caudally, the right supracardinal vein forms the infrarenal segment of IVC.

**D. Others**

Multiple anastomoses form between the posterior and supracardinal veins 3). On each side, a suprasubcardinal anastomosis develops from union of the postsupracardinal and the postsubcardinal anastomoses. Then these anastomoses form the intervening renal segment of the IVC.

Caudally, anastomoses develop between the two posterior cardinal veins and between the posterior and lumbar supracardinal veins. With atrophy of the posterior cardinal veins, blood return from the lower extremities is shunted through the supracardinal system to the suprasubcardinal anastomosis, then to the prerenal division of the IVC.

The embryonic ureter passes posterior to the posterior cardinal veins and anterolateral to the supracardinal vein. Therefore the formation of the postsupracardinal anastomosis
inferiorly and the suprasubcardinal anastomosis at the level of the kidney allows
development of the periureteric venous ring. The embryonic kidneys are initially drained
by paired ventral and dorsal limbs and both dorsal limbs usually regress. On the right
side, the ventral limb is incorporated into the lateral wall of the renal segment of the IVC.
On the left side, the ventral limb forms the left renal vein.

2) Congenital anomaly of the superior vena cava

A. Persistent left superior vena cava; PLSVC (Fig 2)

Persistent left superior vena cava (PLSVC) is rare but important congenital vascular
anomaly. It results when the left superior cardinal vein to the innominate vein fails to
regress2). It is most commonly observed in isolation but can be associated with other
cardiovascular abnormalities including atrial septal defect, bicuspid aortic valve and
coarctation of aorta. This usually drains into the right atrium via the coronary sinus. On
rare occasions, PLSVC drains into the left atrium or the pulmonary veins creating a left to
right shunt4). Individuals with PLSVC usually possess a normal right superior vena cava,
and thus the condition is not routinely detected.

Most cases have been reported as incidental findings during central venous catheter
placement, pacemaker and implantable cardioverter defibrillator (ICD) implantation and
during thoracic surgery for various reasons. When an unknown PLSVC is incidentally
discovered during these procedures, it may cause technical difficulties and life-
threatening complications.

Suspicion of left superior vena cava may arise on the postero-anterior chest X-ray, where
it may appear as widening of the aortic shadow or paramedian stripe. CT with contrast
material and MRI offer voluminous information in delineating the anatomy 5). They also
help identify other cardiac anomalies that might be associated.
Fig.: Persistent left superior vena cava. (a) Coronal reformatted CT image shows a left sided SVC (small arrow). Large arrow indicates normal right sided SVC. (b) This vessel drains into the right atrium (RA) via the coronary sinus (CS).

References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

3) Congenital anomalies of the inferior vena cava

A. Left IVC (Fig 3)

Left IVC has a prevalence of 0.2-0.5% and results from regression of the right supracardinal vein with persistence of the left supracardinal vein. The left IVC typically joins the left renal vein, and together they course anterior to the aorta to join the normal right-sided IVC. This anomaly is of no clinical importance. However, it may be mistaken for left sided paraaortic adenopathy. In addition, transjugular access to the infrarenal IVC for placement of an IVC filter may be difficult.
**Figure 3**

**Fig.**: Left IVC (with retroperitoneal teratoma) Coronal reformatted CT image shows a left-sided IVC (V) joining the left renal vein (LRV), which unites with the right renal vein to form a normal right-sided suprarenal IVC. Ao: aorta. \(\rightarrow\): right gonadal vein * teratoma

**References**: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

**B. Double IVC** (Fig 4)

Double IVC has a prevalence of 1-3% and results from persistence of both the right and left supracardinal veins. The left IVC typically ends at the left renal vein which crosses anterior to the aorta to join the normal right-sided IVC. However, there may be variations in this arrangement. There may be significant discrepancy in the size of the two veins. Double IVC has clinical implications that are similar to those of left IVC and may be mistaken for adenopathy. Doubla IVC should be suspected in cases of recurrent pulmonary embolism following placement of an IVC filter. The options of IVC filter placement vary from a single filter in the common suprarenal IVC, to infrarenal filter in the IVC on each side.
**Fig.**: Double IVC. Contrast-enhanced CT scan obtained inferior to the renal veins shows right (large arrow) and left (small arrow) IVCs.

**References**: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

**C. Absence of hepatic segment of IVC with azygos or hemiazygos continuation** (Fig 5,6)

Absence of hepatic segment of IVC with azygos or hemiazygos continuation results from failed formation of the right subcardinal-hepatic anastomosis, with consequent atrophy of the right subcardinal vein (suprarenal IVC). Consequently blood is shunted from the suprasubcardinal anastomosis through the retrocrural azygos vein, which is partially delivered from the thoracic segment of the right supracardinal vein. The prevalence is 0.6% 1).

The infrarenal IVC continues as the azygos vein and, in case of left IVC, as the hemiazygos vein. The azygos vein joins the superior vena cava at the normal location in the right paratracheal space. The hepatic segment (the posthepatic segment) is usually not truly absent, and it drains directly into the right atrium. This anomaly has become
increasingly recognized in otherwise asymptomatic patients according to the advent of cross-sectional imaging such as CT or MRI 5).

Associated situs anomalies including asplenia or polysplenia syndrome are present in many of these cases. It is important not to misinterpret the enlarged azygos veins as a right sided paratracheal mass or retrocrural adenopathy 6). Preoperative knowledge of the anatomy may be important in planning cardiopulmonary bypass and to avoid difficulties in catheterizing the heart7).

Figure 5

**Fig.**: Absence of hepatic segment of IVC with azygos or hemiazygos continuation (a) Chest radiography shows enlargement of azygos arch (large arrow) and widening of right paraspinal stripe (small arrows) contiguous with azygos arch.

**References:** Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
Fig.: Absence of hepatic segment of IVC with azygos or hemiazygos continuation (same patient as Fig 5) (b,c,e) Contrast-enhanced CT scans show the enlarged azygos vein (*) draining into the superior vena cava (V). (d) The hepatic segment of the IVC is absent. Associated situs anomalies are present. Ao: aorta. L: liver. St: stomach. Sp: spleen.

References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

D. Absent of the infrarenal IVC

Complete absence of the infrarenal IVC with preservation of the suprarenal segment is an extremely rare anomaly. This condition may be a sequel of intrauterine or perinatal thrombosis of the IVC and not truly embryologic in origin. Lower-extremity venous return occurs via the ascending lumbar veins, which drain into the azygos-hemiazygos system. Enlarged collateral vessels may be simulate a paraspinal mass 8).

4) Congenital anomalies of the renal vein
A. Circumaortic venous ring (Fig 7)

A circumaortic venous ring results from persistence of the dorsal limb of the embryonic left renal vein and of the dorsal arch of the renal collar (intersupracardinal anastomosis). The prevalence is 2.4-8.7%. Two left renal veins are present. The superior renal vein crosses the aorta anteriorly and receives the left adrenal vein. The inferior renal vein crosses posterior to the aorta approximately 1-2cm inferior to the normal anterior vein and receives the left gonadal vein. The major clinical significance is in the planning of nephrectomy and can be misdiagnosed as adnopathy.

Fig.: Circumaortic venous ring (with aortic dissection) (a,b) Contrast-enhanced CT scans show the left renal vein coursing anterior (large arrow) and posterior (small arrow) to the aorta (Ao). The posteriorly located renal vein is caudad with respect to the preaortic vein. V: inferior vena cava. *: false lumen of the aortic dissection.

References: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

B. Retroaortic left renal vein (Fig 8)
A retroaortic left renal vein results from persistence of the dorsal arch of the renal collar and the regression of the ventral arch. The prevalence is 2.1%. A single renal vein passes posterior to the aorta. The clinical significance is preoperative recognition of the anomaly. On rare occasions, compression of the renal vein as it courses posterior to the aorta (nutcracker phenomenon) can result in periureteric varices, hypertension, and hematuria 9).

![Image of Retroaortic Left Renal Vein](image)

**Figure 8**

**Fig.**: Retroaortic left renal vein (a,b) Contrast-enhanced CT scans show the left renal vein coursing posterior (arrows) to the aorta (Ao) and draining into the inferior vena cava (V).

**References**: Department of Radiology, Kyorin University School of Medicine - Tokyo/JP

**C. Retrocaval ureter** (Fig 9)

A retrocaval ureter is also termed a circumcaval ureter. Unlike other congenital ureteral obstructions, this embryologic defect lies in the developing vena cava rather than the ureter. The right supracardinal system fails to develop, whereas the right posterior
cardinal vein persists. This anomaly always occurs on the right side. The proximal ureter courses posterior to the IVC, then emerges to the right of the aorta, coming to lie anterior to the right iliac vessels. Patients with this anomaly may develop partial right ureteral obstruction or recurrent urinary tract infections. CT clearly depicts the abnormal course of the ureter. Treatment include surgical relocation of the ureter anterior to the IVC.

**Figure 9**

**Fig.**: Retrocaval ureter (a.b.c) Coronal reformatted CT images show the right ureter (arrows) courses posterior to the inferior vena cava (V) and emerges to the right of the aorta (Ao). The proximal ureter is dilated due to the development of ureteral obstruction.

**References:** Department of Radiology, Kyorin University School of Medicine - Tokyo/JP
a. Three pair of veins  
b. Conceptual framework of the vena cava

Figure 1 Drawing illustrates the embryologic development of the vena cava

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Conclusion

Vena cava anomalies reflect an abnormal regression or persistence of the various embryonic veins. Although most anomalies are asymptomatic and are encountered incidentally, their correct identification is necessary to avoid diagnostic pitfalls and to prevent complications during surgery or catheterization.
References


