64-row MDCT and 1.5 Tesla MR Angiography for the study of congenital anomalies of the vena cava: update and pictorial review.

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Learning objectives

To review the most frequent congenital anomalies of the vena cava that were studied in our department for the last seven years.

To describe the imaging appearance of those congenital anomalies with the available techniques of contrast-enhanced acquisition for MDCT and MR angiography studies.

To review the usefulness of current post-processing acquisition techniques for 64-row MDCT and MR for the study of these anomalies.

Background

The embryogenesis of the vena cava (VC) is a complex process involving the formation of several anastomoses between embryonic veins. The result is numerous variations in the normal anatomy of the VC in thorax, abdomen and pelvis.

Since the development of cross-sectional imaging, congenital anomalies of the VC and its tributaries have become more frequently encountered in asymptomatic patients. Vascular structures are usually identified on computed tomographic (CT) scans of thorax, abdomen and pelvis obtained with intravenously administered contrast material. In addition, with helical acquisition, the venous structures may be imaged during different phases, when little, much or no contrast material is present in the veins.

Therefore, familiarity with these variations is essential for correct interpretation of cross-sectional images, to avoid erroneous diagnosis of retroperitoneal and mediastinal masses or adenopathies, and to alert surgeons and angiographers of the abnormal venous anatomy in order to avoid complications related to the procedures.

In this work, we review the embryogenesis of the VC and we describe the congenital variations in VC anatomy that were retrospectively found in thoracic or abdominal MDCT and MR studies, performed in 330 patients with or without suspected vascular disease.

MDCT and MR imaging appearances of the more frequently encountered anomalies and some unusual variations are presented. In addition, the clinical relevance of the variations is discussed.
Imaging findings OR Procedure details

We have retrospectively reviewed 331 64-MDCT or 1.5 T MR angiographic studies of thorax and abdomen performed at our department between 2006 and 2012. Among them, 32 patients with congenital anomalies of VC in thorax and abdomen were found.

Eleven patients were male and 21 were female. Most of them were clinically suspicious of venous disease (history of abdominal discomfort, swelling/pain of lower extremities, varicosities or collateral circulation); some others, however, were incidental findings of studies required by different clinical disorders.

For MDCT, scanning was performed during a single breath hold in cooperative patients. In all cases, we used a 64-row MDCT which may allow 1-3 mm sections thickness to be made after the study acquisition. After data are acquired, they were transferred to our 3D workstation for multiplanar reformatting shaded surface display, maximum intensity projection, minimum intensity projection, volume rendering. Our software allows real-time manipulation and editing of the volume, which is essential for displaying the vessels in the proper orientation.

For angiographic studies of MDCT and MR, optimal opacification of the veins requires rapid intravenous administration of a bolus of contrast agent as well as precise timing of the data acquisition. Thus, depending of the clinical suspicion, we used three different methods:

1.-) **Patients with low suspicion of venous anomalies/incidental findings** (unspecific discomfort in abdomen, pelvis or lower extremities; unspecific abdominal doppler-ultrasound findings, other clinical indications): We routinely use 100-120 mL of nonionic contrast material injected through a peripheral IV catheter by the upper extremity at a rate of 2-2.5 mL/sec. Scanning is performed during the systemic venous phase at 50-60 seconds after the start of injection.

2.-) **Patients with suspicion of iliocaval venous occlusion or May-Thurner syndrome** (chronic venous insufficiency of lower extremities, varicosities, well documented pathological doppler-ultrasound): we routinely use 100-120 mL of nonionic contrast material injected through a peripheral IV catheter by the lower extremity at a rate of 2.5-3 mL/sec. Scanning is performed during the iliac venous phase at 10-12 seconds after the start of injection and at 50-60 seconds after venous injection; compression over the knee and the groin is previously applied, for increasing the blood flow towards deep venous system.
3.-) **Patients with suspicion of Nut-Cracker syndrome** (pelvic discomfort, or abnormal abdominal doppler-ultrasound), specially in young female patients: we also use 100-120 mL of nonionic contrast material injected through a peripheral IV catheter by the upper extremity at a rate of 2.5-3 mL/sec, but scanning is previously performed during arterial phase at 20-30 seconds after the start of injection, to obtain earlier opacification of both renal veins.

For MR angiographic examinations, we routinely used:

a.-) **Precontrast studies:**
- FFE (BALANCE) sequences on the three planes (axial, sagittal, and coronal)
- Single- Shot T2 sequences on both axial and coronal planes,
- FFE- T1 (IN/OUT phase) sequences on axial plane,
Including the level of both renal veins and iliac bifurcation if pelvic study.

b.-) **Postcontrast studies:**
- We obtain dynamic 3D FFE-T1 sequences after administration of mL of contrast material (gadolinium) injected through a peripheral IV catheter by the upper extremity at a rate of mL/sec. Different phases: basal, arterial, venous, and delayed after the start of injection, are obtained.

In our patients, we routinely used 100-120 mL of nonionic contrast material injected through a peripheral IV catheter at a rate of 2.5-3 mL/sec.

This way, a great variety of congenital venous anomalies were found. They were classified by the involved VC segment and the type of anomaly, like complete/partial agenesis, duplicity, abnormal position, or others. In summary, we found the following cases:

1. **ANOMALIES OF SUPERIOR VENA CAVA (SVC):**

Venous anomalies of the thorax are the result of complex variations in the persistence and regression of segments of three sets of veins during the first 2 months of fetal development: the umbilical, vitelline and cardinal venous systems. Normally, the right anterior cardinal and common cardinal veins form the SVC, and the left anterior cardinal vein regresses.
Thus, the most common reported anomalies of SVC are:

1.1. **Duplicated SVC:**
A double SVC is the result of persistence of the left anterior cardinal vein. A persistent left SVC is an incidental finding in less than 0.5% of the general population but occurs in approximately 4% of patients with congenital heart disease.

A left SVC is occasionally detectable, usually in retrospect, as a focal widening of the mediastinum superior to the left side of the aortic knob on a chest radiograph. Most often, however, a left SVC or left component of a duplicated SVC is revealed by radiography when traversed by an IV catheter.

The left brachiocephalic vein is absent, and the right SVC is smaller than the left in 65% of SVC duplications. In addition, it can be connected or not by brachiocephalic vein. We report two cases of duplicated SVC (FIGs 1-2).

1.2. Persistent Left SVC (PLSVC):

It occurs when the normally persistent right cardinal vein regresses.

A left SVC is most often an incidental finding that is not clinically significant. But it can be incidentally discovered during central venous line placement, intracardiac electrode/pacemaker placement or cardiopulmonary bypass, where it may cause technical difficulties and life-threatening complications.

As in the previous case, left SVC is occasionally detectable, usually in retrospect, as a focal widening of the mediastinum superior to the left side of the aortic knob on a chest radiograph. With a solitary left SVC, the ascending aorta can appear unusually prominent on radiographs because no right SVC obscures it. This circumstance can simulate an abnormal aorta.

Drainage to the left atrium is associated with many types of congenital heart disease, but is rare if the heart is normal. When these circumstances are unrecognized, ligation of the left SVC as part of a cardiac surgical procedure has led to acute coronary venous hypertension and myocardial ischemia. For this reason, we think it is important for radiologists to be able to recognize these incidental abnormalities in order to avoid potential complications.

1.3. Isolated anomalies of Right SVC:

Isolated anomalies of the right SVC are rare. A right SVC can insert low into the right atrium, drain to the left atrium, or be congenitally dilated. On radiography, a congenitally dilated SVC has the appearance of a mediastinal mass. This anomaly is usually an
incidental finding but has been associated with thrombosis leading to embolization and SVC obstruction.

2. ANOMALIES OF INFERIOR VENA CAVA (IVC):

Congenital anomalies of the IVC have become more commonly recognized in asymptomatic patients as incidental findings or unsuspected cases. Thus, specific angiographic studies may be not performed. However, their correct identification is useful in the planning of vascular interventions and prevents their being mistaken for disease. Several congenital anomalies of the IVC are associated with variations of the azygos and hemiazygos anatomy within the retrocrural space.

2.1. AGENESIA OF IVC:

2.1.1. Absent of the hepatic segment of the IVC with azygos continuation, is probably the most well studied of these anomalies with a prevalence of 0.6%. Although previously associated with many other congenital anomalies, it has been increasingly identified in asymptomatic patients. Failure to develop a communication between the right subcardinal and hepatic venous system during embryogenesis, has been postulated to cause interruption of the hepatic segment of the IVC and loss of continuity with the prerrenal IVC. It thus results in shunting of the venous blood from the lower abdomen, pelvis and lower extremities into the azygos or hemiazygos venous system, accessing to the thorax to drain into the superior vena cava. (FIGs.3-4).

2.1.2. Partial absence of the suprarrenal segment of the VCI with preservation of intrahepatic and infrarrenal segments is much uncommon. We describe the case of a middle-age male patient with segmentary absence of the suprarrenal portion of the VCI, and preservation of the hepatic and infrarrenal segments. In addition, stenosis of VCI at the level of hiatus is also present. (FIG. 5).

2.1.3. Complete absence of the infrarrenal IVC:

Complete absence of the infrarrenal IVC with preservation of the suprarrenal segment is a very rare anomaly that suggests that all three paired venous systems failed to develop properly. This entity is very uncommon and results in enlarged retrocrural azigos and hemiazygos veins to drain into the thorax.

There is controversy as whether this condition is the result of perinatal thrombosis of the IVC or a truly embryologic origin, consisting in failure of development of the posterior
cardinal and supracardinal veins. This entity also results in enlarged retrocrural azigos and hemiazygos veins to drain into the thorax.

We show one case of infrarrenal agenesia of VCI with preservation of both iliac veins (FIG.6), and two cases in which both common iliac veins were also absent where venous drainage was made by the gonadal veins, parietal veins, lumbar veins, hemiazygos and inferior mesenteric vein (FIG. 7-8).

Affected patients are prone to develop deep venous thrombosis and chronic venous insufficiency. Lower-extremity venous return in these patients also may occur via the ascending lumbar veins, which drain into the azygous-hemiazygous system. Enlarged collateral vessels may simulate a paraespinale mass.

2.1.4. **Partial/segmentary absence of the infrarrenal IVC:**

It is a very rare anomaly in which only a short segment of VCI is absent, with distal repermeabilization by collateral pathways of retroperitoneal, parietal and paraespinale veins to join a normal infrarrenal VCI. We present one case of a middle-age man with partial atresia of VCI at the origin. (FIG. 9).

2.2. **LEFT IVC:**

Left inferior vena cava results from regression of the right supracardinal vein with persistence of the left supracardinal vein, resulting in a mirror image variant. The average prevalence is 0.2%-0.5 %.

2.2.1. Typically, the **left IVC ends at the left renal vein**, which crosses anterior to the aorta to form a normal right-sided IVC by the union with the right renal one. Thus, diaphragmatic hiatus of VCI is correctly located to the right of both esophageal and aortic hiatus. This was a very common incidental finding we found in our retrospective review (eight patients) (FIGs.10-11).

2.2.2. "**Complete left VCI**" has been less frequently reported in the literature. In this case, the VCI continues with an enlarged hemiazygos vein what crosses through the diaphragm into the thorax to the left of the aortic hiatus. Associated situs anomalies are present in many of these cases.

It is important not to misinterpret the enlarged azygous or hemiazygous veins as lymphadenopathy. Preoperative knowledge of this anomaly is also important in planning cardiopulmonary bypass surgery and can help avoid difficulties in cardiac catheterization.
We present a case of a young asymptomatic patient with "complete" left IVC crossing through diaphragm to the left of the aorta. (FIG. 12).

2.3. DOUBLE IVC:

Duplication of IVC results from persistence of both supracardinal veins. The prevalence is 0.2 -3%.

2.3.1. In double IVC, the left IVC typically ends at the left renal vein, which crosses anterior to the aorta to join the right IVC. There may be significant discrepancy in the size of the two veins. However, there may be variations, such as double IVC with retroaortic right renal vein and hemiazygos continuation of the IVC. In our review, eight patients in which double IVC typically ends at the left renal vein, were found. (FIGs 13-16).

Double IVC has similar clinical implications to those of left IVC and may be mistaken for lymphadenopathy, especially if contrast enhancement of the vein is poor due to technical reasons or thrombosis. Double IVC should be suspected in cases of recurrent episodes of pulmonary embolism despite placement of an IVC filter.

2.3.2. Partial double infrarenal VCI:

This variety is extremely rare, consisting of partial regression of accessory supracardinal left renal vein. We present one case with 3D volumetric reconstruction of a young woman with partial preservation of infrarenal left VCI. (FIG. 17).

2.4. STENOSIS/COMPRESSION OF VCI AT DIAPHRAGMATIC HIATUS:

The lumbar portion of the peripheral diaphragm attaches to the medial and lateral lumbocostal arches (arcuate ligaments) and to the anterolateral surfaces of the lumbar vertebrae as bilateral musculotendinous pillars, known as the diaphragmatic crura. The right one is longer and broader than the left.

A fasciculus of the medial aspect of the left crura crosses the aorta ventrally and runs along the lateral deeper fibers of the right one, toward the vena caval hiatus. The inferior vena cava pierces the diaphragm at T9 through the vena caval foramen in the central tendon of the diaphragm.

We have reviewed four cases for which compression of IVC at the level of the hiatus had clinical repercussion, consisting in discomfort and tenderness or inflammation of lower extremities. (FIGs. 18-20).
Fig. 1: Axial views of double VCS on contrast enhanced MDCT scan and Volume-rendering 3D reconstruction; c-d: sagittal views of both VCS after 3D Volume Rendering post processing; e-f: coronal views of both VCS on MDCT scan and Volume-rendering 3D reconstruction; g: axial view at the level of suprahepatic veins draining separately both hepatic lobes into two different IVC.
**Fig. 2:** Axial views of double VCS after contrast enhanced MDCT scan and Volume-rendering 3D reconstruction at the level of pulmonary trunk; c-f: coronal views of both VCS on MDCT scan and 3D MIP and Volume-rendering reconstruction.
Fig. 3: CT axial images (a-c) and 3D MIP and volumetric reconstructions (d-f), show interruption of the hepatic VCI, with abnormal suprahepatic anatomy and large collateral vessels at the level of the renal hilum and along thoracic and abdominal wall; Infrarrenal VCI connects with an enlarged azygos system to drain the blood from the lower extremities, pelvis and lower abdomen into the thorax. Bilateral iliac thrombosis was confirmed
Fig. 4: Contrast-enhanced CT axial images (a-b) and 3D MIP and volumetric reconstructions (c-f) show interruption of the hepatic VCI with abnormal suprahepatic anatomy and collateral drainage into the thorax by an enlarged azygos system (arrow).
Fig. 5: Contrast-enhanced CT axial and coronal images (a, c), and 3D MIP and Volumetric reconstructions (b, d), show atresia of the infrahepatic segment of VCI with normal preservation of both intrahepatic and infrarrenal venous anatomy. Moderate grade of stenosis at the level of vena cava hiatus is also seen (arrows).
Fig. 14: Coronal views on 3D Volume rendering and MIP reconstructions, (c) sagittal view and (d) axial view of other patient with infrarrenal double VCI
Fig. 15: Coronal and axial views on a venous phase scan show double infrarenal VCI; c-d, 3D Volume rendering coronal views of the same patient, show the left VCI crossing to the right to join the normal righted-sided VCI.
Fig. 16: Coronal views on contrast-enhanced T1-weighted and T2-weighted sequences, showing double left VCI ascending to join left renal vein; (c) axial SSHOT T2-weighted sequence at infrarrenal level; (d), dynamic post-contrast acquisition on venous phase showing double infrarrenal VCI
Fig. 17: Coronal views on 3D Volume rendering and MIP acquisitions of MDCT scan after contrast administration, showing partial infrarrenal double VCI due to partial preservation of an accessory left supracardinal vein. See two left renal veins draining separately, into the accessory left VCI the lower one and into the normal right-sided VCI at the level of right renal hilum, the upper one
Fig. 18: 3D MIP and Volume rendering sagittal views (a, b), axial view of the IVC at the level of diaphragmatic hiatus, and coronal view on 3D MIP reconstruction (d) showing pathological narrowing of the involved portion of IVC.
Fig. 19: 3D Volume rendering images on coronal, sagittal and axial planes at the level of diaphragmatic hiatus, in another patient with stenosis of VCI
**Fig. 13:** Axial and (b-d) coronal views of infrarenal double VCI, on MDCT scan and 3D MIP and Volume Rendering images, where the left VCI ascends to join the left renal vein.
Fig. 12: Axial and coronal views in a patient with "complete left VCI" passing through diaphragm to the left of the aortic hiatus. See the retroaortic right renal vein crossing to the left to join the VCI. E-f, 3D Volume Rendering images on coronal plane of the left VCI, with a retroaortic right renal vein.
Fig. 11: Axial views of infrarrenal left IVC with a stent crossing anterior to the aorta. C-d, 3D MIP and Volume Rendering images show infrarrenal VCI crossing towards right-sided normal position with the stents at the level of the crossing and at the confluence of both common iliac veins, in a patient with previous history of deep vein thrombosis of iliofemoral veins.
Fig. 10: Axial and coronal views show infrarrenal left IVC crossing anteriorly to the aorta at the level of renal hilum to form a normal right-sided suprarrenal IVC by the union with the right renal one. D, 3D Volume Rendering image of the same patient shows the infrarrenal VCI crossing anteriorly to the aorta to reach the normal right-sided VCI.
Fig. 9: Axial view of the absent VCI at the origin, showing collateral venous drainage through retroperitoneal veins (iliopsoas muscle and paraespinical space) and right abdominal wall; axial view at a proximal infrarrenal level where VCI is present; 3D Volume Rendering images show atresia of VCI at its origin with the collateral pathways. Both iliac veins, however, are present.
**Fig. 8:** Coronal view (a) shows VCI interruption: suprarrenal segment of IVC is present; (b) axial view at the level of iliac bifurcation; (c-d) 3D MIP and Volume Rendering images of the collateral pathways along both enlarged gonadal veins in a patient with absence of VCI and common iliac veins. Genitourinary congenital anomalies (agenesia of uterus) are also seen.
Fig. 7: Axial view at the level of renal hilum, 3D MIP and Volume Rendering images of enlarged and prominent collateral pathways along both gonadal veins and inferior mesenteric vein in a patient with absence of VCI and common iliac veins. Suprarrenal segment of IVC is present.
**Fig. 6:** Contrast-enhanced CT axial image below the level of renal hilum (a) and 3D MIP and Volumetric reconstructions on coronal views (b, c), show absence of infrarenal VCI, with normal preservation of suprarenal segment, formed by the confluence of both renal veins. Common iliac veins are present.
Fig. 20: 3D Axial and coronal views on FFE T2-weighted sequences of MR studies performed after inspiration and expiration at the level of diaphragmatic hiatus, show mild oscillation on the narrowing of the involved segment of VCI with respiratory movements.
Conclusion

The complexity of embryology of the VC can lead to a wide spectrum of variations in the basic plan of venous return from thorax, abdomen and veins of extremities.

Although vascular structures can usually be easily identified on contrast-enhanced MDCT scans, identification of these unusual variations may be difficult in those cases in which intravenous contrast material or RX are contraindicated (such as pregnant women, children, patients with renal insufficiency). In such cases, MR vascular imaging may be used to identify and distinguish those aberrant vessels from masses or adenopathies by demonstrating flow voids or flow-related enhancement.

Many of those anomalies may have significant clinical implications. Thus, MDCT and MR angiography studies, with MIP and Volume-rendering 3D reconstructions for thoracic and abdominal veins can play an important role to identify unsuspected (or suspected) VC anomalies, mainly when abdominal discomfort, swelling/varicosities in extremities, deep vein thrombosis or pulmonary thromboembolism as related complications, are present.

Finally, it is important for radiologists to recognize the spectrum of those congenital anomalies in order to avoid possible diagnostic pitfalls, to plan an early endovascular or surgical treatment if necessary, and prevent potential severe cardiovascular complications in unsuspected patients.

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