Developmental Anomalies of the Inferior Vena Cava and its Tributaries: What the Radiologist Needs to Know?

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Abstract

Inferior vena cava (IVC) can be involved by a wide gamut of developmental anomalies owing to its complex embryogenesis. Developmental anomalies of the IVC are not infrequent, seen in approximately 8.7% of the general population. Although most of the anatomical variations are asymptomatic, identification of these variations is important before planning any vascular surgery or interventional procedure in relation to the IVC to avoid inadvertent complications. Conventional venography has largely been replaced by noninvasive cross-sectional imaging modalities for detecting IVC abnormalities. Ultrasonography, often used for initial evaluation, is highly operator-dependent and the infrarenal part of IVC is often obscured by bowel gases. While magnetic resonance imaging is devoid of radiation risks, its use is limited due to limited availability and the frequent need for sedation. Computed tomography (CT) venography plays a pivotal role in the detection of these anomalies as it has an excellent spatial resolution along with availability of multiple postprocessing tools such as multiplanar reconstruction with generation of maximum intensity projection and volume-rendered images. This pictorial review focuses on the embryogenesis of IVC, various developmental anomalies of the IVC and its tributaries, their appearance on CT venography and conceivable clinical relevance.

Keywords
► computed tomography
► developmental anomalies
► inferior vena cava

Introduction

Inferior vena cava (IVC) is a venous trunk, formed by the confluence of right and left common iliac veins, which drains into the right atrium. The tributaries of IVC include the lumbar veins, bilateral renal veins, right adrenal vein, right gonadal veins, and the hepatic veins.1 It courses to the right of aorta in the retroperitoneum and ascends into thorax across the diaphragm, through an aperture in the central tendon, at the level of D8 vertebra. Developmental anomalies of IVC are not infrequent, existing in approximately 8.7% of the general population.2 Knowledge of these variations is important before planning any vascular surgery or interventional procedure in relation to the IVC to avoid potential complications.
Role of Imaging

Conventional venography, once the gold standard, has largely been replaced by noninvasive cross-sectional imaging modalities for detecting IVC abnormalities. Ultrasonography is often used for initial evaluation; however, it is an operator-dependent modality and scanning of the infrarenal part of IVC is suboptimal due to obscuration of IVC by bowel gases. Orthogonal techniques such as computed tomography (CT) or magnetic resonance (MR) venography are commonly used for imaging of IVC and characterization of its variants. The advantages of CT include excellent spatial resolution and availability of postprocessing tools such as multiplanar reconstruction with generation of maximum intensity projection and volume-rendered images. With the introduction of advanced dose reduction techniques, the required information can be acquired at very low doses. Routine imaging done at 60 to 70 seconds (portal-venous phase) after intravenous contrast administration depicts the renal and supra-renal IVC with contrast admixture in the infrarenal part. Delayed imaging done at 70 to 90 seconds provides optimal enhancement of the entire IVC. At our institute, CT venography (CTV) for IVC is performed on a dual-source CT scanner after administration of 1.5 to 2 mL/kg contrast material via a peripheral vein at the rates varying from 1 to 4 mL/s. The optimal delay for scanning (at 60 seconds, 80 seconds, or both) is decided on a case-to-case basis.

MR venography (MRV) is a useful modality for detecting the developmental IVC anomalies. It can be performed by both noncontrast and contrast-enhanced techniques. It has an advantage over CTV in terms of lack of radiation as well as lack of contrast requirement while planning noncontrast scans. While the contrast-enhanced MRV remains the workhorse, the noncontrast techniques can be used standalone when there is a contraindication to contrast administration. In the contrast-enhanced scans, mask subtraction of enhanced and unenhanced data generates maximum intensity projection images without background soft tissue signal and with good depiction of venous vasculature and variants. Time-resolved MR angiography is also a promising technique to separately depict arterial and venous phases and venous flow. The various noncontrast sequences for MRV include balanced steady state free precession, quiescent interval single shot, time of flight (TOF), and three-dimensional turbo spin echo techniques. During preoperative imaging in children with congenital heart diseases or those scheduled to receive transplant, a fast multistation TOF protocol can also be used, as it allows avoidance of anesthesia. Although MR is devoid of radiation risks, its use is limited due to limited availability and the frequent need for sedation, especially in pediatric patients.

Relevant Embryology

The development of IVC is a complex sequential process occurring during the fourth to eighth week of gestation. It involves three primitive pairs of venous channels, namely, the posterior cardinal vein, the subcardinal vein, and the supracardinal vein (lumbar and thoracic parts) which appear and are replaced in order (Fig. 1). A wide gamut of anomalies occur due to defect in the appearance or regression of these embryonic venous structures.

Development of Posterior Cardinal Veins

In the early stages of development, the paired posterior cardinal veins drain the caudal aspect of the embryo which are connected by interpostcardinal anastomosis. Meanwhile, the paired anterior cardinal veins drain the cranial aspect of the embryo and ultimately form the venous drainage of the head and neck region. Blood from the visceral organs is drained by the vitelline vein.

Fig. 1 Embryological development of the inferior vena cava.
Development of Subcardinal Veins
Subsequently, there is formation of paired subcardinal veins on the ventromedial aspect of posterior cardinal veins, anterior to the aorta, which are connected by intersubcardinal anastomosis. There is development of anastomosis between the posterior cardinal and subcardinal veins (postsbcardinal anastomosis). Later the drainage of the caudal embryo occurs via the subcardinal vein with the regression of posterior cardinal vein. The cranial right subcardinal vein forms the suprarenal IVC and intersubcardinal anastomosis forms the left renal vein. The connection develops between cranial subcardinal vein and hepatic part of IVC (formed from vitelline vein) via subcardinal–hepatic anastomosis. 

Development of Supracardinal Veins
Further, there is formation of paired supracardinal veins on the dorsomedial aspect of the posterior cardinal veins and posterior to aorta. A complex suprasubcardinal and intersupracardinal anastomosis also develops. The supracardinal veins divide into the cranial (azygous) and the caudal (lumbar) part. The final venous drainage of caudal embryo occurs via the supracardinal veins with the right supracardinal vein forming the infrarenal IVC. There is subsequent shunting of blood from left to right via the complex anastomosis and regression of left-sided structures. The azygous and hemiazygous veins are derived from supracardinal veins in the thoracic region.

Development of Renal Veins
The renal collar consists of a ventral arch (intersubcardinal anastomosis) and dorsal arch (intersupracardinal anastomosis). The embryonic kidney is drained via the anterior and posterior limbs. There is regression of the posterior limb. The right-sided anterior limb gets incorporated into the IVC wall and on the left side forms the left renal vein.

► Table 1 summarises the embryology of the various vascular structures discussed above.

Anomalies of IVC and Its Tributaries
There is no definite classification of IVC variations. In 1920, Huntington and McClure studied IVC development in domestic cats and proposed that there could be 14 possible variations.

Anomalies of IVC Proper
Absent IVC
Congenital absence of IVC is rare and includes two variations as follows:

- Absent infrarenal IVC with normal suprarenal part.
- Complete absence of entire IVC. It occurs due to failure of development of all three primitive veins (posterior cardinal, subcardinal, and supracardinal veins).

This congenital variation is usually detected incidentally and the patients remain asymptomatic because of the development of collateral vessels which may become aneurysmal at times and mimic paraspinal masses. Absence of part or all of IVC is associated with lower limb deep venous thrombosis (DVT) and chronic venous insufficiency (varices) due to blood stasis because of insufficient collateralization. DVT most commonly involves iliac veins with propensity for bilateral and recurrent involvement. Absent IVC should be suspected in a young patient with DVT without any predisposing factor or clotting defects.

Persistent Left-Sided IVC

Duplicated IVC
A double IVC occurs in 1 to 3% of the population and is caused due to persistent right and left supracardinal veins. The left-sided IVC usually crosses at the level of left renal vein to join right IVC (►Fig. 2). However, many variations have been described in this arrangement:

- Asymmetrical sizes of both IVC (►Fig. 3).
- Double IVC with retro-aortic right renal vein and hemiazygous continuation of IVC.

This variation occurs due to persistent left lumbar and thoracic supracardinal veins and left suprasubcardinal

<table>
<thead>
<tr>
<th>Sr. No.</th>
<th>Vascular structure</th>
<th>Derived from</th>
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<tbody>
<tr>
<td>1</td>
<td>Hepatic IVC</td>
<td>Vitelline vein</td>
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<tr>
<td>2</td>
<td>Suprarenal IVC</td>
<td>Right subcardinal vein and subcardinal–hepatic anastomosis</td>
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<td>3</td>
<td>Renal IVC</td>
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<td>4</td>
<td>Infrarenal IVC</td>
<td>Right supracardinal vein</td>
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<tr>
<td>5</td>
<td>Left renal vein</td>
<td>Intersubcardinal anastomosis</td>
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<tr>
<td>6</td>
<td>Common iliac veins</td>
<td>Posterior cardinal veins</td>
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<tr>
<td>7</td>
<td>Azygous and hemiazygous veins</td>
<td>Thoracic supracardinal veins</td>
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Abbreviation: IVC, inferior vena cava.
Fig. 2  Coronal maximum intensity projection image (A) and volume-rendered image (B) showing presence of bilateral inferior vena cava (IVC). The left IVC ends at the level of the left renal vein (‘C3’ in (B)) crossing to the right and draining into the right IVC.

Fig. 3  Coronal maximum intensity projection image (A) and volume-rendered image (B) showing presence of bilateral inferior vena cava (IVC) with asymmetrical sizes of both IVCs. A communication (‘*’) between the two IVCs is seen at the caudal end.
anastomosis with nonformation of right subcardinal–hepatic anastomosis. In addition, the dorsal limb of renal collar persists with regression of the ventral limb. Right IVC and right renal vein meet and course posterior to aorta to join the left IVC, which continues as a hemiazygous vein. The hemiazygous vein can join the azygous vein, drain into coronary sinus via persistent left superior vena cava or drain into left brachiocephalic vein via an accessory hemiazygous vein.

- Double IVC with retroaortic left renal vein and azygous continuation of IVC.²

It occurs due to persistence of left supracardinal vein and dorsal limb of renal collar with failed development of ventral limb and right subcardinal–hepatic anastomosis.

This anatomical variation has many clinical implications. It can be misinterpreted as mass or lymph node on imaging, if there is poor contrast enhancement. This anomaly should be ruled out in case of recurrent pulmonary emboli despite adequate anticoagulation and IVC filter placement.¹³

**Isolated Left-Sided IVC**

An isolated left-sided IVC has a prevalence of 0.2 to 0.5%.⁹ It occurs due to persistent left supracardinal vein with regression of the right supracardinal vein. At the level of renal hilum, the left IVC courses anterior or posterior to aorta to occupy its normal position on the right side.¹ It can be misdiagnosed as a lymph node. Chronic mesenteric ischemia has been reported in the literature due to compression of celiac trunk when left-sided IVC crosses anterior to the aorta.¹⁴ In the presence of left-sided IVC, the transjugular access for filter placement is difficult.⁹

**Interruption of Intrahepatic IVC**

This variant occurs in 0.6% of the population⁸ and results from failure of formation of subcardinal–hepatic anastomosis and resultant atrophy of right subcardinal vein.¹⁵

**Azygous Continuation**

Due to interruption of IVC between the suprarenal and hepatocardiac segments, the blood from infrarenal IVC is shunted via suprasubcardinal anastomosis into the dilated retrocraural azygous vein with drainage into the superior vena cava. Hepatic veins are drained into the right atrium via the posthepatic segment (► Fig. 4). The common associations of this anomaly include heterotaxy syndromes (left isomerism) and intestinal malrotation.¹⁵ It is imperative to be aware of this variation before planning cardiopulmonary bypass surgeries.⁹ An enlarged azygous vein can be misinterpreted as lymphadenopathy on imaging.

**Hemiazygous Continuation**

The drainage of left-sided IVC via the hemiazygous vein can have the following variations in drainage:

- Hemiazygous vein → azygous vein → right superior vena cava.
- Via accessory hemiazygous vein → persistent left superior vena cava → coronary sinus (► Fig. 5).
- Via accessory hemiazygous → left superior intercostal vein → left brachiocephalic vein.¹⁶

**High Right Atrial Insertion**

Anomalous high insertion of IVC is extremely rare with very few cases published in the literature. Most of the cases are associated with cardiac diseases but can also occur as an isolated anomaly.¹⁷
Anomalous Drainage of IVC to Left Atriums

During embryogenesis, there is communication of sinus venosus with the primitive atrium by right and left valves. In normal development, the sinus venosus shifts to the right, the left valve disappears, and the right valve atrophies. In case the right valve persists, it fuses with septum secundum and drainage of IVC occurs into the left atrium. Half of the cases are associated with atrial septal defect with other associated abnormalities being anomalies of pulmonary drainage and pulmonary arteriovenous fistula. Although the individuals with this anomaly are usually asymptomatic, they can present with cyanosis, dyspnea, and features of systemic embolism. On imaging, IVC has a normal position in lower chest but then curves to drain into the left atrium. This variation can be misinterpreted as inferior sinus type of atrial septal defect. Due to this variation, problems can be encountered to gain access to right atrium by the femoral route.

IVC Webs

Occurrence of an IVC web is rare and most commonly occurs in Asian and South African countries. It can occur in the form of membrane or a fibrous occlusion of variable length. The membrane can be complete or fenestrated and almost always occurs in proximity to the IVC drainage into the right atrium or infradiaphragmatic level (Fig. 6). Prominent collaterals develop in intrahepatic and extrahepatic regions. The origin of this anomaly has been proposed to result from congenital vascular malformation or organized thrombus. Clinically, the web leads to hepatic venous outflow obstruction causing congenital Budd–Chiari syndrome.

IVC Aneurysm

IVC aneurysm is a rare abnormality with potential clinical significance. Predisposing factors include trauma, inflammation, and uncontrolled hypertension. Gradman and Steinburg have proposed the following classification system:

- Type I: suprahepatic IVC aneurysm with no obstruction.
- Type II: IVC aneurysm above and below the hepatic veins.
- Type III: infrarenal IVC aneurysm without any anomaly.
- Type IV: miscellaneous.

Treatment of IVC aneurysms is recommended due to potential complications as thrombosis and pulmonary embolism. Surgical correction is the treatment of choice. However, medical management is preferred in asymptomatic type I aneurysms. An endovascular stent graft could be an alternate option.

Anomalies of Tributaries

Anomalous Left Renal Vein

Variations of left renal vein are more common than right owing to its longer length and complex embryogenesis. Retroaortic left renal vein is present in 1.7 to 3.4% of the population and the prevalence of circumaortic vein is 2.4 to 8.7%. Knowledge of renal vein anomalies is imperative for surgeons and interventionists before performing renal vein
catheterizations, nephrectomy, left renal transplantation, or creation of splenorenal shunt. The classification of congenital anomalies of left renal vein is as follows:

- **Type I**: single left renal vein bifurcating into periaortic and retroaortic branches.
- **Type II**: two left renal veins: one preaortic and other retroaortic.
- **Type III**: characterized by anastomosis between preaortic and retroaortic veins or occurrence of multiple preaortic or retroaortic veins without anastomosis.

### Retroaortic Left Renal Vein

It occurs due to persistent intersupracardinal anastomosis with involution of intersubcardinal anastomosis with the left-sided renal vein coursing posterior to aorta. Due to this anatomical variation, the left renal vein can get compressed between aorta and vertebral body (posterior nut cracker syndrome) causing flank pain and hematuria (Fig. 7). There can be associated varicoceles and pelvic varices. In the presence of symptoms, surgical correction is needed.

Fig. 6 Sagittal (A) and coronal (B) images showing discrete significant stenosis of the suprahepatic part of the inferior vena cava (IVC) with a calcific speck at the level of the stenosis. Axial image (C) shows downstream dilatation of the IVC. Volume-rendered image (D) depicts the extensive collateralization over the chest and abdominal walls.

Fig. 7 Oblique axial (A) and coronal (B) images demonstrate the retroaortic course of the left renal vein (arrow heads). AA, abdominal aorta.
**Circumaortic Left Renal Vein**
It occurs due to persistent intersupracardinal as well as intersubcardinal anastomosis resulting in the formation of two left renal veins: the superior one receiving the left suprarenal vein with a course anterior to aorta and the inferior one receiving left gonadal vein and coursing posterior to the aorta to drain into the inferior vena cava (IVC). (Fig. 8).

**Abernethy Malformation**
Abernethy malformation also called as congenital extrahepatic portosystemic shunt (CEPS) occurs rarely and is characterized by shunting of portal blood to the systemic circulation by passing the liver via a complete or partial shunt. The most common drainage site is the IVC. As the nutrients as well as toxins are diverted from the liver to the systemic circulation, it results in varied clinical presentations as liver dysfunction, hepatic encephalopathy, and hepato-pulmonary syndrome.

Embryologically, the portal vein (PV) develops from the right and left vitelline vein and intervitelline anastomosis around the duodenum. The selective regression of the anastomotic channels forms PV. Type 1 CEPS occurs due to excessive involution of the periduodenal venous channel with resultant shunt due to persistent right or left vitelline vein. Type 2 CEPS occurs due to persistent primitive anastomosis between the vitelline and subcardinal veins. Morgan and Superina have classified CEPS as follows:

- Type 1: end to side anastomosis with absence of intrahepatic part of PV. Splenic vein and superior mesenteric vein can drain separately (type 1a) or via a common trunk (type 1b) (Fig. 9).
- Type 2: intrahepatic portal venous radicles are present with partial diversion of the blood through a side-to-side anastomosis (Fig. 10).

This anomaly especially type 1 is associated with multitude of congenital anomalies and heart defects. Hepatic
ischemia and subsequent compensatory increase in arterial flow to the liver lead to increased incidence of hepatic nodular lesions. Liver transplantation is the only effective treatment in cases of symptomatic type 1 CEPS, whereas type 2 CEPS can be managed with surgical closure or coil embolization.

Interventional Radiology and Developmental Anomalies of IVC

The knowledge of congenital anomalies of IVC and renal veins is essential before planning any interventional procedure such as IVC filter placement. In cases where duplicated IVC is present, filter may be placed either in both IVCs or in the suprarenal location. If the other IVC is a small accessory cava which communicates with the main IVC at both common iliac vein and renal vein levels, coil embolization of the smaller IVC followed by placement of the filter in the dominant IVC may be considered. In cases of circumaortic left renal vein, there are chances of embolization via the venous ring if the filter is placed between two vein orifices, so it must be placed below the retrocaval component of the ring, which usually drains at a lower level in the IVC. Similarly, if the retroaortic left renal vein is present, a filter must be placed below its orifice. In patients with short-segment IVC obstruction causing Budd–Chiari syndrome, IVC angioplasty and stenting is an effective alternative to surgical portosystemic shunts with satisfactory long-term patency. Also, single or multisession endovascular shunt closure can be done in cases of type 2 Abernethy malformation.

Conclusion

IVC can be involved by a gamut of developmental anomalies owing to its complex embryogenesis. Although most of the anatomical variations are asymptomatic, their identification is of utmost importance before planning surgeries and any interventional procedure to avoid inadvertent complications. CTV has a pivotal role in the detection of such anomalies.
Especially with the advent of dual-source scanners and dose-reduction techniques, diagnosis can be made at very low doses.

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Conflict of Interests
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