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Pictorial review of congenital anomalies of the inferior vena cava and its tributaries

Revisión iconográfica de las anomalías congénitas de la vena cava inferior y sus tributarias

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Abstract

The anomalies of the inferior vena cava (IVC) represent a broad group of entities that are not uncommon, mostly being asymptomatic. Their development has its starting point in embryogenesis, representing a complex sequence of processes that occur during the fourth to eighth week of gestation. This process involves complex anastomoses and regressions of embryonic veins. The IVC is the main venous return structure for the extremities and abdominal organs. Proper identification of the IVC is often crucial, as it has direct clinical and pathological implications. Additionally, some anomalies will require a different approach in the planning of vascular interventions, while recognizing others will be useful to differentiate them from pathological conditions. With current multislice tomography, high-resolution spatial images can be acquired, allowing for high-quality reconstructions and highly detailed representations of the anatomy. There is no definitive classification for IVC anomalies. There are numerous possibilities, including agenesis, duplication, left-sided location, interruption of the intrahepatic portion, and Abernethy malformation. There are also anomalies of the tributary veins. In this article, we will describe the different entities, their embryological development, and their correlation with diagrams and tomographic images.

Keywords: Inferior vena cava. Congenital anomalies. Computed tomography. Embryology.

Resumen

Las anomalías de la vena cava inferior (VCI) representan un amplio grupo de entidades que no son infrecuentes y en su mayoría son asintomáticas. Su desarrollo tiene como punto de partida la embriogénesis, con una secuencia compleja de eventos que ocurren durante la cuarta a octava semana de gestación. Dicho proceso involucra anastomosis complejas y regresiones de venas embrionarias. La VCI es la principal estructura de retorno venoso de las extremidades y los órganos abdominales. La correcta identificación de sus anomalías es fundamental, ya que algunas tendrán implicancia clínico-patológica directa, otras determinarán un abordaje diferente en la planificación de intervenciones vasculares y, en algunos casos, será de utilidad para diferenciarlas de condiciones patológicas. Con los actuales tomógrafos multislice se pueden adquirir imágenes con alta resolución espacial, lo que hace posible reconstrucciones de alta calidad y representaciones altamente detalladas de la anatomía. No existe una clasificación definitiva para las anomalías de la VCI. Se han descripto numerosas variantes, entre las que se destacan: agenesia, duplicación, localización izquierda, interrupción de la porción intrahepática y malformación de Abernethy; existen también anomalías de las venas tributarias. En este artículo describiremos las diferentes entidades, su desarrollo embriológico y su correlación con diagramas e imágenes tomográficas.

Palabras clave: *Vena cava inferior. Anomalías congénitas. Tomografía computada. Embriología.*

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Reception date: 21-07-2023
Acceptance date: 25-03-2024
DOI: 10.24875/RAR.23000064

Available online:28-08-2024
Rev Argent Radiol. 2024;88(3):116-124
www.revistarar.com

Introduction

Anomalies of the inferior vena cava (IVC) and its variants were first described by Abernethy in 1793, in a 10-month-old patient with polysplenia and dextrocardia who had a congenital mesocaval shunt and continuation of the vena cava with azygos vein.[1] These anomalies represent a broad spectrum of pathologic conditions that may be seen in as much as 8.7% of the general population. The genesis of these entities is linked to embryologic development, which involves complex anastomoses and regression of embryonic veins occurring during the fourth to eighth week of gestation.[2,3]

The role of imaging, mainly multislice computed tomography (MSCT), is crucial for the detection and characterization of these anomalies. Even if most entities are asymptomatic incidental findings, they can often have a direct clinical and pathological impact, mimic other conditions or pose a challenge for planning vascular procedures.[3] Furthermore, they may lead to the presence of venous insufficiency in the lower limbs, deep venous thrombosis and pelvic congestion syndrome.[4-6]

The aim of this pictorial essay is to describe the various congenital entities involving the IVC and its tributaries, considering their embryological correlate and their appearance on MSCT.

Imaging techniques

Color Doppler ultrasound is a useful modality for initial evaluation. However, it is operator dependent and visualization of the IVC may be limited by bowel gas or the patient's adverse body habitus.[7]

As MSCT is commonly used to evaluate multiple abdominal symptoms, it is often by this imaging modality that many IVC anomalies are first detected incidentally.3 In this context, images are obtained at 60-70 seconds after intravenous injection of contrast material, which allows good opacification of the suprarenal IVC. Nevertheless, images acquired at 70-90 seconds may provide better opacification of the infrarenal IVC.[8]

The advantages of MSCT include high spatial resolution, which allows imaging with isotropic data acquisition, and the availability of tools that enable postprocessing reconstructions, such as 3D volume-rendered and maximum intensity projection (MIP) images.[2,8]

Magnetic resonance imaging (MRI) is a valid alternative, particularly in patients who cannot receive iodinated contrast material. The absence of ionizing radiation is another important feature when patients belong to the pediatric and young adult populations.[8]

Embryogenesis of IVC

The mature IVC has four segments: the hepatic, suprarenal, renal and infrarenal IVC. The genesis of IVC involves various processes that include the formation of anastomoses and regression of embryonic venous structures.[3] Congenital anomalies of IVC result from abnormal persistence or regression of these fetal venous structures.[9]

These venous structures are mainly made up of the vitelline vein and a series of paired veins on either side of the midline, including the posterior cardinal, subcardinal and supracardinal veins.

The hepatic segment of the IVC is derived from the right vitelline vein. The suprarenal IVC arises from the cranial right subcardinal vein. The renal segment of the IVC is formed by the anastomoses between the supracardinal veins posteriorly and the subcardinal veins anteriorly.[10] A portion of the right supracardinal vein persists as the infrarenal segment. The posterior cardinal veins (the caudal aspects) become the iliac veins. Finally, the azygos system results from persistence of the superior supracardinal veins (Fig. 1).[3]

Agenesis of the IVC

La ausencia de VCI es rara e incluye dos variantes: ausencia de VCI infrarrenal con preservación del segmento suprarrenal o ausencia completa de VCI[2]. Esta última sugiere que existe una falla en el desarrollo de los tres pares de venas embrionarias, mientras que la primera implica una alteración en la génesis de las venas cardinales posteriores y supracardinales. Es complejo establecer un único evento embriológico, por lo que se plantea que pueda ser resultado de trombosis venosa perinatológica[11,12]. El retorno venoso en estos pacientes ocurre a través de venas lumbares ascendentes que drenan hacia el sistema ácigos y hemiácigos, dando como resultado colateralidad prominente de estructuras venosas que pueden simular masas paraespinales (Fig. 2)[7,13].

Duplicación de la VCI

The absence of IVC is rare and includes two variants: absent infrarenal IVC with normal suprarenal part or

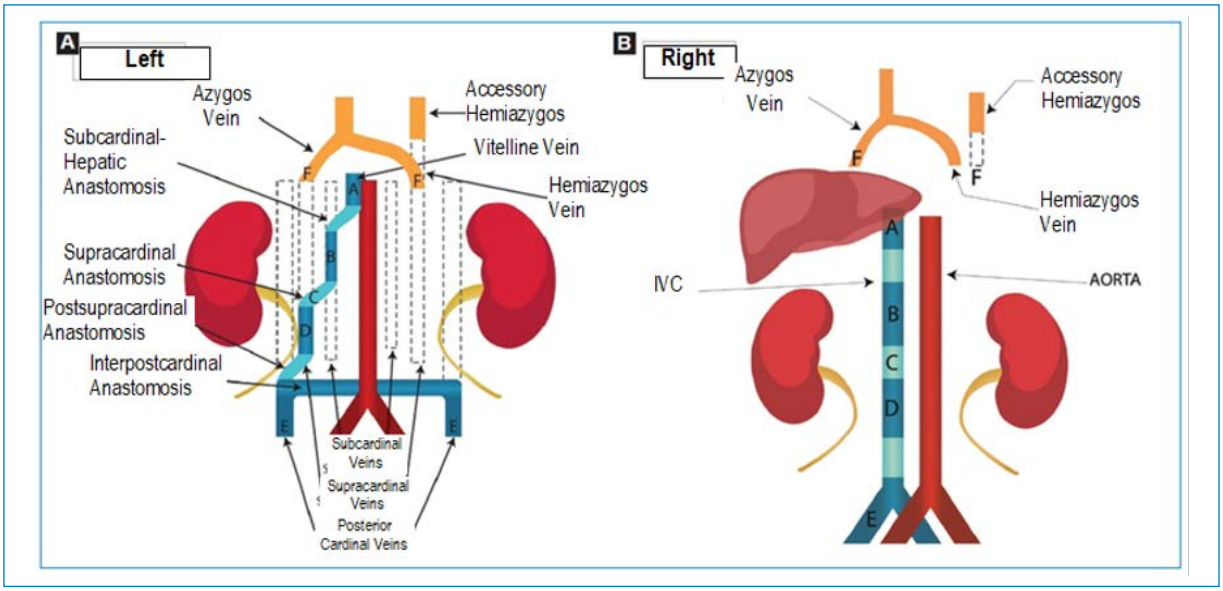

Figure 1. Drawing of the IVC embryogenesis (**left**) and normal final anatomic configuration in adults (right). The mature IVC (**dark blue**) is composed of four segments (**hepatic, suprarenal, renal and infrarenal**), which are derived from anastomoses (**light blue**) and regressions of embryonic veins. The hepatic segment of the IVC is derived from the right vitelline vein (**A**). The suprarenal IVC arises from the cranial segment of the right subcardinal vein (**B**). The suprasubcardinal anastomosis (**C**) will become the renal segment. A portion of the right supracardinal vein (**D**) persists as the infrarenal segment. The caudal portions of the posterior cardinal veins (**E**) become iliac veins. The azygos system results from persistence of the superior supracardinal veins (**F**).

complete absence of entire IVC.[2] The latter implies a failure of development of all three paired embryonic veins, while the former implies an impairment in the genesis of the posterior cardinal and supracardinal veins. As it is complex to identify a single embryonic event, it has been postulated that these conditions may result from perinatal venous thrombosis.[11,12] Venous return in these patients occurs via the ascending lumbar veins, which drain into the azygous and hemiazygous system, resulting in a prominent collateralization of venous structures that may mimic paraspinal masses (Fig. 2).[7,13]

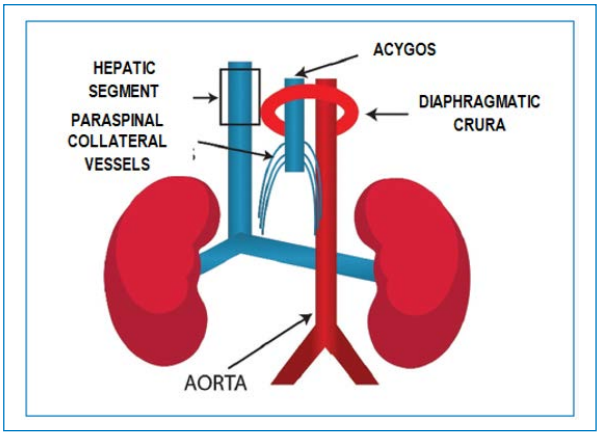

Figure 2. Schematic representation of IVC agenesis.

Left-sided IVC

The prevalence of this entity ranges from 0.2% to 0.5%.[10] It is secondary to regression of the right supracardinal vein and abnormal persistence of the left supracardinal vein.[3] A left-sided IVC courses cranially to the left of the abdominal aorta, joins the left renal vein (LRV) and together they course anterior to the aorta to drain into a normal suprarenal IVC (Fig. 4).[3,7] This anomaly by itself has no clinical significance; however, it may mimic paraaortic adenopathies.[7,14]

Anomalous Continuation of the IVC

This anomaly occurs as a result of an embryonic failure to form the right subcardinal-hepatic anastomosis. The suprarenal IVC drains into the azygos-hemiazygos system and returns to the heart through the superior vena cava. Because of the absence of the intrahepatic segment of the IVC, the suprahepatic

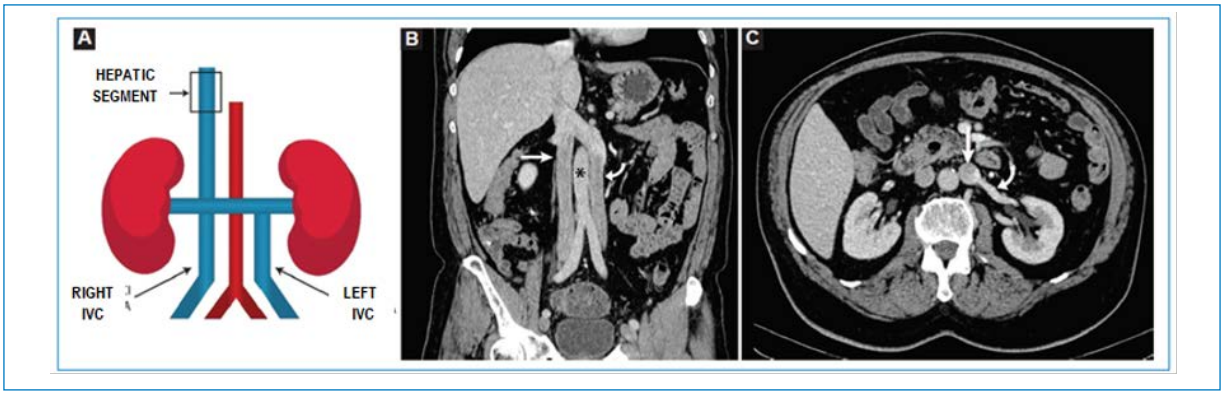

Figure 3. Duplication of the IVC. **A:** the drawing shows the right and left IVC and the convergence of the LRV with the latter. **B:** Intravenous contrast-enhanced coronal CT scan shows duplication of the IVC on the right side (straight arrow) and left side (curved arrow) of the aorta (asterisk), and convergence at suprarenal level. **C:** Axial CT scan shows double IVC with LRV (curved arrow) draining into the ipsilateral IVC (straight arrow).

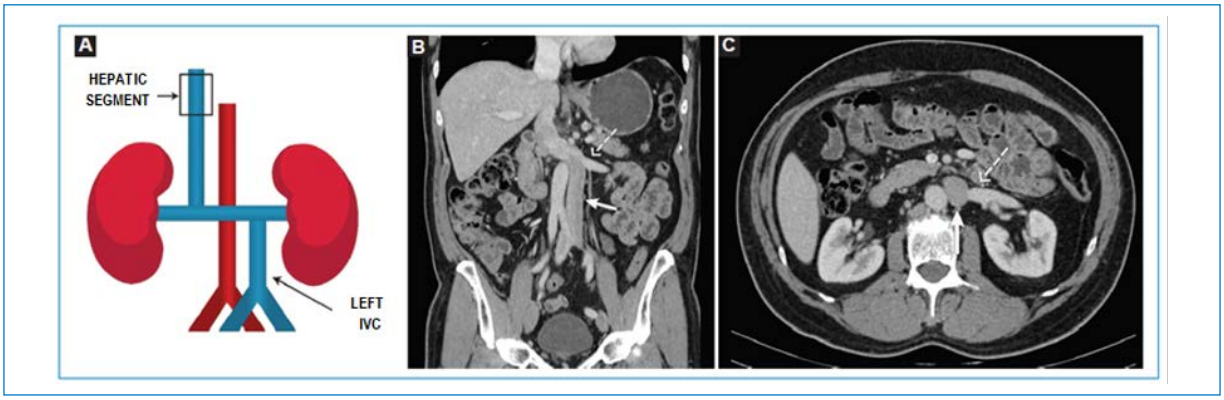

Figure 4. Left-sided IVC. **A:** the drawing illustrates the IVC on the left side of the aorta draining into the ipsilateral renal vein. **B** and **C:** Coronal and axial CT scans, respectively, with intravenous contrast. IVC on the left side (straight arrow) and convergence with the ipsilateral renal vein (dashed arrow).

veins drain directly into the right atrium.[3] (Fig. 5). The azygos vein is usually dilated and may mimic a paratracheal mass or retrocrural lymphadenopathy.[10,15] In addition, the possibility of accidental ligation of the azygos vein during chest surgery should be considered, as this would constitute a fatal event for the patient.[10] It is imperative to consider this variant prior to planning cardiopulmonary bypass surgery.[10,16] This anomaly is most commonly associated with heterotaxy syndromes (left isomerism) and intestinal malrotation.[17]

Retrocaval ureter

Unlike the usual anatomy of the infrarenal IVC, which arises from the right supracardinal vein (located posteromedial to the ipsilateral ureter), in this anomaly the infrarenal IVC develops from the right posterior cardinal vein (which lies anterior and lateral to the ureter). As a result, the proximal ureter becomes confined posteriorly to the IVC. This can cause compression, resulting in hydronephrosis or recurrent urinary tract infections (Fig. 6).[16] This anomaly has a prevalence of 0.06% to 0.17% and occurs more commonly in men, on the right side.[10,18]

Membranous obstruction of the intrahepatic IVC

Although rare in most countries worldwide, membranous obstruction of the IVC is the most common cause of hepatic venous outflow obstruction in Asia and South

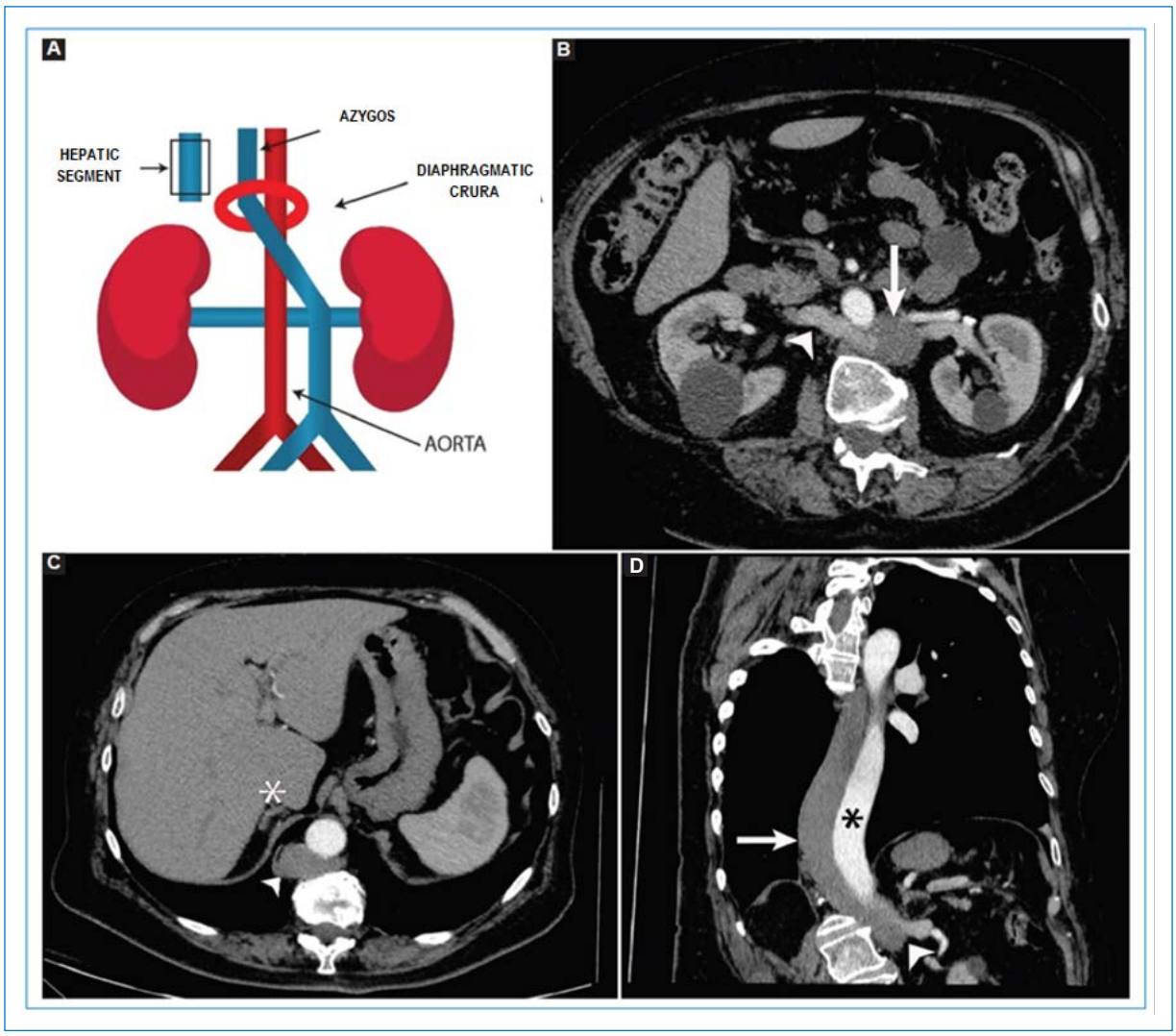

Figure 5. Anomalous continuation of the IVC. **A:** the drawing shows left IVC, with absence of the hepatic segment and abnormal drainage into the azygos system. **B:** Axial CT scan shows IVC on the left (straight arrow) and a retroaortic course of the right renal vein (arrowhead). **C:** axial image showing absence of the hepatic segment of the IVC (asterisk) and prominent azygos vein (arrowhead). **D:** oblique coronal CT scan shows dilated azygos vein (straight arrow) accompanying the course of the descending aorta (asterisk) due to abnormal drainage of left IVC (arrowhead).

Africa. It appears as a generally complete occlusive lesion of the IVC, but occasionally with a small central opening, located near the entry of the IVC into the right atrium or just below the diaphragm. The etiology remains uncertain. However, two possible origins have been suggested: it might be the result of a congenital vascular malformation or a sequel of thrombosis in the hepatic portion of the IVC.[19] Prominent intrahepatic and extrahepatic collateral vessels may develop and, clinically, it may cause obstruction of the hepatic outflow, congenital Budd-Chiari syndrome and hepatocellular carcinoma.[3]

Extrahepatic portocaval shunt (Abernethy malformation)

Abernethy malformation, also known as congenital extrahepatic portosystemic shunts (CEPS) is a rare condition in which portal blood drains partially or completely into the systemic circulation via an abnormal communication. Embryologically, the portal vein (PV) develops from the right and left vitelline veins and from intervitelline anastomoses around the duodenum. Selective regression of anastomotic channels forms the PV. It is thought that these extrahepatic shunts result from excessive involution

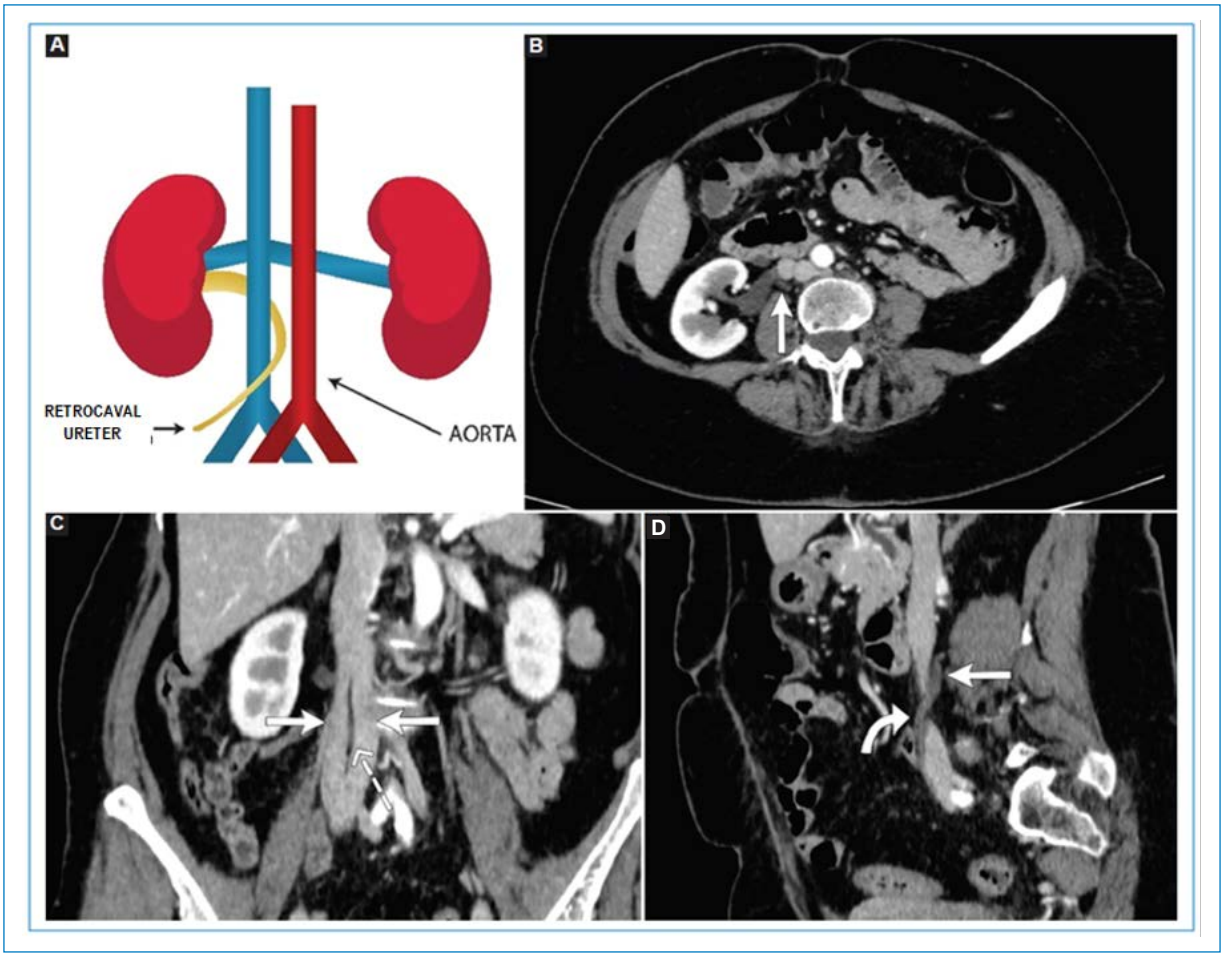

Figure 6. A. Drawing of the retrocaval ureter. **B:** oblique axial image showing right ureter coursing posterior to the IVC (straight arrow). **C:** oblique coronal image showing an association with right incomplete duplication of the IVC, with the ureter (dashed arrow) becoming trapped within it and forming a "periureteral ring" (straight arrows). **D:** oblique sagittal images showing the retrocaval course of the proximal third of the ureter (straight arrow); it courses anterior to the IVC after crossing the ring (curved arrow).

of the vitelline vein or failure of the vitelline vein to establish an anastomosis with the hepatic sinusoids or suprahepatic veins.[3] It is classified into two categories:[20]

- Type 1 (complete): it is characterized by complete shunting of portal blood into the IVC and congenital absence of the portal vein. This type, in turn, is divided into two subtypes:

• Ia: where the splenic vein (SV) and the superior mesenteric vein (SMV) drain independently into a systemic vein.

• Ib: where the SV and the SMV join to form a common trunk which drains into a systemic vein.

- Type 2 (partial): partial anastomosis between an intact PV and the IVC.[3]

In type 1 shunts, the PV drains extrahepatically into the IVC at any point inferior to the suprahepatic veins confluence. However, cases of the PV draining directly into the suprahepatic IVC or the right atrium have been reported in the literature (Fig. 7).[22,23]

Clinical features of CEPS can be divided into those related to shunting of the portal circulation (hepatopulmonary syndrome, metabolic dysfunction and hepatic encephalopathy), associated congenital abnormalities (cardiac, arterial and venous, visceral and musculoskeletal anomalies) and those secondary to hepatic lesions (including, but not limited to, regenerative nodules, focal nodular hyperplasia and hepatocellular carcinoma).[21]

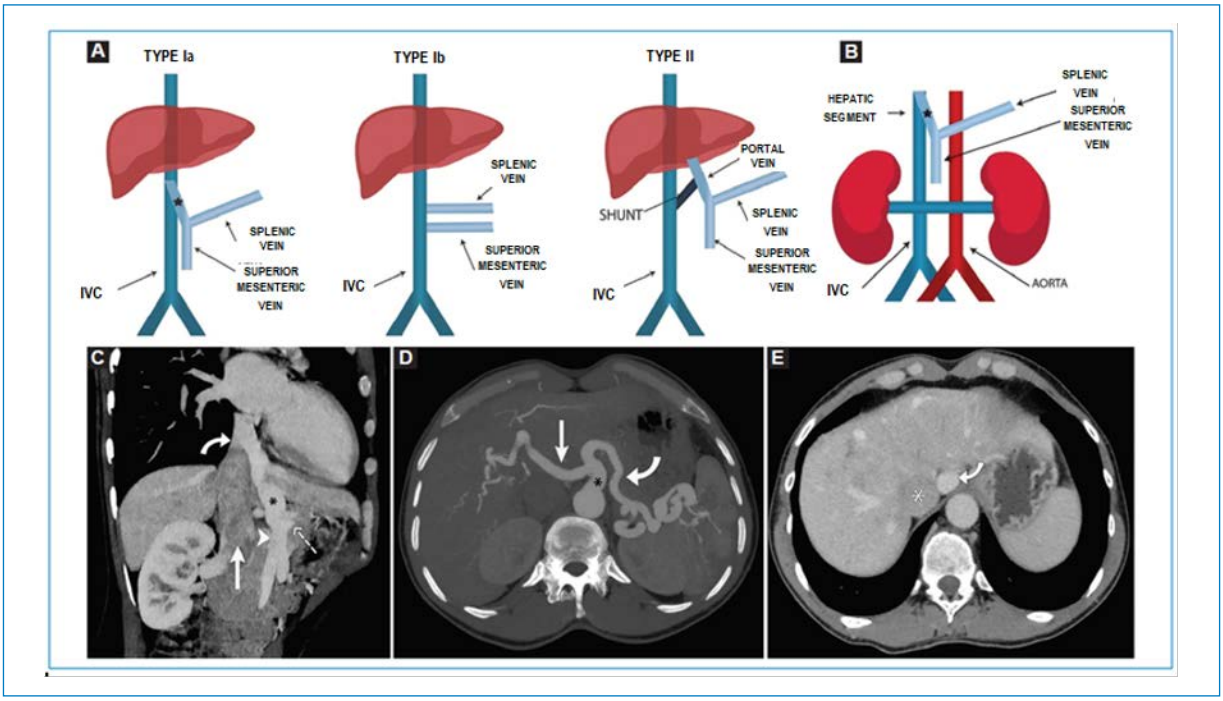

Figure 7. **A:** drawing of the different types of Abernethy malformation. **B:** schematic representation of the extrahepatic portocaval shunt, showing a splenic vein (SV) and superior mesenteric vein (SMV) joining to form a common trunk (asterisk) draining into the suprahepatic IVC (arrow). **C:** oblique coronal MIP reconstruction. Normal position of the IVC (straight arrow). SMV (arrowhead) joining the SV (dashed arrow) to form a common trunk (asterisk) that drains into the suprahepatic IVC (curved arrow). **D:** axial MIP reconstruction showing the prominence of the celiac trunk (asterisk), hepatic artery (straight arrow) and splenic artery (curved arrow). **E:** Axial CT scan, immediately caudal to the convergence of the IVC (asterisk) and the abnormal extrahepatic portal vein (curved arrow). The scan also shows heterogeneous post-contrast enhancement of the hepatic parenchyma, caused by the disturbance in perfusion typical of this entity.

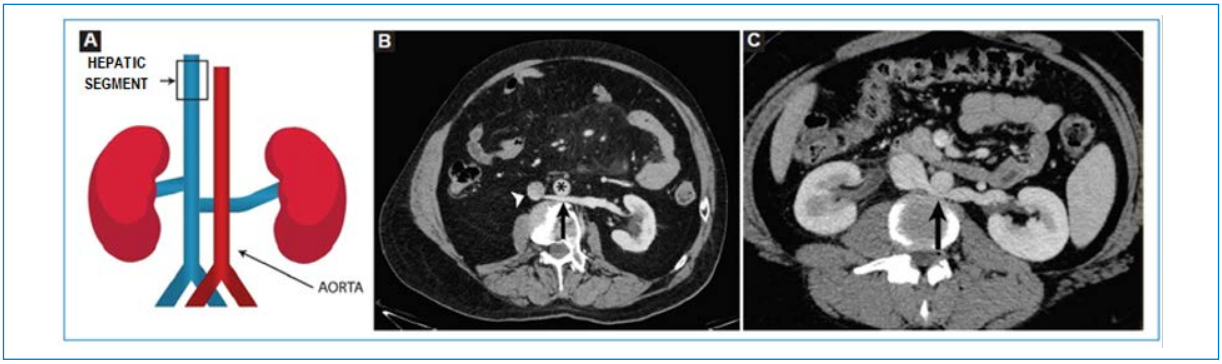

Figure 8. Variant of the tributaries: retroaortic LRV. A: Drawing of the LRV in retroaortic position. **B:** oblique axial CT scan shows the LRV (straight arrow) coursing posterior to the aorta (asterisk) and its confluence with the IVC (arrowhead). **C:** axial CT scan performed in a different patient, with the same finding and slight compression of the LRV by the aorta (straight arrow).

Anomalies of tributaries (Retroaortic and circumaortic LRV)

Anomalies of the IVC also include other variants derived from tributary veins. Normally, the LRV is derived from intersubcardinal anastomoses, which course anterior to the aorta.

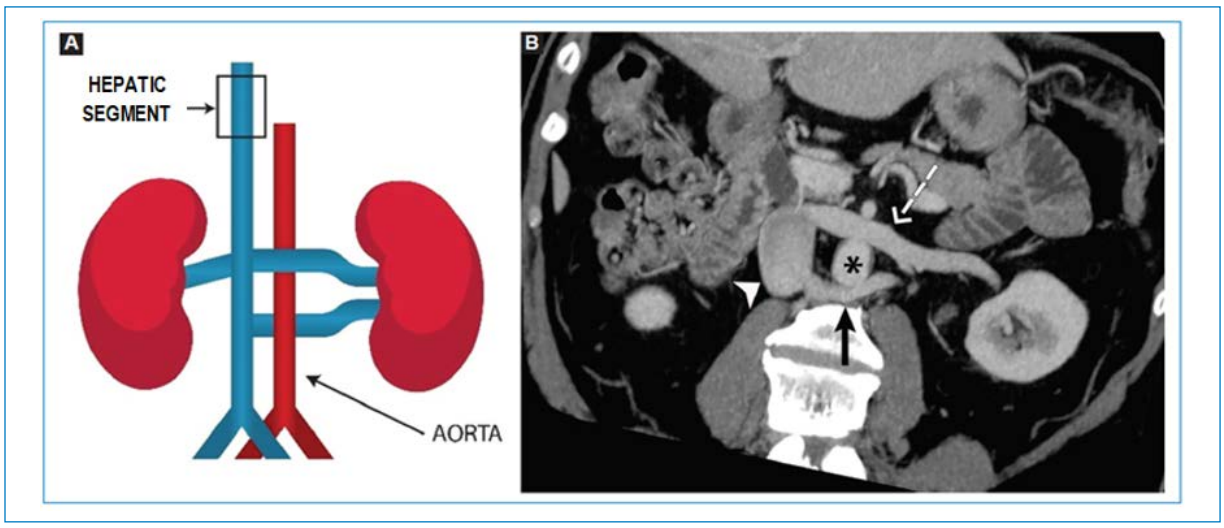

Figure 9. Variant of the tributaries: circumaortic LRV. **A:** Drawing of the LRV with circumaortic course. **B:** oblique axial MIP reconstruction. Bifurcation of the LRV, with one tributary coursing anterior (dashed arrow) and the other posterior (straight arrow) to the aorta (asterisk) to drain into the IVC (arrowhead).

The LRV is retroaortic when it derives from intersupracardinal veins, which lie posterior to the aorta. This occurs in 1.7% to 3.4% of individuals (Fig. 8).

Persistence of both the intersupracardinal and intersubcardinal veins results in circumaortic venous ring (with one vein anterior and the other posterior to the aorta). The prevalence of this anomaly is 2.4% to 8.7% (Fig. 9).

These variants play a significant role in the preoperative planning of nephrectomy. They can also mimic adenopathy. On rare occasions, compression of the posterior course of the renal vein may occur, resulting in periuretic venous insufficiency and hematuria, among other conditions.[7]

Conclusion

Imaging techniques play an essential role in the detection of the various congenital anomalies of the IVC. A correct characterization of these entities, mainly by intravenous contrast-enhanced CT scan, is crucial, since they may have direct clinical implications, mimic other conditions or pose a challenge for planning vascular procedures.

Funding

The authors declare that no funding was received for this study.

Conflicts of interest

The authors declare no conflicts of interest.

Ethical responsibilities

Protection of human subjects and animals. The authors declare that no experiments were performed on humans or animals for this investigation.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for text generation. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, or for the creation of images, graphics, tables, or their corresponding captions.

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