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Is IVC looking different ?

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Learning Objectives

- To review the embryogenesis of the inferior vena cava (IVC) and describe the most common variations in IVC anatomy

- To recognize the clinical and surgical implications of identifying these anomalies

Background

- The IVC is composed of four segments (hepatic, adrenal, renal, and infrarenal) which are formed by the successive development and regression of three paired veins: the posterior cardinal, subcardinal, and supracardinal systems.
- Abnormal or absent regression or persistence of any of these venous structures results in different anomalies, which present in approximately 4% of the population.
- Congenital anomalies of IVC and its tributaries have become more commonly recognized in asymptomatic patients since the development of cross-sectional imaging.
- Familiarity with these variations is essential for correct interpretation of cross-sectional images, to avoid erroneous diagnosis of retroperitoneal and mediastinal masses or adenopathy and to alert the surgeon and angiographer of potential sources of complications preoperatively.

Background

Embryogenesis of the IVC

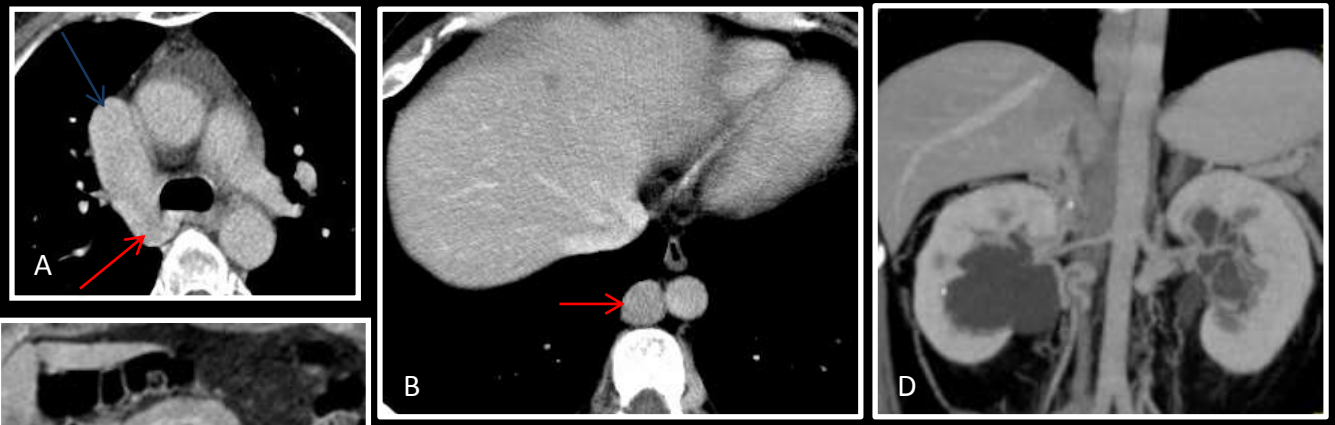
- The IVC develops between the 6th and 8th embryonic week, formed after the regression of three pairs of primitive veins (in order of appearance): posterior cardinal veins, subcardinal veins and supracardinal veins.
- The posterior cardinal veins are progressively replaced, first by the subcardinal veins and later by the supracardinal veins, which together form the subhepatic IVC.
- The posterior cardinal system regresses without forming any of the normal IVC. The subcardinal veins are ventromedial and the supracardinal veins dorsomedial to the posterior cardinal veins.
- The right subcardinal system forms the suprarenal segment of the IVC and anastomoses with the developing hepatic vessels, which are derived from the right vitelline vein to form the intrahepatic IVC.
- Anastomotic channels between the supracardinal and subcardinal veins form the intervening renal segment of the IVC. Anastomosis of the right and left subcardinal systems forms the normal left renal vein. The remainder of the subcardinal system regresses.
- The right supracardinal system forms the azygus system cephalad to the renal veins and the infrarenal segment of the IVC. It forms an anastomosis with the iliac veins, which are derived from the persistent posterior cardinal veins.
- The left supracardinal system gives rise to the hemiazygus system cephalad to the renal veins and normally regresses caudal to the renal vein.

Clinical Findings

Anomalies of the suprarenal segment of the IVC

Interruption of the IVC with Azygous or Hemiazygous Continuation

- Failure of 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 derivered from the thoracic segment of the right supracardinal vein.
- The prevalence is 0.6%.
- The IVC is normal below the level of renal vein but has no hepatic segment. 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.



Absence IVC hepatic segment with azygos continuation. CT images shows: (A) enlarged azygos vein (red arrow) draining into the superior vena cava (blue arrow). (B and C) the vessel parallel to the aorta under the crus is the azygos vein (red arrow). (D) Absence of IVC in its normal location.

Association with situs anomalies (asplenia or polysplenia syndrome) and congenital heart malformations.

Azygos-hemiazygos system dilatation may lead to a misdiagnosis of mediastinal, retrocrural and/or retroperitoneal lymphadenopathies.

Important for surgical planning for cardiopulmonary bypass.

Technical difficulties during cardiac catheterization.

Clinical Findings

Anomalies of the renal segment of the IVC

Normally the left renal vein is derived from the intersubcardinal anastomoses, which course anterior to the aorta.

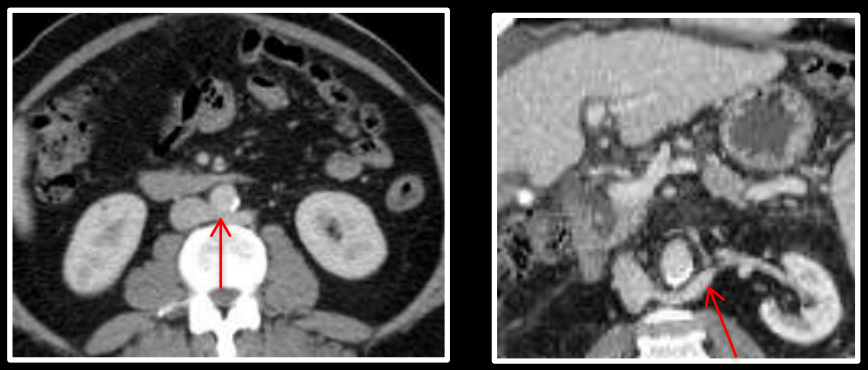
Circumaortic Left Renal Vein

- Persistence of the both intersubcardinal and intersupracardinal anastomosis system, resulting in a circumaortic venous ring. The prevalence is 2,4-8.7%.
- Thus, there are two left renal veins, the one that normally crosses anterior to the aorta and the dorsal left renal vein, which originates below the previous and crosses behind the aorta, both draining into the IVC.

Contrast-enhanced craniocaudal CT scans show the left renal vein coursing anterior (red arrow) and posterior (blue arrow) to the aorta.

Retroaortic Left Renal Vein

- Regression of the intersubcardinal anastomosis (ventral arch). The left renal vein derives from the intersupracardinal veins, which lie posterior to the aorta. The prevalence is 1.7%-3.4%.



Two cases of retroaortic left renal vein. A single renal vein passes posterior to the aorta.

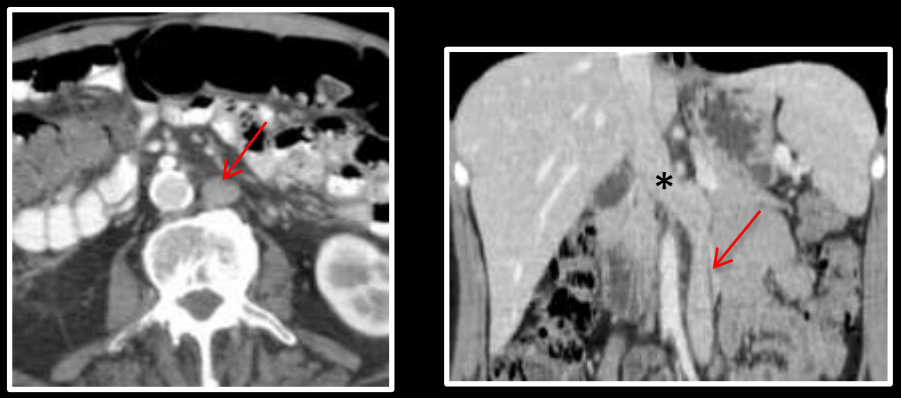
Preoperative planning of nephrectomy
Misdiagnosis as retroperitoneal lymphadenopathy
Association with the “Nutcracker phenomenon”

Clinical Findings

Anomalies of the infrarenal segment of the IVC

Left IVC

- Anomalous regression of the right supracardinal vein with persistence of left supracardinal vein. The prevalence is 0.2%-0.5%.
- The left IVC joins the left renal vein and crosses anterior to the aorta to join the normal right-sided (prerenal) IVC.



Two cases of a left sided IVC. CT images show a large single IVC to the left of the spine at levels below the renal veins (red arrow). A left-sided IVC (red arrow) joining the left renal vein, which unites with the right renal vein to form a normal right-sided suprarenal IVC (*).

Misdiagnosis as left para-aortic lymphadenopathy
Complications during aortic aneurysm repair surgery
Difficult placement of an IVC filter (transjugular access)

Double IVC

- Persistence of both supracardinal veins. The prevalence is 1%-3%.
- The left IVC drains into the left renal vein, which then joins the right IVC.
- Asymmetry in the sizes of the left and right veins.

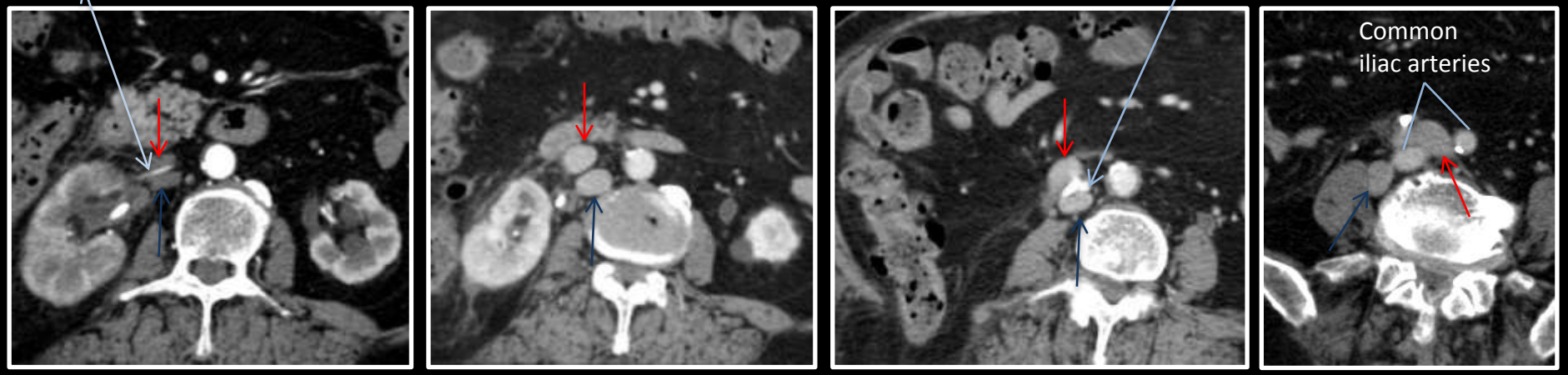
Two cases of a double IVC. Contrast enhanced CT shows right and left infrarenal IVCs (red arrows). The left IVC opens into the left renal vein (blue arrow), which crosses anterior to the aorta to form a single right-sided suprarenal IVC (*).

Double IVC has clinical implications that are similar to those of left IVC.
This anomaly should be suspected in cases of recurrent episodes of pulmonary embolism despite placement of an IVC filter

Clinical Findings

Anomalies of the infrarenal segment of the IVC

High bifurcation of the IVC



High bifurcation of the IVC – incidental finding in a patient with complicated pyelonephritis and a right double-J.

The union of the left (red arrow) and right (blue arrow) iliac veins to form the IVC occurs more cranially than usual, 2cm bellow the emergence of the left renal artery.

The suprarenal IVC (*) is normal.

Conclusions

- The IVC embryogenesis is complex and involves multiple anastomosis formed between the three primitive paired veins and the involution of any of these pairs during embryonic development.
- Vena cava anomalies reflect an abnormal regression or persistence of those various embryonic veins and results in anomalies of the anatomy of the inferior vena cava: left IVC, double IVC, absence of the hepatic segment of the IVC with azygos continuation, circumaortic left renal vein, retroaortic left renal vein, among others.
- Knowledge of the various developmental anomalies of the venous system and recognition of their CT appearances are critical for proper image interpretation (avoids diagnostic pitfalls) and patient management (prevent complications during surgery or catheterization).
- The diagnostic radiologist should always aware the treating physician prior to surgical and interventional procedures.

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