Forgotten and frequent congenital anomalies of the inferior vena cava. What we need to know.

Poster No.: C-1459
Congress: ECR 2012
Type: Educational Exhibit
Authors: M. M. Mendigana Ramos¹, A. Sáez de Ocáriz García², C. Antón Munárriz², J. Agreda Sadaba²; ¹Etxauri/ES, ²Pamplona/ES
Keywords: Congenital, Contrast agent-intravenous, CT, Thorax, Abdomen
DOI: 10.1594/ecr2012/C-1459

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR’s endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method ist strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slide shows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

To illustrate the different congenital anomalies of the inferior vena cava and its embryonic development.

To present a revision in a 64-slice multidetector computed tomography of the most frequently encountered anomalies of the inferior vena cava.
Background

Congenital anomalies of the inferior vena cava (IVC) are most commonly recognized in asymptomatic patients since the development of cross-sectional images. In addition to this, these anomalies could have significant clinical implications. Awareness of these anomalies is necessary to avoid diagnostic pitfalls.

It is important a correct interpretation in diagnostic images so as to avoid false positives such as retroperitoneal adenopathies or masses. Besides, it is also useful for the surgeon being aware of these anomalies before an intervention.
Imaging findings OR Procedure details

Basic knowledge of the embryogenesis of the vena cava is essential to recognize and understand the anomalies of the vena cava on CT images. We reviewed 500 thoracoabdominal scanners with intravenously administered contrast material in a 6 months period, where we found the most frequent anatomical variants of the IVC: left IVC, double IVC, azygos continuation of the IVC, circumaortic left renal vein, retroaortic left renal vein and circumcaval ureter.

1. Embryogenesis of the IVC:

The embryogenesis of the IVC is a complex process of multiple regression and anastomosis. The IVC is developed between the 6th and 8th week of embryonic life. It is formed from the regression of three pairs of embryonic veins known as: Posterior cardinal veins, subcardinal veins and supracardinal veins. Finally, a complex system of the IVC thus forming four segments from top to bottom are known as: Hepatica, adrenal, renal and infrarenal segment (1) (Fig. 1).

2. Congenital anomalies of the IVC:

A. Left IVC

A left IVC results from regression of the right supracardinal vein with persistence of the Leith supracardinal vein. The prevalence is 0.2%-0.5% (2).

The left IVC joins the left renal vein, and both are directed anteriorly from the aorta to join the normal course on the right side IVC (Fig. 2,3,4,5). This anomaly has no clinical significance. But it could be confused with a left paraaortic adenopathy, so it is important to recognize it in the diagnostic images. Besides, when needing to access to the infrarenal IVC with a transjugular approach for a placement of an IVC filter, it could be difficult.

B. Double IVC

Duplication of the IVC results from the persistence of both supracardinal veins. The prevalence is 0.2% to 3% (3). The left IVC ends at the left renal vein, crossing the front of the aorta in the normal way to join the right (2). The major clinical significance of this anomaly is the misdiagnosis as adenopathy, especially when the vein does not have enough contrast
enhancement. This anomaly should be suspected in patients with recurrent episodes of pulmonary embolism after the placement of an IVC filter (3) (Fig. 6,7).

C. Azygos continuation of the IVC

Azygos continuation of the IVC is also known as the absence of the hepatic segment of the IVC with azygos continuation.

The embryonic event is the failure of the formation of the right subcardinal-hepatic anastomosis, resulting on atrophy of the right subcardinal vein. Consequently, blood is shunted from the suprasubcardinal anastomosis through the retrocruural azygos vein, which is partially derived from the thoracic segment of the right supracardinal vein. Azygos vein joins the superior vena cava in its normal place in the right paratracheal space. (Fig. 8,9,10,11,12). The prevalence is 0,6 % (2).

The clinical and radiological findings lies it is frequently associated with congenital cardiac malformations or asplenia or polysplenia syndrome. However, most patients are asymptomatic. This congenital anomaly could be confused with adenopathy.

The knowledge of the existence of this anomaly of the IVC can make improve surgical planning of cardiopulmonary "bypass" surgery and also prevents technical difficulties during cardiac catheterization.

D. Circumaortic Left Renal Vein

A circumaortic left renal vein results from the persistence of the intersupracardinal and intersubcardinal veins, forming a venous ring, in which the superior renal vein crosses the aorta anteriorly (receives the left adrenal vein) and the inferior renal vein crosses posterior to the aorta (receiving the left gonadal drainage) (Fig. 13,14,15,16). The prevalence is 2,4 % - 8.7%. (3).

Knowing the existence of this abnormality is important in the planning of nephrectomy and can be misdiagnosed as an adenopathy.

E. Retroaortic Left Renal Vein
There is a persistence of the dorsal segment of the left renal vein in embryo with a regression of the ventral arch (2). A single renal vein crosses posteriorly to the aorta. (Fig.17,18,19,20). The prevalence is 2.1%.

This abnormality is related to Nutcracker phenomenon, which is defined as the compression of the renal vein (because of its posterior course to the aorta) resulting in periureteric varices, hypertension and hematuria.

F. Circumcaval ureter or retrocaval ureter

This embryologic defect occurs due to an anomaly of the development of vena cava, rather than ureteral cause.

The infrarrenal IVC develops from the right posterior cardinal vein, which lies anterior and lateral to the ureter. As a result, part of the right ureter becomes trapped posterior and medial to the IVC (3).

Patients with this anomaly may develop hydrenephrosis because of the right ureter compression or recurrent urinary tract infections.

The diagnosis is often made with an intravenous urography or CT which confirm the anomalous way of the proximal ureter. The treatment of this abnormality consists on surgical relocation of the ureter anterior to the IVC.
Fig. 1: Drawing illustrates the embryologic development of the IVC. Three pairs of veins—the posterior cardinal (blue), subcardinal (vertical red), and supracardinal (purple) veins—develop in succession on either side of the aorta (gray). Portions of these venous channels regress (dashed colored lines), whereas others persist (solid colored lines) to form the infrahepatic IVC. The right subcardinal vein forms the suprarenal segment of the IVC, and the right supracardinal vein forms the infrarenal segment. The hepatic segment is derived from the vitelline vein (green). Anastomotic channels (black) connect various segments of the IVC. An intersubcardinal anastomosis, that passes anterior to the aorta, forms the left renal vein (horizontal red line), whereas intersupracardinal and interposterior cardinal anastomoses normally regress. The supracardinal veins continue...


**Fig. 2:** A,B. Left IVC in a 62-year-old patient with pancreatic carcinoma. A,C. Contrast enhanced axial CT image shows a left-sided IVC (arrow).

© Hospital de Navarra - Etxauri/ES
**Fig. 3:** A,B. Left IVC in a 62-year-old patient with pancreatic carcinoma. B,D. Coronal reformatted images show a left IVC (arrow).

© Hospital de Navarra - Etxauri/ES
**Fig. 4:** C,D. Left IVC in a 51-year-old patient with massive hemoptysis A,C. Contrast enhanced axial CT image shows a left-sided IVC (arrow).

© Hospital de Navarra - Etxauri/ES
**Fig. 5:** C,D. Left IVC in a 51-year-old patient with massive hemoptysis. B,D. Coronal reformatted images show a left IVC (arrow).

© Hospital de Navarra - Etxauri/ES
Fig. 6: Double IVC in a 57-year-old patient with cirrhosis, ascites descompensation and adenocarcinoma of the rectum. A. Coronal reformatted images.

© Hospital de Navarra - Etxauri/ES
**Fig. 7:** Double IVC in a 57-year-old patient with cirrhosis, ascites descompensation and adenocarcinoma of the rectum. B. Axial CT scan obtained inferior to the renal veins shows left (small arrow) and right (large arrow) IVCs.

© Hospital de Navarra - Etxauri/ES
Fig. 8: Interruption of the IVC with Azygos continuation in a 58-years-old patient with breast carcinoma. A,B,C,D,E. Contrast-enhanced CT scans show the enlarged azygos vein (arrows) draining into the superior vena cava. A. CT scans obtained the level of the azygos vein arch.

© Hospital de Navarra - Etxauri/ES
**Fig. 9:** Interruption of the IVC with Azigos continuation in a 58-years-old patient with breast carcinoma. A,B,C,D,E. Contrast-enhanced CT scans show the enlarged azygos vein (arrows) draining into the superior vena cava. B. The hepatic segment of the IVC is absent.

© Hospital de Navarra - Etxauri/ES
**Fig. 10:** Interruption of the IVC with Azigos continuation in a 58-years-old patient with breast carcinoma. A,B,C,D,E. Contrast-enhanced CT scans show the enlarged azygos vein (arrows) draining into the superior vena cava.

© Hospital de Navarra - Etxauri/ES
Fig. 11: Interruption of the IVC with Azygos continuation in a 58-years-old patient with breast carcinoma. A,B,C,D,E. Contrast-enhanced CT scans show the enlarged azygos vein (arrows) draining into the superior vena cava.

© Hospital de Navarra - Etxauri/ES
**Fig. 12:** Interruption of the IVC with Azigos continuation in a 58-years-old patient with breast carcinoma. A,B,C,D,E. Contrast-enhanced CT scans show the enlarged azygos vein (arrows) draining into the superior vena cava.

© Hospital de Navarra - Etxauri/ES
Fig. 13: A,B. Circumaortic left renal vein in a 59-year-old patient with colon cancer and liver metastases. Contrast enhanced CT scans show the left vein coursing anterior and posterior (A,D) to the aorta (arrows).

© Hospital de Navarra - Etxauri/ES
**Fig. 14:** A,B. Circumaortic left renal vein in a 59-year-old patient with colon cancer and liver metastases. Contrast enhanced CT scans show the left vein coursing anterior and posterior (A,D) to the aorta (arrows).

© Hospital de Navarra - Etxauri/ES
Fig. 15: C,D. Circumaortic left renal vein in a 43-year-old patient with esophagus carcinoma. Contrast enhanced CT scans show the left vein coursing anterior and posterior (A,D) to the aorta (arrows).

© Hospital de Navarra - Etxauri/ES
Fig. 16: C,D. Circumaortic left renal vein in a 43-year-old patient with esophagus carcinoma. Contrast enhanced CT scans show the left vein coursing anterior and posterior (A,D) to the aorta (arrows).

© Hospital de Navarra - Etxauri/ES
**Fig. 17:** A,B. Retroaortic left renal vein in a 54-year-old man with lung cancer and liver metastases. CT scans show the left renal vein (arrows) descending to cross posterior to the aorta.

© Hospital de Navarra - Etxauri/ES
Fig. 18: A,B. Retroaortic left renal vein in a 54-year-old man with lung cancer and liver metastases. CT scans show the left renal vein (arrows) descending to cross posterior to the aorta.

© Hospital de Navarra - Etxauri/ES
Fig. 19: C,D. Retroaortic left renal vein in a 62-year-old man with lung cancer. CT scans show the left renal vein (arrows) descending to cross posterior to the aorta.

© Hospital de Navarra - Etxauri/ES
**Fig. 20:** C,D. Retroaortic left renal vein in a 62-year-old man with lung cancer. CT scans show the left renal vein (arrows) descending to cross posterior to the aorta.

© Hospital de Navarra - Etxauri/ES
Conclusion

We made a review of 500 thoracoabdominal scanners with intravenously administered contrast material during 6 months, finding different abnormalities of IVC in 38 patients, such as: left IVC (5.4%), double IVC (2.7%), azygos continuation of the IVC (5.4%), circumaortic left renal vein (18.9%), retroaortic left renal vein (67.6%) and circumcaval ureter (0%).

Congenital anomalies of the inferior vena cava frequently are incidental findings in asymptomatic patients. Knowledge of IVC anomalies is essential to avoid diagnostic pitfalls and the identification is necessary to prevent complications during surgery or catheterization.
References

