Congenital anomalies of the inferior vena cava and appropriate interventional radiology

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Learning objectives

To discuss the knowledge of congenital anomalies of the inferior vena cava (IVC), renal vein and azygos system for successful interventional treatment. To illustrate the radiological imaging of complications of interventional treatment or central venous catheterization on CT.
Background

Most congenital anomalies of the IVC are asymptomatic. Detecting these anomalies is essential for successful central venous catheterization (CVC). These anomalies are also recognized as a possible risk factor for deep vein thrombosis, particularly in young adult. Knowledge of the image characteristics of these anomalies is important to place of IVC filter correctly.
**Embryogenesis** ¹⁻³)

Embryogenesis of the IVC and renal venous system is complex. The infraheptic IVC develops between the 6th and 8th weeks of embryonic life as a composite structure formed from the continuous appearance and regression of three paired embryonic veins. In order of appearance, they are posterior cardinal, the subcardinal, and the supracardinal veins. The supracardinal veins are a paired system lying dorsal to the developing aorta; multiple midline anastomotic channels exist between left and right supracardinal veins. Right-sided IVC is created from right supracardinal vein, with concomitant obliteration of the left supracardinal and of the retroaortic anastomotic channels.

**Anomalies of IVC (Left IVC, Double IVC)**

**Left IVC** ²⁻⁵)

Left IVC, with a prevalence of 0.2-0.5%, has as its embryological substratum a persisting left supracardinal vein with regression of the right, the mirror image of a normal situation. As a consequence at the level of the renal hila, a left IVC passes either in front of or behind the aorta and runs to the right, assuming the normal position (Fig. 1). The major clinical significance of this anomaly is the potential for misdiagnosis as left-sided paraaortic adenopathy. Transjugular access to the infrarenal IVC for placement of an IVC filter may be difficult. Central venous catheterization of left IVC from femoral vein has risk of venous thrombosis at level of joint of left renal vein (Fig. 2).

**Double IVC** ²⁻⁶)

Duplication of the IVC results from persistence of both supracardinal veins. The prevalence is 0.2-3%. The left IVC typically ends at the left renal vein, which crosses anterior to the aorta in the normal fashion to join the right IVC. Pelvic venous variations of double IVC are classified into 5 types according to the patterns of the interiliac communicating veins (Fig. 3): double IVC with no interiliac communication (Fig. 4), double IVC with interiliac communication from left common iliac vein (Fig. 5), double IVC with interiliac communication from right common iliac vein (Fig. 6), double IVC with interiliac communication from left internal iliac vein (Fig. 7) and double IVC with interiliac communication from right internal iliac vein. The most common type is a double IVC with no interiliac communication (Fig. 4). Differential diagnosis of double IVC is dilated gonadal veins (Fig. 8). The gonadal veins may be misinterpreted as a double IVC because they run close to the ipsilateral IVC lumen, particularly the left gonadal vein that drains into the left renal vein. Evaluation of the peripheral connection of these veins is crucial in distinguishing them from IVC anomalies because the gonadal veins definitely originate from the ovaries or testes (Fig. 9). In contrast, the double IVC originated from iliac veins.
IVC filter for patient with double IVC should be replaced in both IVC s or suprarenal IVC. IVC filter for patient with ovarian vein thrombosis should be replaced suprarenal IVC (Fig. 10).

Right double IVC is a very rare IVC anomaly which is defined as 2 postrenal IVCs lying to the right of the abdominal aorta. Characteristic CT findings of right double IVC are ventral-dorsal vessels originates from the right subcardinal vein, whereas the dorsal vessels originates from the right supracardinal vein. The differential diagnosis of right double IVC is the periureteral venous ring. However, in periureteral venous ring, the anastomic relationship of these 2 vessels is medial and lateral vessels.

**Retrocaval ureter**

Retrocaval (circumcaval) ureter is an uncommon venous anomaly in which the right ureter courses posterior to the inferior vena cava and partially encircles it. Retrocaval ureter results from persistence of the posterior cardinal venous system that anomalously forms the inferior vena cava, and subsequently courses anterior to the ureter for a variable distance. This can cause varying degrees of ureteral obstruction, and surgical intervention is often necessary. The embryological significance of the retrocaval ureter is, strictly speaking, an anomaly affecting the inferior vena cava and not the ureter. In the majority of patients, symptoms are due to ureteral obstruction and resulting hydronephrosis. Pain can be intermittent, dull and aching. Haematuria in varying degrees is also present in many cases. They are classed into two types on the basis of radiographic criteria. Type 1, the more common form, has severe or moderate hydronephrosis with extreme medial deviation of the middle ureteral segment, usually medial to the pedicle or across the midline at the L3 level. An "S" or "fish hook" deformity is present at the point of obstruction (Fig.11). In type 2 there is mild hydronephrosis and less medial deviation of the ureter. The ureter is noted to be sickle shaped at the level of obstruction (Fig.12). There have been reports of various complications and associations with retrocaval ureters and most of these have implications regarding its possible medical, radiological and surgical management.

**Anomalies of IVC associated with horseshow kidney**

Horseshoe kidney results from the fusion of metanephric buds between weeks 4 and 8 of embryogenesis, blocking their cephalic migration and normal rotation. The incidence of IVC anomalies with horseshoe is higher than that reported in the general population. Because the embryogenesis of the renal parenchyma and its drainage system in IVC occur simultaneously during gestational weeks 4-10, it is plausible that horseshoe kidney and abnormal IVC are the consequence of a shared disturbed signal during the development of these retroperitoneal structures. IVC anomalies are associated with horseshoe kidney includes double IVC (Fig. 13), left IVC (Fig. 14) and preismic IVC (Fig. 15). Right retrocaval ureter is associated with preismic IVC. Almost preismic IVC is associated with retrocaval ueter. The aetiology of retrocaval ureter is that there is an
anomalous infrarenal IVC because of persistence of either the right posterior cardinal or the right subcardinal venous supply.

**Azygos continuation of IVC** 2, 3, 13, 14

Azygos continuation of IVC has been termed absence of the hepatic segment of the IVC with azygos continuation. The embryonic event is theorized to be failure to from the right subcardinal-hepatic anastomosis, with resulting atrophy of the right subcardinal vein. Consequently, blood is shunted from the supracardinal anastomosis through the retrocrural azygos vein, which is partially derived from the thoracic segment of the right supracardinal vein. The prevalence is 0.6%. The renal portion of IVC receives blood return from both kidneys and passes posterior to the diaphragmatic crura to enter the thorax as the azygos vein (Fig). The azygos vein joins the SVC at the normal location in the right parayrtacheal space and almost ayzgos vein arch is enlarged. The hepatic segment (often termed the posthepatic segment) is ordinarily not truly absent; rather, it drains directly into the right atrium.

Azygos continuation of IVC is predominantly associated with severe congenital heart disease, asplenia or polysplenia syndromes (Fig. 16, 17) and heterotaxy syndrome (Fig. 18).

**Development of renal vein** 2, 15

The development of the renal vein is a part of the complex deneomental process of the IVC. The process starts from the fourth week of conception and ends at about the eighth week. The IVC is formed from vast network of three pairs of pararell veins in communication. During the development of the IVC, there are ananotootic communications between the subcardinal and supracardinal channels that form a collar of veins encircling the aorta. The ventral portion of the circumaortic collar persists as the normal left renal vein.

Left renal vein anomalies are generally classified into four types. In type I, the ventral preaortic limb of the left renal vein is obliterated, but the dorsal retroaortic limb persists and joins the IVC in the orthotopic position. Type II results from the obliteration of the ventral preaortic limb of the left renal vein, and the remaining dorsal limb turns into the RLRV. The left renal vein lies at the level of L4 to L5 and joins the gonadal and ascending lumbar veins before joining the IVC. Type III is the circumaortic left renal vein or venous collar. This type is due to the persistence of subsupracardial and intersupracardial anastomoses and the dorsal limb of the left renal vein. In type IV, the ventral preaortic limb of the left renal vein is obliterated, and the remaining dorsal limb becomes the retroaortic left renal vein. Then, the retroaortic left renal vein courses obliquely and caudally behind the aorta to join the left common iliac vein.

**Circumaortic left renal veins (Circumaortic venous ring)** 13, 15-17

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Circumaortic venous ring results from persistence of a segment of the left supracarcinal vein and of a midline anastomosis, dorsal to the aorta; this dorsal anastomotic vessel, along with normally placed left renal vein, forms a ring or collar about the aorta, with a preaortic and retroaortic limb. When the midline anastomosis is at level of the left renal vein, the "ring" configuration is clear; however, this midline anastomosis can occur at a cranial distance caudally due to retention of a longer segment of the left supracardinal vein (Fig.19). The left renal vein often communicates with the retroperitoneal veins including the lumbar, ascending lumbar, and hemiazygos veins. Occasionally difficult to determine whether a small vein connected to the left renal vein coursing posterior to the aorta and draining into the inferior vena cava should be defined as circumaortic renal vein or as communication of a retroperitoneal vein, such as the lumbar vein or ascending lumbar vein, to the left renal vein on CT. Circumaortic aortic left renal is the cause of hematuria, varicocele, and ureteropelvic junction obstruction (Fig.20).

**Retroaortic left renal vein** \(^{13, 15-17}\)

Retroaortic left renal vein arises from persistent intersupracardinal anastomosis flowing dorsally with respect to the aorta, with the regression of intersupracardinal anastomosis, which normally give rise to the left renal vein in its correct position (Fig. 21). Incidence of retraortic left renal vein is 3.7-8.7%.

The most common type of retroaortic left renal vein was type I. Multiple retroaortic veins are not rare (Fig. 22).

Retroaortic left renal vein is usually asymptomatic. Compression of the retroaortic left renal vein between the aorta and the vertebra is known to be the cause of urological problems such as hematuria, varicocele, and ureteropelvic junction obstruction.

**Anomaly of IVC and renal veins for IVR** \(^{18, 19}\)

The safety of the placement technique and the efficacy of thee implanted device are directly related to the anatomy of each individual patient. It is important to document the number, location, and size of entering renal veins and collateral or potential collateral pathways of veins before filter placement. The presence of undetected anomalies may have been responsible for some recurrent pulmonary emboli reported in previous studied of vena caval filters. Large accessory renal veins or retroaortic left renal veins are essentially no different from main renal veins in that positioning of the filter legs of certain types inthese veins in these veins can result in tilting of the filter and possible loss of filtration efficacy. With respect to location, renal veins closer in distance than one vertebral height are close enough together that it would be very difficult in most circumstances to place a filter between them.

Anomalies of IVC have been recognized as possible risk factor for deep vein thrombosis, particularly in young adults. Regarding the pathophysiology of deep vein thrombosis in individuals with an anomaly of IVC, blood return may be inadequate in spite of prominent
collaterals. This inadeguate blood return may increase the blood pressure in the veins of the lower extremities, with ensuing venous stasis and subsequent deep vein thrombosis.
Fig. 0: Fig. 1 Left IVC CT images show that infrarenal IVC located in left side of aorta. RIVC: right IVC, Ao: aorta, LRV: left renal vein

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**Fig. 0:** Fig. 2 Venous thrombus associated to central venous catheterization Central venous catheter (yellow arrows) is inserted from right femoral vein into left IVC, and venous thrombus (red arrows) is noted adjacent to catheter top.

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Fig. 3: Fig. 3 Schema of pelvic venous variations. Pelvic venous variations of IVC anomalies are classified into 5 types.

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**Fig. 0:** Fig. 4 Double IVCs with no interiliac communication CT images show ascending aorta between double IVCs without interiliac communication. RIVC: right IVC, LIVC: left IVC, RCIV: right common iliac vein, LCIV: left common iliac vein

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Fig. 0: Fig. 5 Double IVCs (type 2) CT images show double IVCs with interiliac communication from left CIV. RIVC: right IVC, LIVC: left IVC, LCIV: left common iliac vein, CV: communicated vein (red arrows)

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Fig. 6: Double IVCs (type 4) CT images show double IVCs with interiliac communication from left internal iliac vein. RIVC: right IVC, LIVC: left IVC, LCIV: left common iliac vein, RCIV: right common iliac vein, CV: communicated vein (red arrows), LIIV: left internal iliac vein

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Fig. 0: Fig. 7 Double IVCs (type 5) CT images show double IVCs with interiliac communication from right internal iliac vein. RIVC: right IVC, LIVC: left IVC, LCIV: left common iliac vein, RCIV: right common iliac vein, CV: communicated vein (red arrows), RIIV: right internal iliac vein

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Fig. 0: Fig. 8 Dilated left ovarian vein due to Nut cracker syndrome CT images show left renal vein stenosis (black arrow) between aorta and superior mesenteric artery and dilated left ovarian vein (blue arrows).

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Fig. 0: Fig. 9 Dilated ovarian veins Axial and coronal CT images show marked dilated ovarian veins bilaterally.

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Fig. 0: Fig. 10 Suprarenal IVC filter placement for left ovarian venous thrombus. Compression of bilateral ovarian vein due to a large myoma uteri is noted before operation. Post operative CT image shows left ovarian venous thrombus (red arrow). IVC filter is placed to suprarenal IVC.

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**Fig. 0**: Fig. 11 Retrocaval ureter (type 1) Venography after contrast enhanced CT shows S-shaped dilatation of retrocaval ureter. Operation of retrocaval ureter was performed, however hydonephrosis was noted due to postoperative scar on axial CT images. RU: retrocaval ureter

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**Fig. 0:** Fig. 12 Retrocaval ureter (type 2) Contrast enhanced CT images show right ureter runs behind IVC. Note mild stretched retrocaval ureter on CT urography. U: retrocaval ureter

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**Fig. 0:** Fig. 12 Retrocaval ureter (type 2) Contrast enhanced CT images show right ureter runs behind IVC. Note mild stretched retrocaval ureter on CT urography. U: retrocaval ureter

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**Fig. 14**: Double IVCs associated with horseshoe kidney CT images show double IVCs behind horseshoe kidney.

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Fig. 0: Fig.15 Preisthmic IVC associated with horseshoe kidney CT images show preisthmic IVC and retrocaval ureter. CT urography shows stenosis of right retrocaval ureter (yellow arrow) and mild hydronephrosis. RU: retrocaval ureter

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**Fig. 0:** Fig. 16 IVC interruption azygos continuation associated with polysplenia CT images show dilated azygos arch and vein in right side. Note polysplenia in left upper abdomen (blue circle).

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Fig. 0: Fig. 17 IVC interruption azygos continuation associated with polysplenia and situs ambiguous CT images show dextracardia, right aortic arch, dilated azygos arch in left side and symmetric liver. Intrahepatic segment of IVC (white arrow) is present.

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**Fig. 0:** Fig. 18 IVC interruption azygos continuation associated with polysplenia and heterotaxy syndrome CT images show dilated azygos arch and vein in right side, spolysplenia, symmetric liver, right sided stomach and left sided gall bladder. Both lungs have two lobes.

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**Fig. 0:** Fig. 19 Circumaortic left renal veins CT images show that upper left renal vein runs in front of aorta and lower left renal vein runs behind aorta consistent with circumaortic venous ring. LRV: left renal vein

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Fig. 0: Fig. 20 UP junction stenosis due to "circumaortic left renal veins" CT images show cicumaotic left renal veins and UP junction stenosis due to a lower renal vein. A lower left renal vein runs behind left common iliac artery and inserts into common iliac vein. CT urography show left hydronephrosis and UP junction stenosis. U:ureter, IMV: inferior mesenteric vein

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**Fig. 0:** Fig. 21 Retroaortic left renal vein CT images show one left renal vein runs behind aorta and inserts into right IVC (blue arrows)

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Fig. 0: Fig. 22 Multiple retroraortic left renal veins CT images show three retroaortic left renal veins. LCIV: left common iliac vein

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Conclusion

Knowledge of pathogenetic venous variation and recognition of radiological findings are beneficial for successful interventional radiology. Thin slice axial and multi planar reformatted CT images reveal venous variations accurately. Thrombotic complications are common and sepsis associated with catheter-related infections is serious on CVC to these various veins. Placement of IVC filter correctly is important to prevent pulmonary embolism in patients with congenital anomalies.
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