

Various congenital anomalies of the inferior vena cava: review of cross-sectional imaging findings and report of a new variant

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Abstract

The inferior vena cava (IVC) is an important structure receiving a large amount of venous return and is associated with various congenital disorders. Advances in diagnostic imaging and its increasing accessibility have led to an increase in the incidental detection of IVC anomalies. Congenital anomalies of the IVC are not uncommon and are occasionally critical to treatment planning. However, they are frequently overlooked in abdominal imaging. The IVC is composed of four segments (intrahepatic, suprarenal, renal, and infrarenal), and each segment arises from different embryonic structures in a complex process. Anomalies of the IVC can be classified according to the involved segment. Familiarity with the variety of IVC anomalies seen on imaging is vital for correctly diagnosing and managing patients in daily practice.

Key words: Vena cava, inferior—Congenital abnormalities—Tomography, x-ray computed

Abbreviations

IVC	Inferior vena cava
CT	Computed tomography

Anomalies of the inferior vena cava (IVC) are not uncommon, existing in up to 8.7% of the population [1]. The embryogenesis of the IVC is complex; it develops between the 6th and 8th weeks after conception [2]. With the increased use of cross-sectional imaging, various anomalies of the IVC are frequently encountered in

clinical practice and can be misdiagnosed as mass-like lesions or lymph nodes [3]. Although most patients with congenital anomalies of the IVC are asymptomatic, some could present with symptoms or abnormal laboratory findings. It is especially important to recognize an IVC anomaly prior to vascular surgery or other interventional procedures because unexpected complications can arise [4–6]. For example, in patients with double IVC, an IVC filter should be inserted into the suprarenal IVC. Otherwise, pulmonary embolism could recur after IVC filter insertion [7]. Radiologists must therefore be familiar with IVC anomalies to correctly interpret them on cross-sectional images.

In this article, we describe the embryogenesis of the IVC and present imaging findings of various congenital anomalies, including interruption of the IVC with azygous continuation, retroaortic left renal vein, circum-aortic left renal vein, retrocaval ureter, left-sided IVC, double IVC, and preaortic iliac confluence. In addition, we report one new variant: left IVC with hemiazygos continuation, retroaortic right renal vein, and the presence of a right suprarenal IVC.

Imaging techniques

Computed tomography (CT) is the most common imaging modality for initial identification of IVC anomalies. The portal venous phase (60–70 s after intravenous administration of 120–150 mL of contrast material at a rate of 3–5 mL/s) is widely used to evaluate IVC, although delaying the image after contrast material administration to 70–90 s shows uniform enhancement of the entire IVC [8]. To evaluate an IVC-anomaly-associated ureter, the excretory phase (12–15 min after administration of contrast material) is useful [8]. Multi-

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planar reformatted CT images can help practitioners recognize complex IVC anomalies. Magnetic resonance imaging can also be used to detect IVC anomalies without radiation exposure, making it especially valuable for pediatric patients [8, 9].

Embryonic development of the IVC

The IVC is composed of four segments (intrahepatic, suprarenal, renal, and infrarenal), and each segment arises from a different embryonic structure [1, 9]. During embryologic development, three pairs of cardinal veins (subcardinal, supracardinal, and posterior cardinal) run along each side of the aorta, and those veins form the segments of the IVC [1, 8, 9]. The vitelline vein arises from a capillary plexus of the yolk sac and develops into the intrahepatic segment. The subcardinal and supracardinal veins form the suprarenal, renal, and infrarenal segments of IVC. The subcardinal and supracardinal veins are ventromedial and dorsomedial to the posterior cardinal veins, respectively. Normally, the right vitelline, right subcardinal, and right supracardinal veins form the IVC, and the left subcardinal and supracardinal veins regress during embryogenesis. The azygos and hemiazygos veins are derived from the supracardinal veins in the thoracic region. Several midline anastomotic channels occur between the paired veins: the inter-subcardinal, inter-supracardinal, and inter-posterior cardinal anastomoses. Among these anastomotic channels, the inter-subcardinal vein develops into the left renal vein [1, 2, 7, 9]. Both iliac veins arise from the posterior cardinal veins, and the inter-posterior cardinal anastomosis forms the common iliac venous confluence posterior to the aorta in the pelvis (Table 1, Fig. 1) [9–11].

Anomalies of the IVC

Anomalies of the IVC can be classified according to the involved segment; each will be discussed in turn. Most IVC anomalies are related to the left renal vein or double IVC.

Anomaly of the suprarenal segment

Interruption of the IVC with azygos continuation

Azygos continuation of the suprarenal IVC results from a failure to develop the right subcardinal-hepatic anas-

tomosis (Fig. 2A). This anomaly, termed absence of intrahepatic IVC, has a prevalence of 0.6% and is attributable to atrophy of the right subcardinal vein [6, 9]. The suprarenal segment of the IVC continues posteriorly to the diaphragmatic crura and subsequently enters the thorax as the azygos vein [12]. Elongated azygos and hemiazygos veins can be misidentified as lymphadenopathy [13]. Interruption of the IVC with azygos continuation is also commonly associated with heterotaxy syndrome (Fig. 2) [14].

Anomalies of the left renal vein

Retroaortic left renal vein

The prevalence of a retroaortic left renal vein is as high as 3.4% [12]. This anomaly results from a persistent inter-supracardinal anastomosis with regression of the inter-subcardinal anastomosis (Fig 3A). The inter-subcardinal anastomosis is positioned in front of the aorta and normally forms the left renal vein. In contrast, the inter-supracardinal anastomosis goes behind the aorta and typically develops into the retroaortic left renal vein [7] (Fig. 3). Two or more retroaortic left renal veins can exist (Fig. 4). It is important to recognize a retroaortic left renal vein before performing a left nephrectomy.

Circumaortic left renal vein (circumaortic venous ring)

A circumaortic left renal vein arises from the persistence of both the inter-subcardinal and inter-supracardinal anastomoses (Fig. 5A). Thus, two or more left renal veins are located ahead of and behind the aorta, forming a ring-like appearance. The retroaortic renal vein is derived from the inter-supracardinal anastomosis and is caudally located with respect to the preaortic renal vein, which arises from the inter-subcardinal anastomosis [12] (Fig. 5). The cranially located preaortic vein receives the left adrenal vein, and the caudally positioned retroaortic vein receives the left gonadal vein [9]. Posterior nutcracker syndrome can occur when a persistent posterior branch is compressed between the aorta and a vertebral body [15].

Anomalies of the infrarenal segment

Retrocaval ureter (circumcaval ureter)

A retrocaval ureter is an uncommon anomaly that presents with the ureter partially encasing the IVC. In this condition, the infrarenal segment of the IVC consists of the right posterior cardinal vein instead of the right supracardinal vein (Fig. 6A). In contrast to the supracardinal vein, which is located posterior and medial to the ureter, the posterior cardinal vein lies anterior and lateral to the ureter. As a result, the ureter goes along the posterior and medial aspect of the IVC and partially encases

Table 1. Vessels and their normal embryologic origins

Vessels	Embryologic origin
Intrahepatic IVC	Vitelline vein
Suprarenal/renal IVC	Right subcardinal vein
Left renal vein	Inter-subcardinal anastomosis
Infrarenal IVC	Right supracardinal vein
Azygos/hemiazygos veins	Thoracic supracardinal veins
Both common iliac veins	Pelvic posterior cardinal veins
Common iliac venous confluence	Inter-posterior cardinal anastomosis

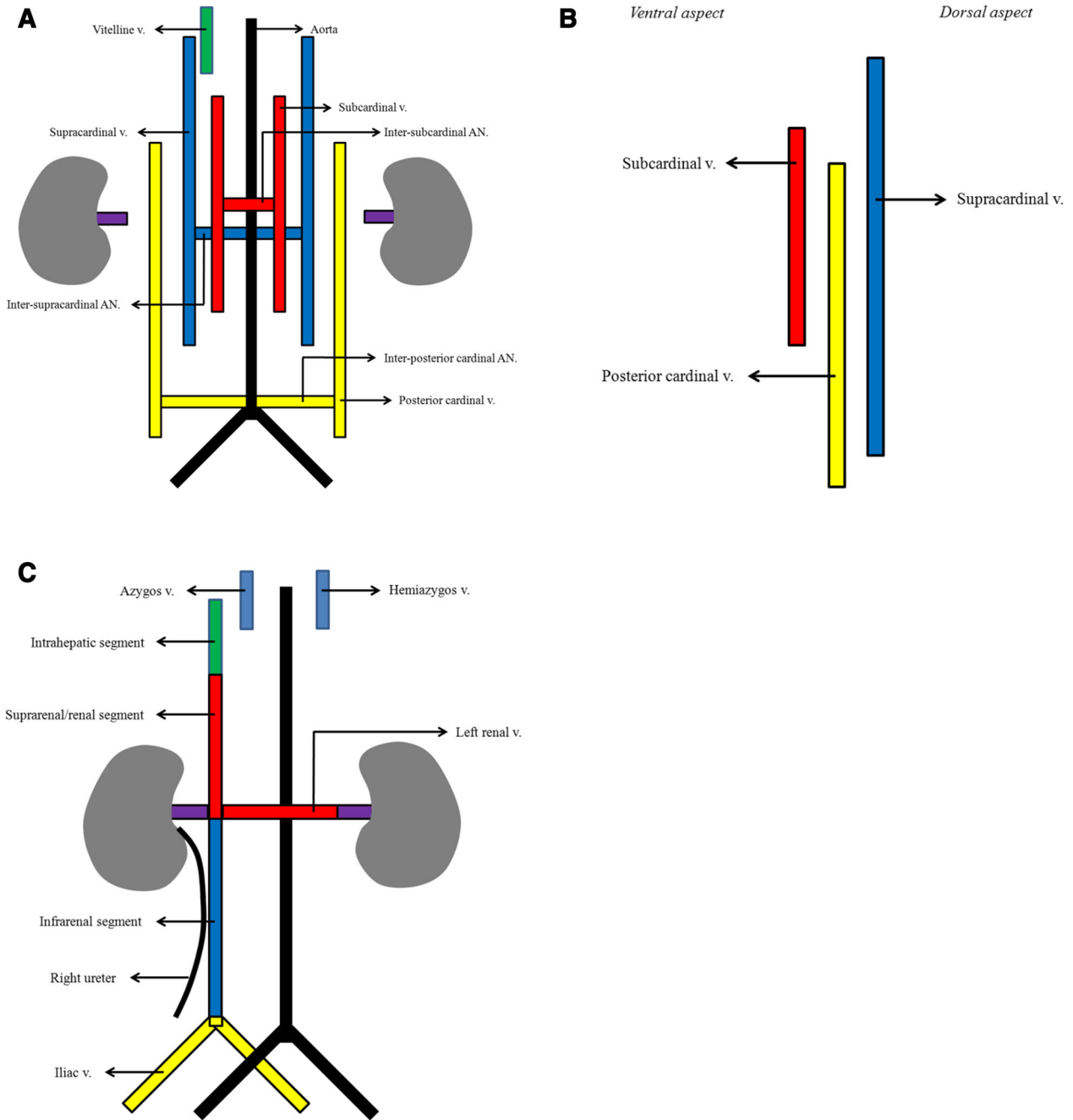
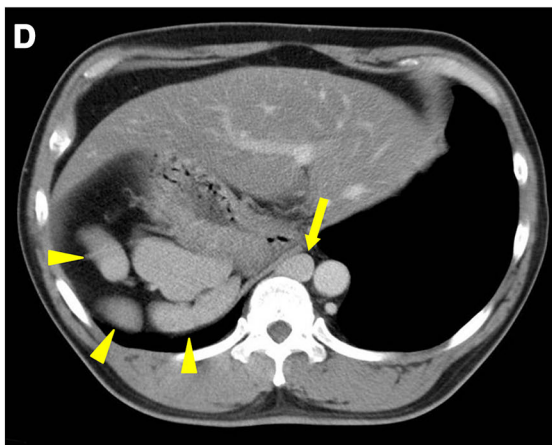
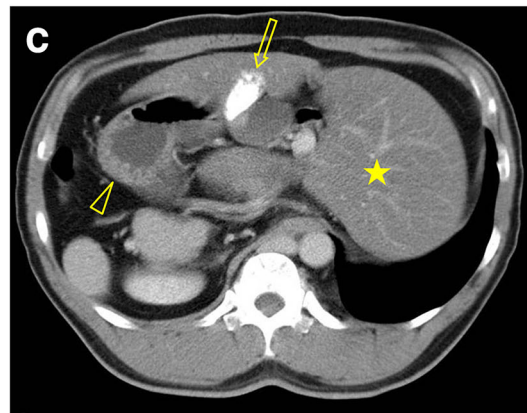
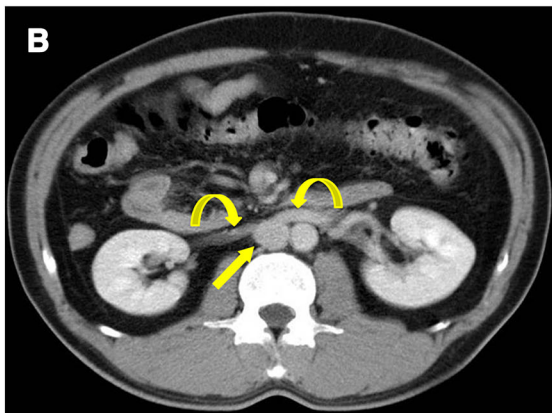
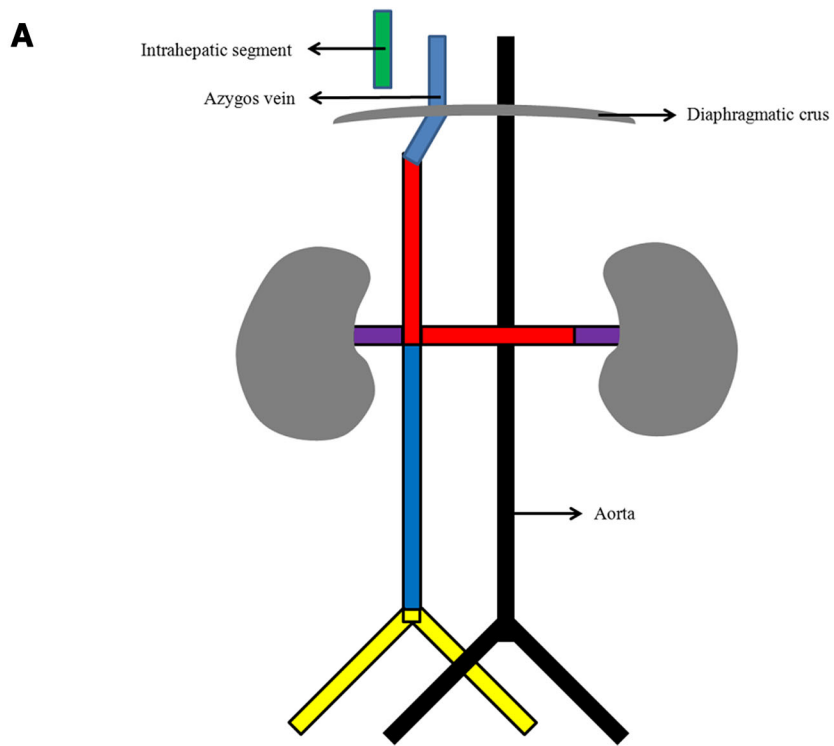


Fig. 1. Illustration of the development of the IVC. **A** Ventral view of the embryonic vessels. Three paired embryonic vessels develop on each side of the aorta (black): the subcardinal (vertical red), supracardinal (vertical blue), and posterior cardinal (vertical yellow) veins. Both posterior cardinal veins are located lateral to the sub- and supracardinal veins. There are anastomoses between the paired vessels: inter-subcardinal (horizontal red), inter-supracardinal (horizontal blue), and inter-posterior cardinal (horizontal yellow). The inter-subcardinal and inter-supracardinal anastomoses are located at the level of the renal hilum. The inter-supracardinal anastomosis is caudally located with respect to the inter-subcardinal anastomosis. **B** Sagittal view of the embryonic vessels. The subcardinal vein (red) is the most ventrally located, and

the supracardinal vein (blue) is the most dorsal. The posterior cardinal vein (yellow) is positioned between the sub- and supracardinal veins. **C** Schematic of a normal IVC. The intrahepatic segment arises from the vitelline vein (green). The suprarenal and renal segments derive from the right subcardinal vein (red). The right supracardinal vein (blue) forms the infrarenal segment, and both thoracic supracardinal veins (blue) develop into the azygos and hemiazygos veins. Both iliac veins originate from the posterior cardinal veins (yellow). Among the anastomotic channels, the inter-supracardinal anastomosis regresses, and the inter-subcardinal and inter-posterior cardinal anastomoses develop into the left renal vein and common iliac venous confluence, respectively. V vein, AN anastomosis.



◀ **Fig. 2.** IVC interruption with azygos continuation in a 45-year-old man. **A** Schematic shows discontinuation between the intrahepatic and suprarenal segments of the IVC. The suprarenal IVC continues up to the azygos vein. **B–D** Axial CT scans from caudal to cranial show heterotaxy syndrome with an IVC anomaly. **B** The IVC (arrow) at the level of the renal hilum is normal and receives both renal veins (curved arrows). **C** The liver (*closed star*) and contrast-filled gallbladder (open arrow) are located in the midline abdomen and the stomach (open arrowhead) is positioned on the right side of the abdomen. **D** The IVC continues cephalad to the enlarged azygos vein (arrow). Note the polysplenia (arrowheads) on the right side of the abdomen.

it [7]. Most often, a retrocaval ureter occurs on the right side [9] (Fig. 6). Recurrent urinary tract infections and ureteral obstruction are frequent in patients with a circumcaval ureter. If necessary, treatment involves surgical relocation of the ureter [7, 9].

Left IVC

A left-sided IVC results from the persistence of the left supracardinal vein with regression of the right supracardinal vein (Fig. 7A). It has a prevalence of 0.2% to

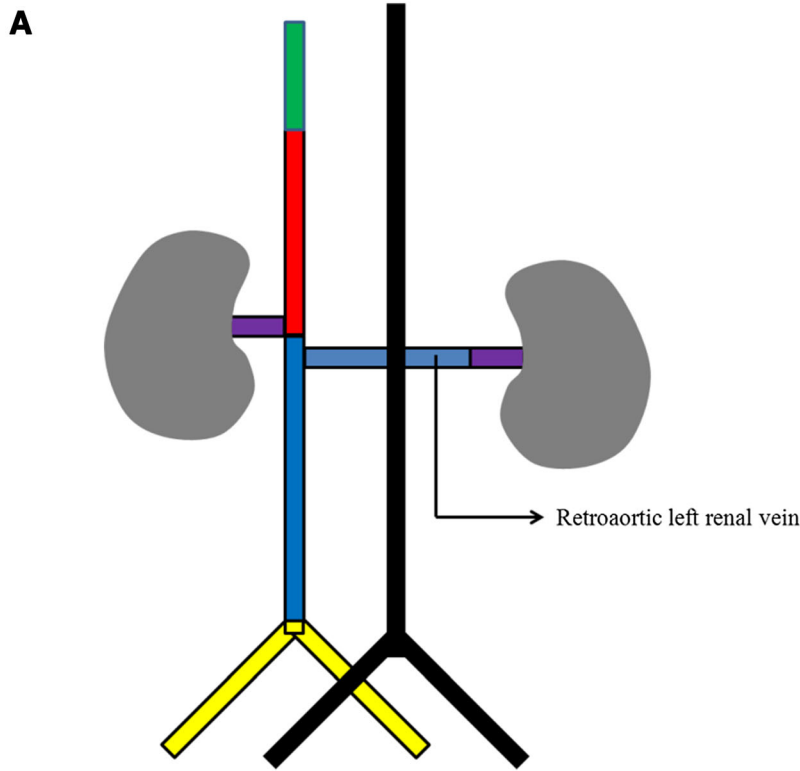


Fig. 3. Retroaortic left renal vein in a 60-year-old man. **A** Schematic shows a left renal vein behind the aorta. **B, C** Curved reformatted CT scans show the left renal vein (arrow) crossing posterior to the aorta.

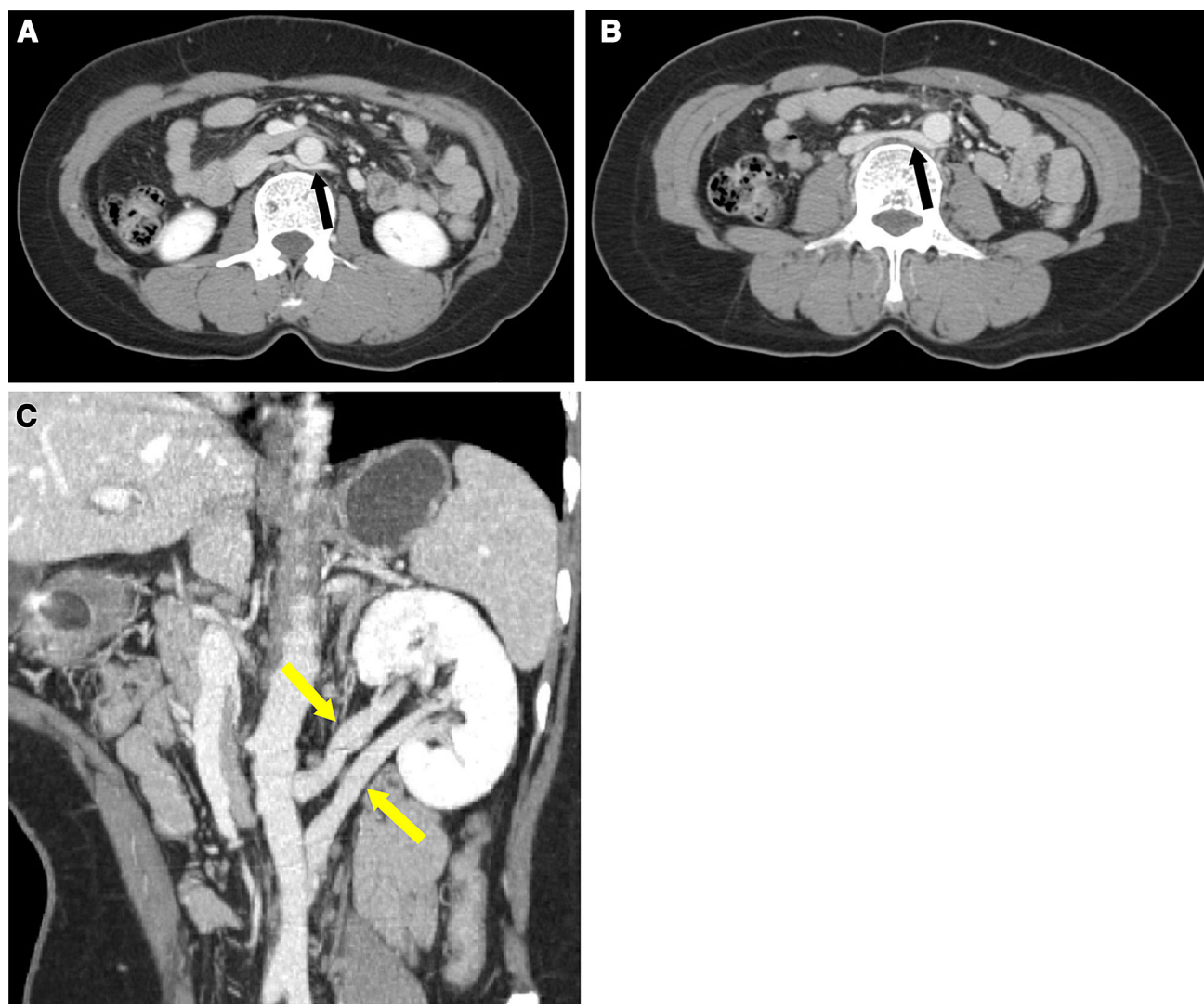


Fig. 4. Two retroaortic left renal veins in a 47-year-old woman. **A, B** Axial CT scans show two left renal veins (arrows) behind the aorta. **C** Curved reformatted CT scan shows the left renal veins (arrows) posterior to the aorta.

0.5% and represents the mirror image of the normal configuration. The left-located IVC ends at the left renal vein, which runs anterior to the aorta and finally forms a normal right-sided suprarenal IVC [9, 16]. Caudally, a left IVC connects with both iliac veins (Fig. 7).

Double IVC

The prevalence of a double IVC might be as high as 3%. This anomaly represents the persistence of both supracardinal veins [2, 9] (Fig. 8A). The IVC along the left side of the aorta joins the left renal vein, as a left IVC would. The right-sided IVC connects to the right common iliac vein, whereas the left-sided IVC continues on to the left common iliac vein (Fig. 8) [7]. Recurrent episodes of pulmonary embolism after IVC filter insertion occur in patients with a double IVC. On cross-sectional imaging,

the left-located IVC can be misdiagnosed as lymphadenopathy.

Anomaly of the common iliac venous confluence

Preaortic iliac confluence

A preaortic iliac confluence is a very unusual anomaly of the IVC. In patients with this anomaly, the iliac vein confluence or the left common iliac vein is located anterior to the aortic bifurcation or right common iliac artery (Fig. 9). The first report of a preaortic iliac confluence was in 1929 [17], but few cases have been described since then [11, 18–20]. During the first trimester of pregnancy, the circumumbilical venous ring surrounds the future common iliac arteries. Normally, the ventral limb of the circumumbilical venous ring regresses,

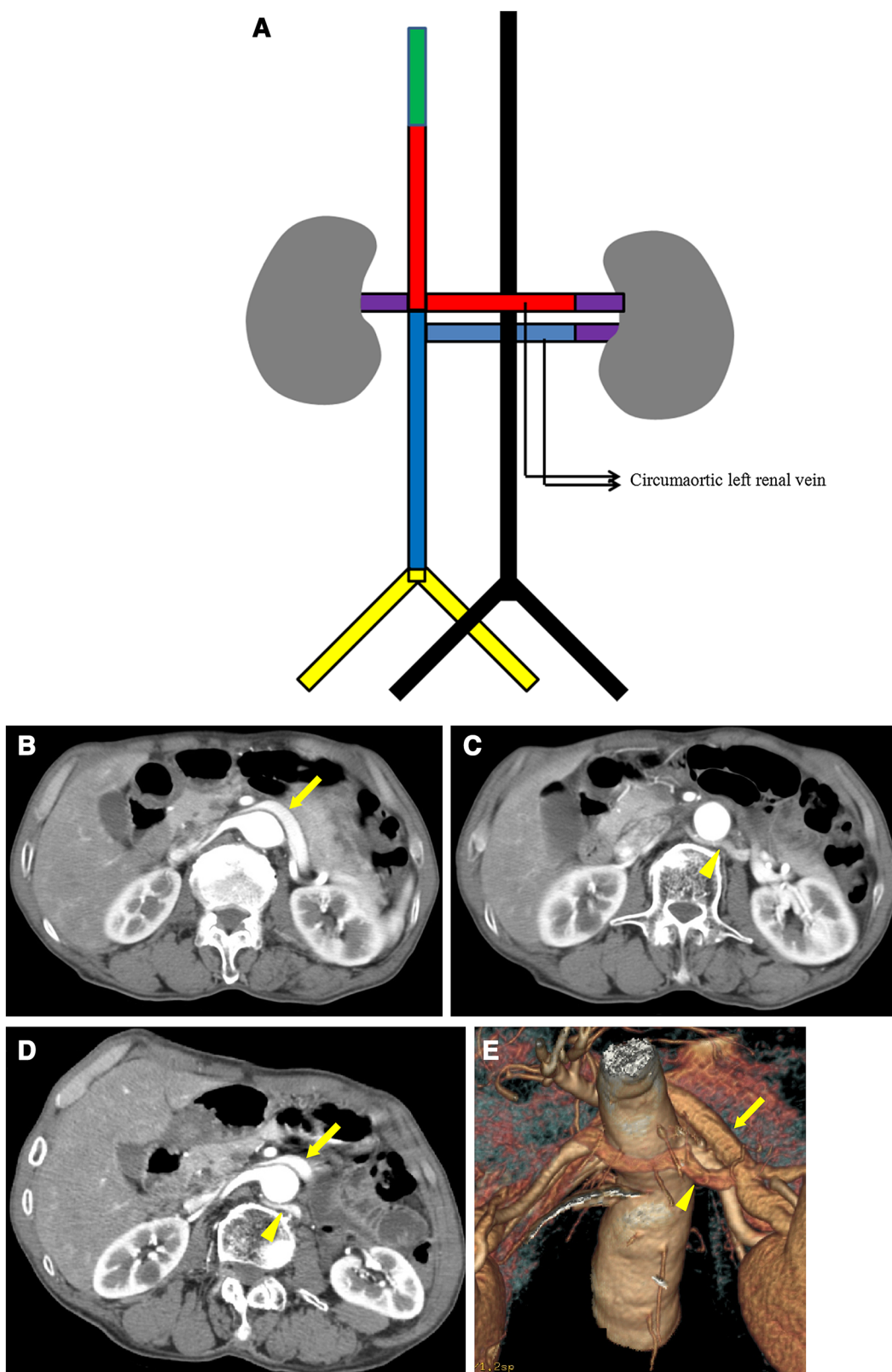


Fig. 5. Circumaortic left renal vein in a 79-year-old man. **A** Schematic shows two left renal veins located ahead of and behind the aorta. **B–D** Axial and curved reformatted CT scans show two left renal veins. The preaortic left renal vein (arrow)

is superior to the retroaortic left renal vein (arrowhead). **E** 3D reconstructed image shows two left renal veins (arrow and arrowhead) encircling the aorta.

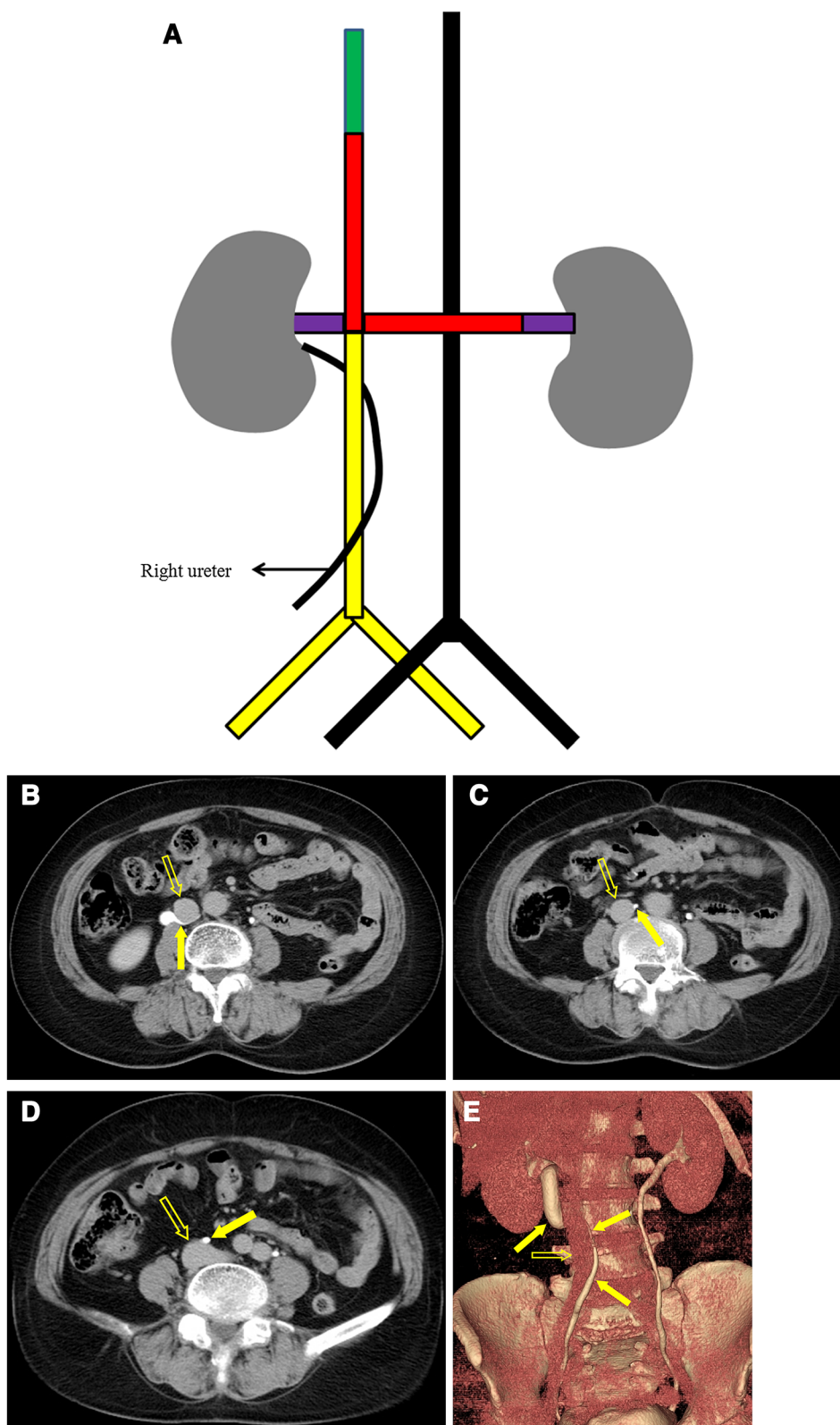


Fig. 6. Retrocaval ureter in a 63-year-old woman. **A** Schematic shows the right ureter encasing the infrarenal segment of the IVC. **B–D** Axial excretory phase CT scans from cranial to caudal show the circumcaval ureter. **B, C** The right ureter (arrow) runs posterior to the IVC (open arrow), and

then runs along the left side of the IVC. **D** Eventually, the ureter (arrow) is located anterior to the IVC (open arrow). **E** 3D reconstructed image shows the retrocaval ureter (arrows) encircling the IVC (open arrow).

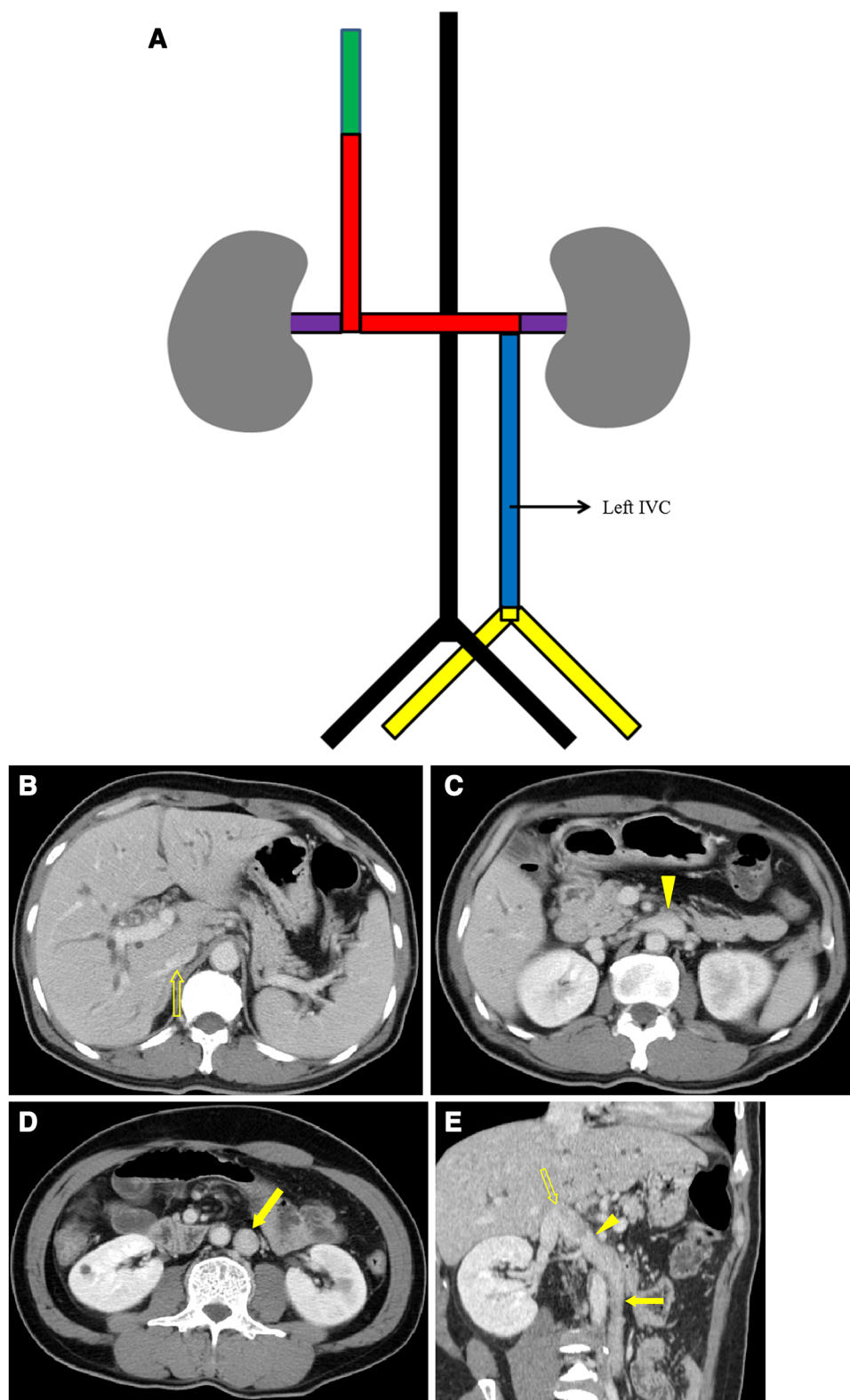


Fig. 7. Left IVC in a 50-year-old man with biliary stones. **A** Schematic shows a left-located infrarenal IVC that terminates at the left renal vein. **B–D** Axial CT scans from cranial to caudal show the left IVC. **B, C** Note the normal right suprarenal IVC (open arrow) and left renal vein (arrowhead). There is dilatation of the bile duct caused by the

biliary stones. **D** The infrarenal segment of the IVC (arrow) is along the left side of the aorta. **E** Curved reformatted image shows the course of the IVC. The infrarenal IVC (arrow) ends at the left renal vein (arrowhead), which crosses anterior to the aorta and continues to the right suprarenal IVC (open arrow).

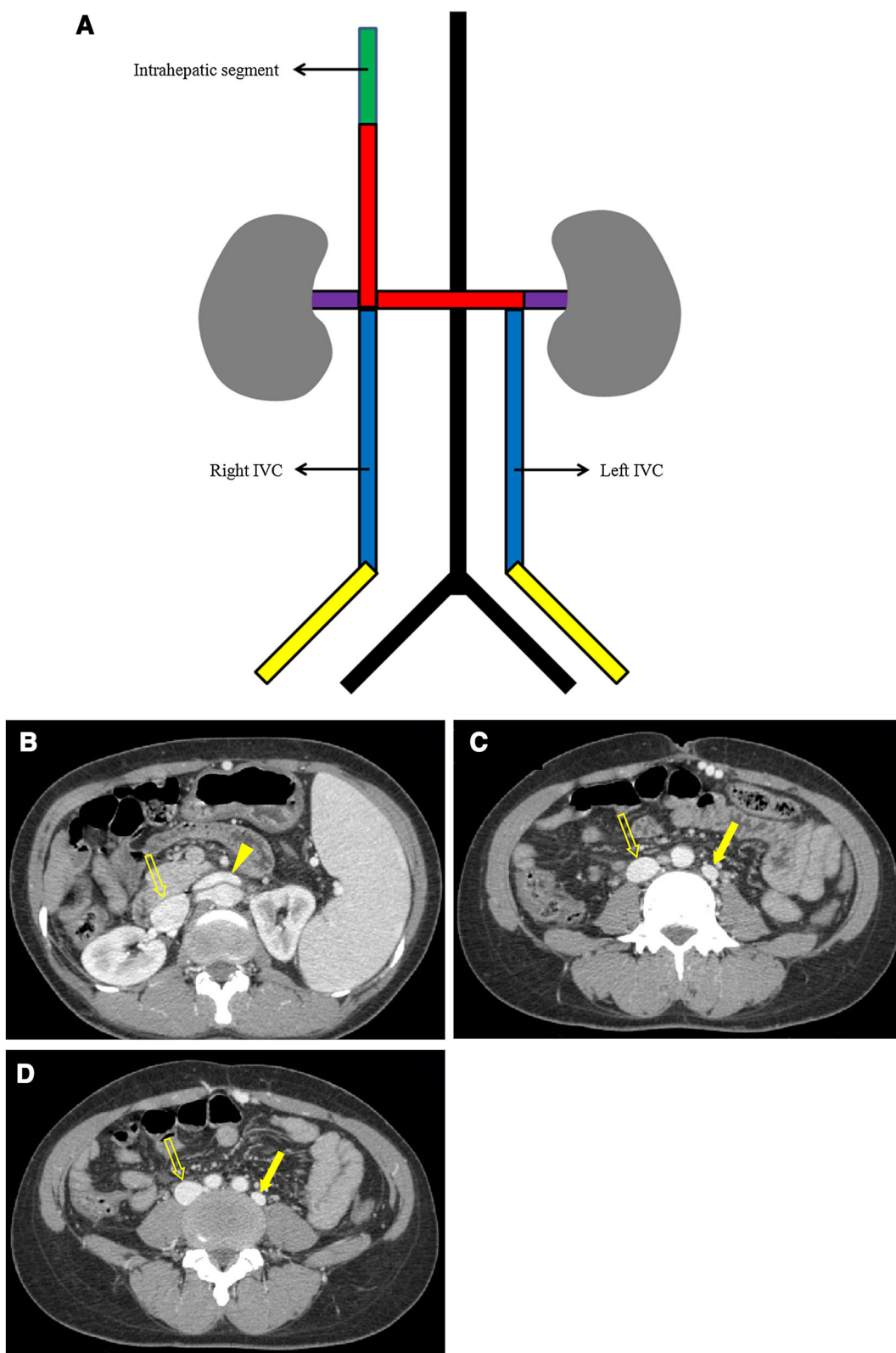


Fig. 8. Double IVC in a 37-year-old woman with liver cirrhosis. **A** Schematic shows two infrarenal IVCs on either side of the aorta. **B–D** Axial CT scans from cranial to caudal show the anomaly. **B** CT scan shows a normal right suprarenal IVC

(open arrow) and left renal vein (arrowhead). **C** CT scan below the kidney shows a right (open arrow) and left (arrow) IVC on either side of the aorta. **D** Each IVC continues to either side of the common iliac veins.

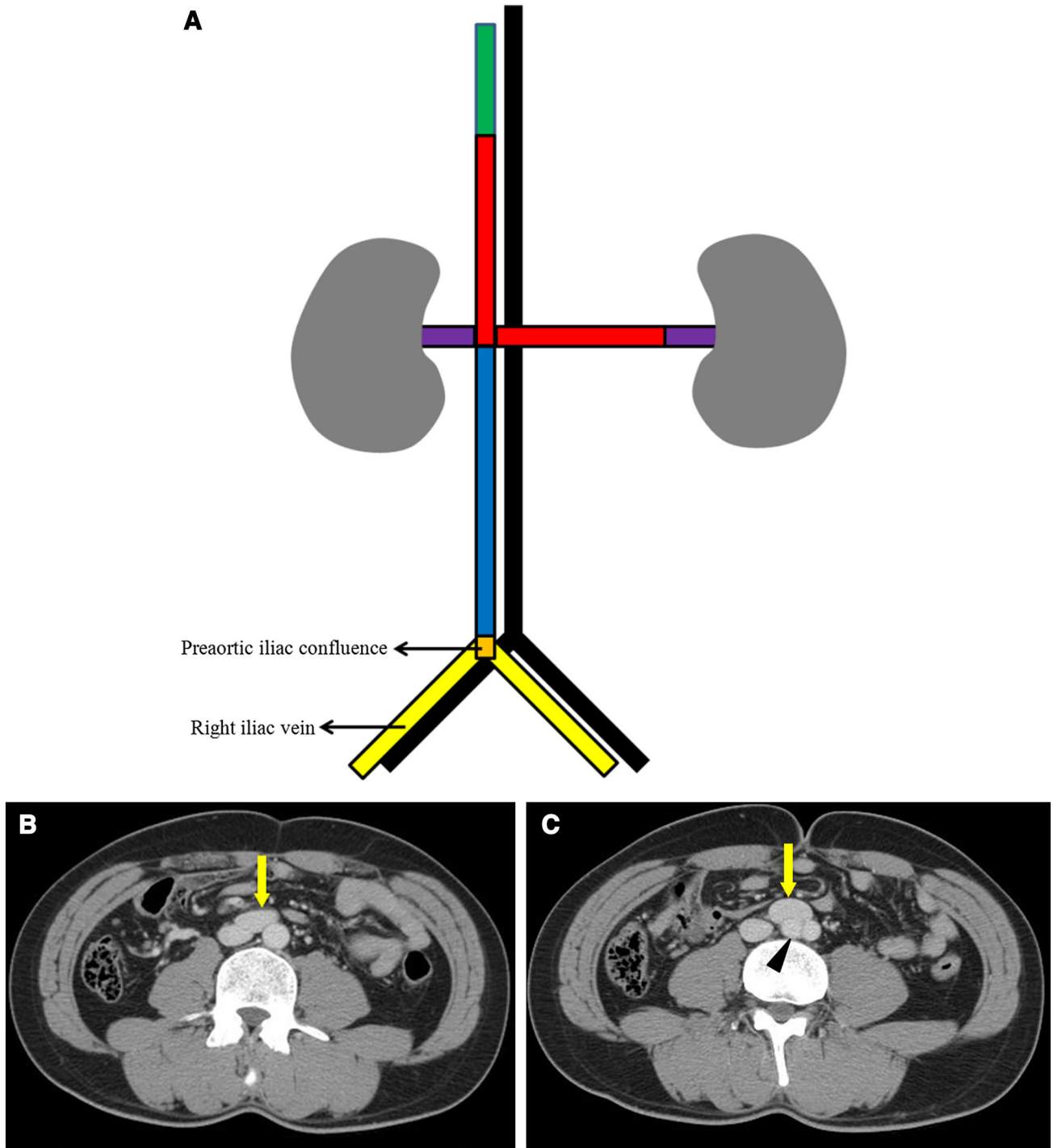


Fig. 9. Preaortic iliac confluence in a 36-year-old man. **A** Schematic shows the common iliac venous confluence (orange box) derived from the ventral limb of the circum-

bilical venous ring in front of the right common iliac artery. **B**, **C** Axial CT scans show the left common iliac vein (arrow) anterior to the right common iliac artery (arrowhead).

whereas the dorsal limb, which consists of the interposterior cardinal anastomosis, forms the common iliac venous confluence behind the aortic bifurcation (Fig. 1A, C). In patients with a preaortic iliac confluence,

however, the dorsal limb of the circumumbilical venous ring is interrupted, and the common iliac venous confluence is derived from the persistent ventral limb, which lies anterior to the aortic bifurcation. [1, 10, 11, 21].

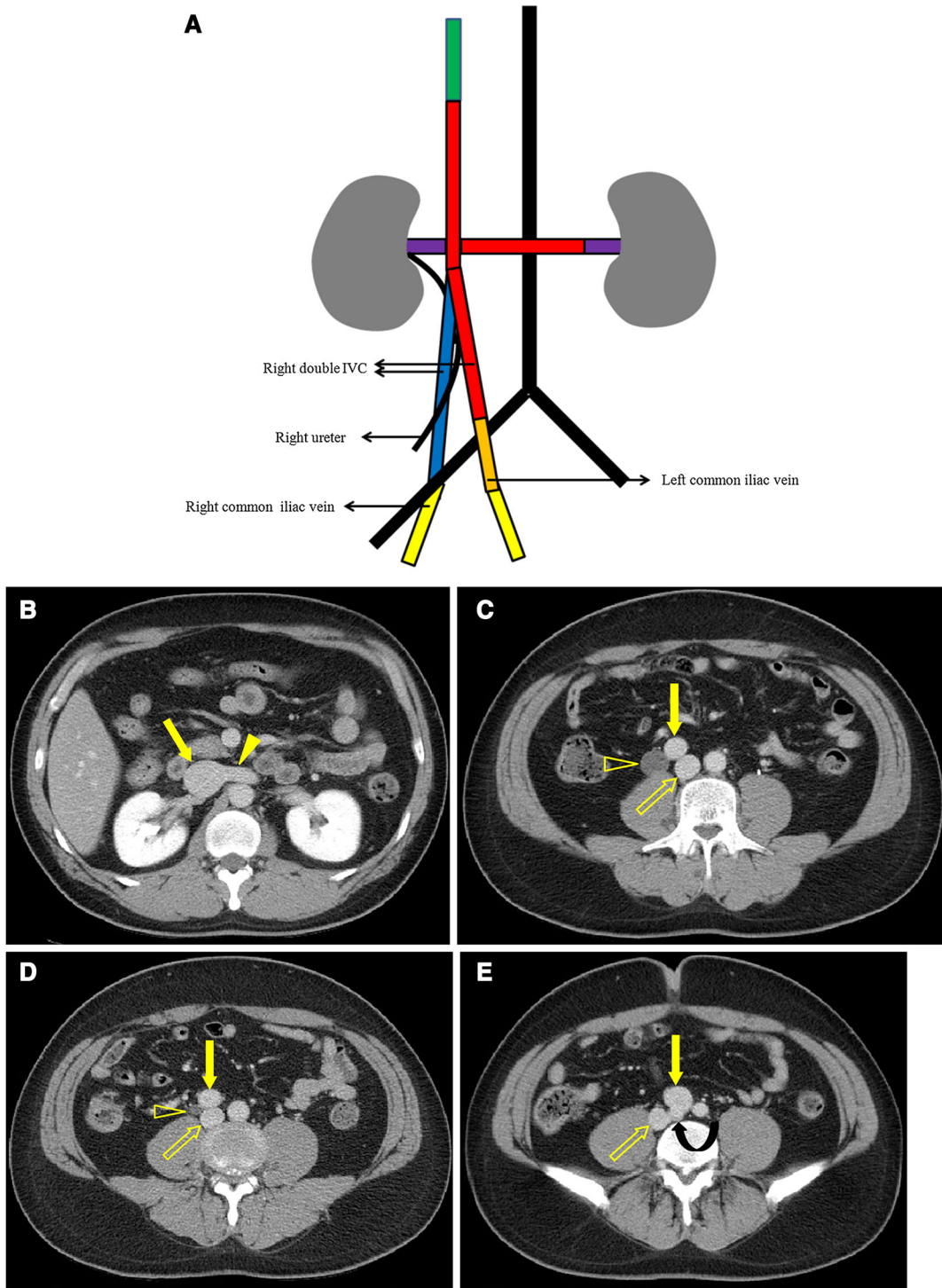


Fig. 10. Right double IVC with retrocaval ureter in a 35-year-old man. **A** Schematic shows a right double IVC with the right ureter encircling the IVC. The right common iliac artery passes between the two common iliac veins. **B–F** Axial CT scans from cranial to caudal show the anomaly. **B** One IVC (arrow) receives the left renal vein (arrowhead) at the level of the renal hilum. **C** The infrarenal IVC is separated into two right IVCs (arrow and open arrow). Note the dilated right ureter (open arrowhead). **D** The right ureter (open arrowhead) runs between the two infrarenal IVCs (arrow and open arrow). **E**,

F The ventral IVC continues to the left common iliac vein (arrow) in front of the right common iliac artery (curved arrow), and the dorsal IVC connects to the right common iliac vein (open arrow). Note the right common iliac artery (curved arrow) between the two common iliac veins (arrow and open arrow). **G** Curved reformatted image shows the right double IVC with a ventral (arrow) and dorsal (open arrow) relationship. **H** Curved reformatted image shows the dilated right ureter (open arrowhead) passing between the right double IVCs (arrow and open arrow).

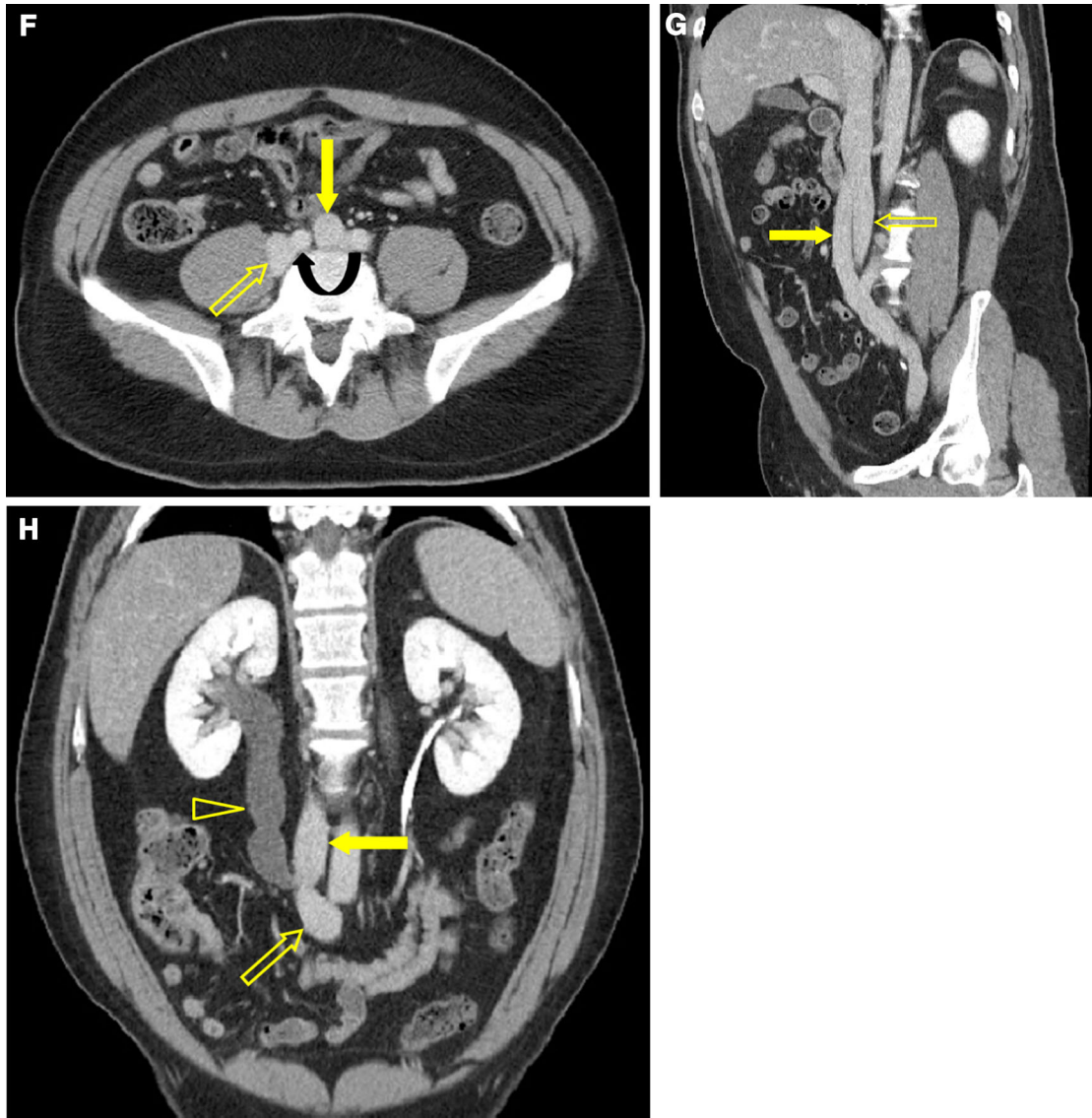


Fig. 10. continued.

While uncommon in humans, a preaortic iliac confluence is normal in marsupials; thus, this anomaly is often called a “marsupial cava” [11].

Complex anomalies

More than one IVC anomaly can coexist in a patient. Complex anomalies of the IVC are often associated with congenital disorders.

Right double IVC with retrocaval ureter and preaortic left iliac vein

A right double IVC is an extremely rare anomaly that occurs when two infrarenal IVCs lie to the right of the aorta. Doyle et al. [22] first described ipsilateral dupli-

cation of the IVC, and since then, only around a dozen cases have been reported [23–28]. Most cases of a right double IVC have shown a ventral and dorsal relationship between the two vessels. Nagashima et al. [25] suggested a theory for the development of a right double IVC: the right sub- and supracardinal veins both persist (Fig. 10A). The IVC derived from the right subcardinal vein is located ventrally, and the IVC arising from the right supracardinal vein is positioned dorsally. The ventral limb of the circumumbilical venous ring forms the left common iliac vein, similar to a marsupial cava, and connects with the ventral IVC anterior to the right common iliac artery. The dorsal-sided IVC continues to the right common iliac vein, passing behind the right common iliac artery. As a result, the right common iliac

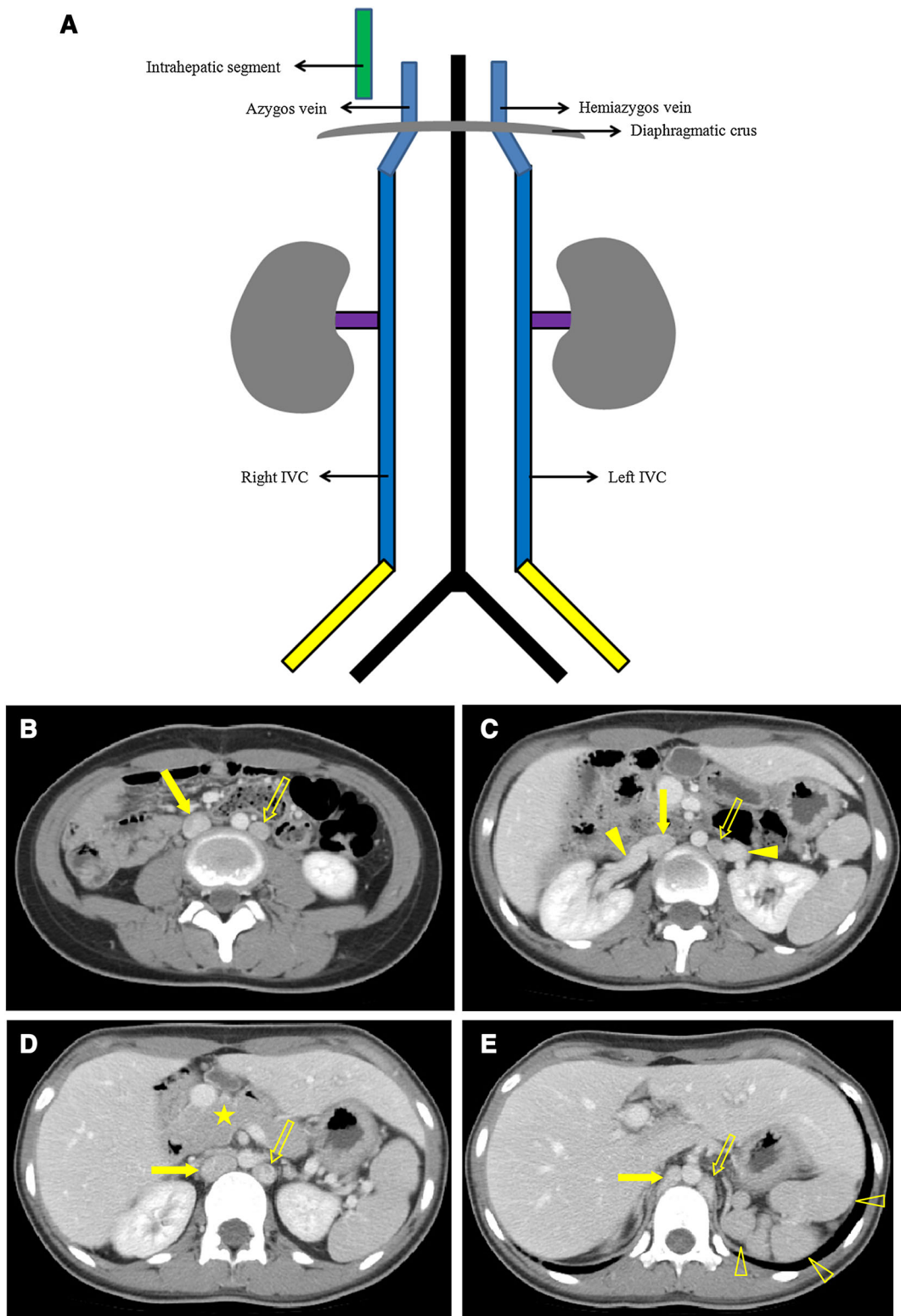


Fig. 11. Double IVC interruption with azygos and hemiazygos continuation in a 17-year-old woman. **A** Schematic shows a double IVC continuous with the azygos and hemiazygos vein. There is no anastomosis between the IVCs below the diaphragm. **B–F** Axial CT scans from caudal to cranial show situs ambiguous with the IVC anomaly. **B** There are two infrarenal IVCs (arrow and open arrow), one on each side of the aorta. **C** Both IVCs (arrow

and open arrow) receive the ipsilateral renal veins (arrowheads), but there is no midline-crossing renal vein. **D–F** Each IVC continues to the azygos and hemiazygos veins (arrow and open arrow), one on each side of the aorta. Note the short pancreas (*closed star*) and polysplenia (open arrowheads). **G, H** Barium studies show intestinal malrotation with a duodenum that does not cross the midline and an abnormally positioned colon.

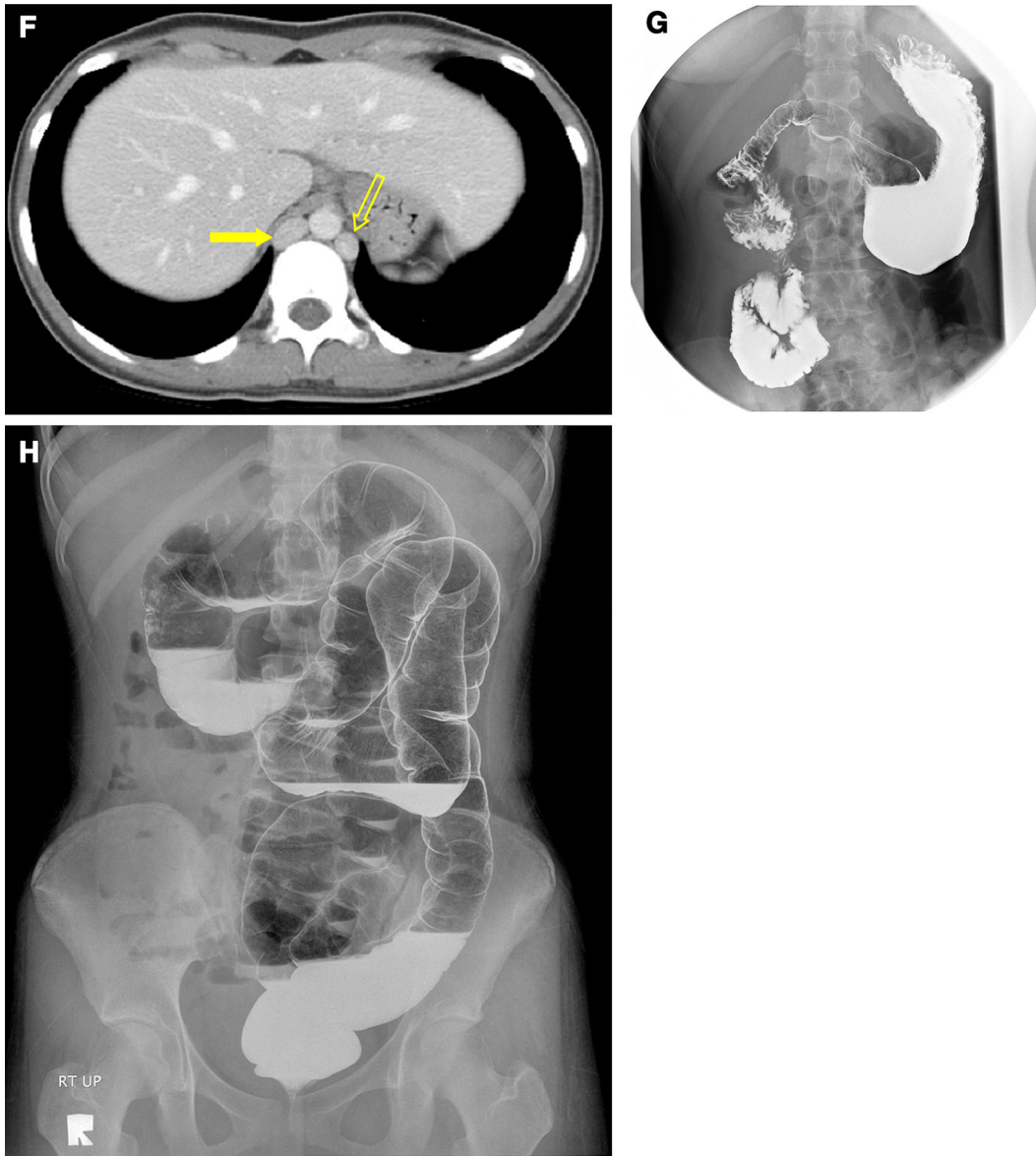


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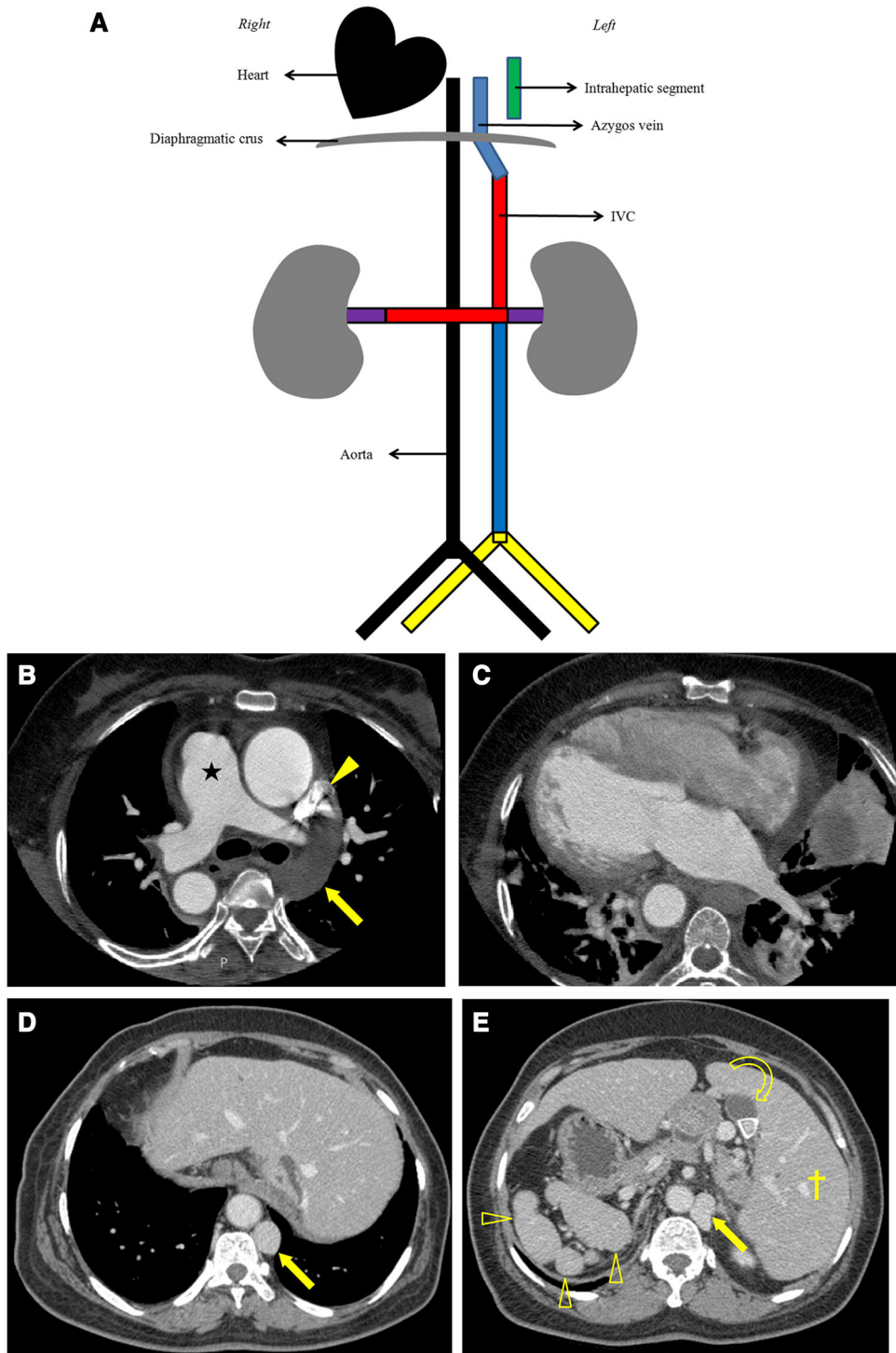


Fig. 12. IVC interruption with azygos continuation in a 53-year-old woman with situs inversus totalis. **A** Schematic shows the absence of anastomosis between the intrahepatic and suprarenal IVC. The heart and aorta are on the right side, and the IVC and anatomical azygos vein are positioned on the left side. **B–F** Axial CT scans from cranial to caudal show the anomaly. **B, C** The pulmonary trunk (*closed star*) is positioned to the right of the ascending aorta and the base-to-apex axis of the heart points to the right, indicating dextrocardia. Note the azygos vein (*arrow*) joining the anatomic right atrium

(*arrowhead*). **D** There is an enlarged azygos vein (*arrow*), and the IVC is interrupted. **E** The liver (*+*) and gallbladder (*open curved arrow*) are on the left side of the abdomen, in contrast to the polysplenia (*open arrowheads*), which is seen on the right side of the abdomen. There is an anastomosis between the IVC and the azygos vein (*arrow*). **F** The IVC receives the anatomic left renal vein (*curved arrow*). **G** Coronal reformatting image shows that the IVC (*open arrow*) continues to the anatomical azygos vein (*arrow*), which is positioned to the left of the aorta.

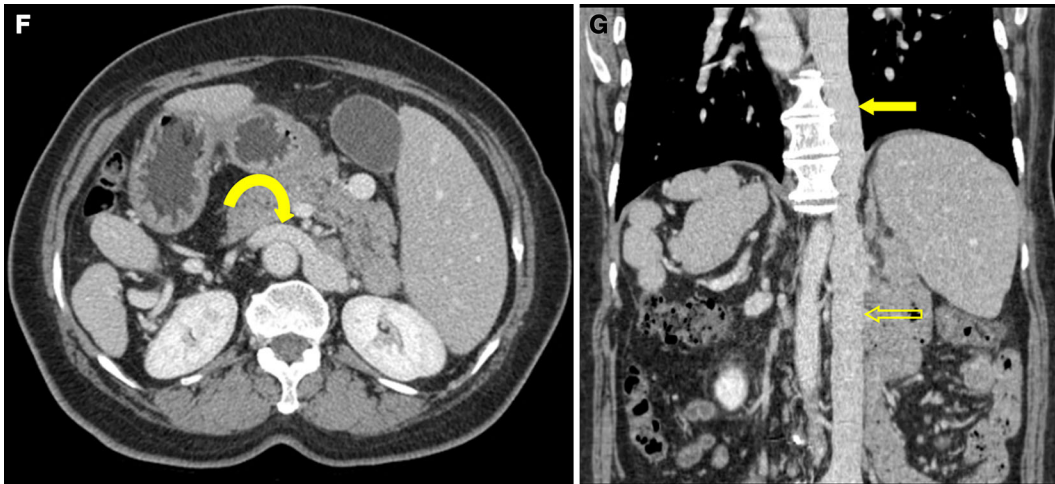


Fig. 12. continued.

artery lies between the two common iliac veins [25, 28]. There might or might not be an anastomosis between the iliac veins, and the right ureter can go between the right double IVCs and be compressed by them (Fig. 10) [28].

Double IVC interruption with azygos and hemiazygos continuation in heterotaxy

The interruption of a double IVC with azygos and hemiazygos continuation arises from agenesis of both subcardinal veins, accompanied by the persistence of both supracardinal veins (Fig. 11A) [12, 29]. As mentioned earlier, interruption of the IVC is associated with heterotaxy syndrome [30]. We found a double IVC interruption with azygos and hemiazygos continuation in a patient with situs ambiguous presenting with a midline liver, polysplenia, and a short pancreas (Fig. 11). The right-sided IVC continued up to the azygos vein, and the left-sided IVC was connected to the hemiazygos vein. Renal venous flow drained to both ipsilateral IVCs without a midline-crossing renal vein, thus forming an independent trunk for each IVC below the diaphragm.

IVC interruption with azygos continuation in a patient with situs inversus totalis

Situs inversus totalis refers to the total transposition of the thoracic and abdominal viscera, a mirror image of the normal configuration, which is termed situs solitus. The prevalence is 0.01%. The bilobed lung, cardiac apex, stomach, and spleen are right-sided structures, and the trilobed lung, liver, gallbladder, and IVC are left-sided structures in patients with situs inversus totalis [31–33]. In this situation, a left-located retrocaval vein functions

as an anatomical azygos vein and a right-positioned vein functions as an anatomical hemiazygos vein. IVC interruption can exist in patients with situs inversus totalis, and the failure to form a subcardinal–intrahepatic anastomosis is similar to what is seen in IVC interruption with azygos continuation in a situs solitus patient (Fig. 12).

Left IVC interruption with hemiazygos continuation, retroaortic right renal vein, and the presence of a right suprarenal IVC

We discovered a patient with left IVC interruption with hemiazygos continuation, a retroaortic right renal vein, and a right suprarenal IVC. In this subject, both common iliac veins formed the left-sided IVC, which continued up to the hemiazygos vein. Subsequently, the hemiazygos vein crossed the midline mediastinum posterior to the right atrium and finally joined the superior vena cava. In addition, there was a small right suprarenal IVC in the normal position with one retroaortic right renal vein. The larger left IVC and smaller suprarenal right IVC communicated via a retroaortic right renal vein without a midline-crossing left renal vein (Fig. 13). Usually, the retroaortic right renal vein is accompanied by a double IVC interruption with hemiazygos continuation, and in that situation, there are bilateral infrarenal IVCs and a left suprarenal IVC [9, 30] (Fig. 14). Our subject, however, showed a left infrarenal IVC and bilateral suprarenal IVCs with left IVC interruption with hemiazygos continuation. To the best of our knowledge, this particular IVC anomaly has not been reported prior to this article.

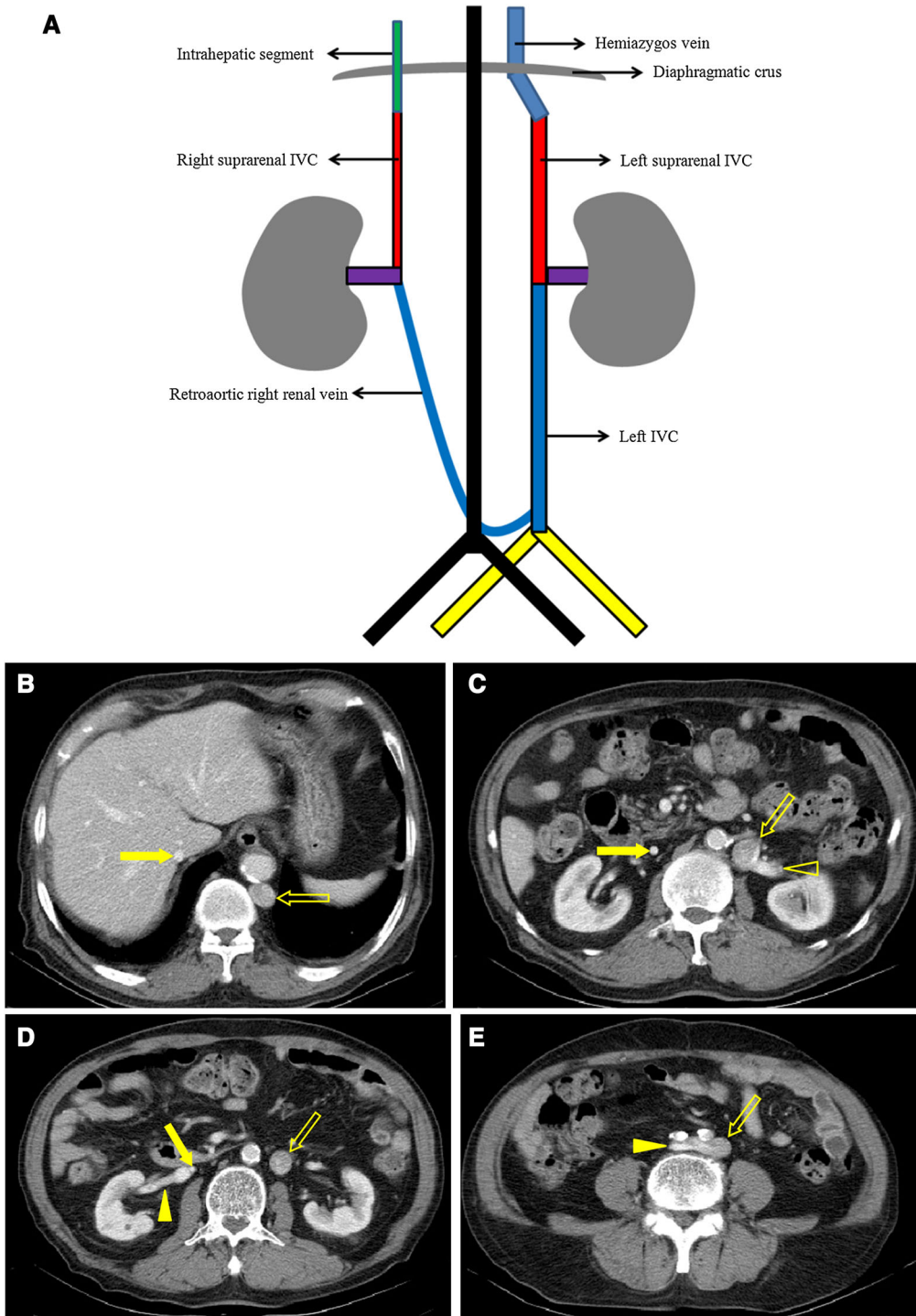


Fig. 13. Left IVC interruption with hemiazygos continuation, retroaortic right renal vein, and the presence of a right suprarenal IVC in a 77-year-old man. **A** Schematic shows two suprarenal IVCs and a left infrarenal IVC. The right suprarenal IVC continues up to the intrahepatic segment of the IVC, and the left suprarenal IVC continues to the hemiazygos vein. The right suprarenal IVC connects to the left infrarenal IVC via a midline-crossing retroaortic right renal vein. **B–E** Axial CT scans from cranial to caudal show the anomaly. **B** There is a hypoplastic

intrahepatic IVC (arrow) and an enlarged hemiazygos vein (open arrow). **C, D** The left IVC (open arrow) receives the left renal vein (open arrowhead). Note the hypoplastic right suprarenal IVC (arrow) connected to the right renal vein (arrowhead). **E** The right renal vein (arrowhead) crosses posterior to the aortic bifurcation to join the left IVC (open arrow). **F, G** Curved reformatted images show a retroaortic right renal vein (arrowhead) communicating between the left IVC (open arrow) and the right suprarenal IVC (arrow).

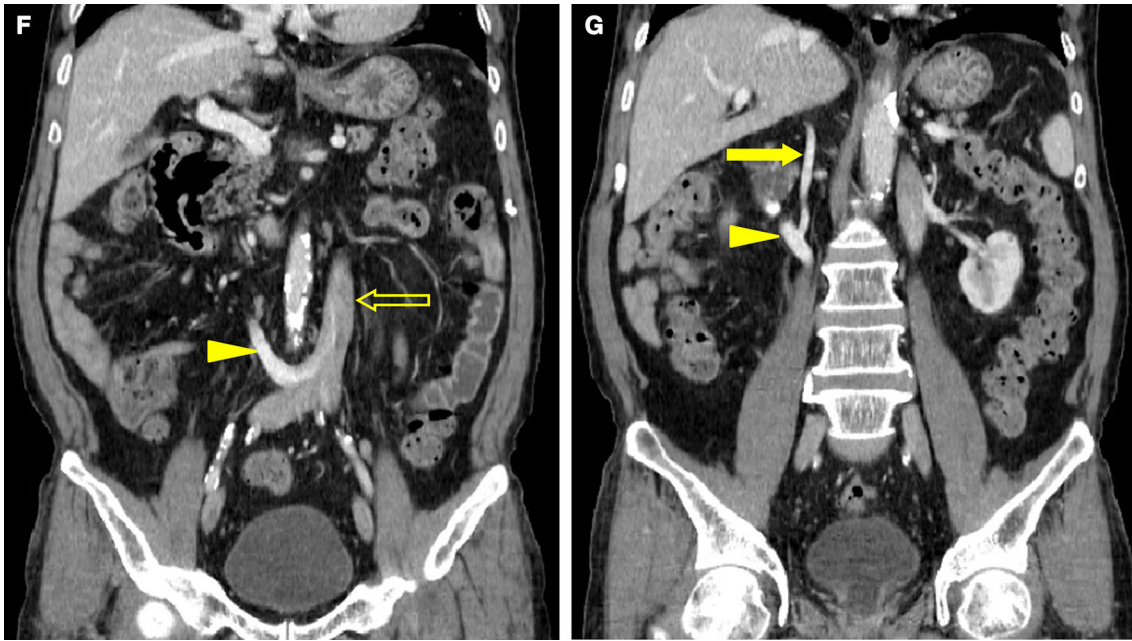


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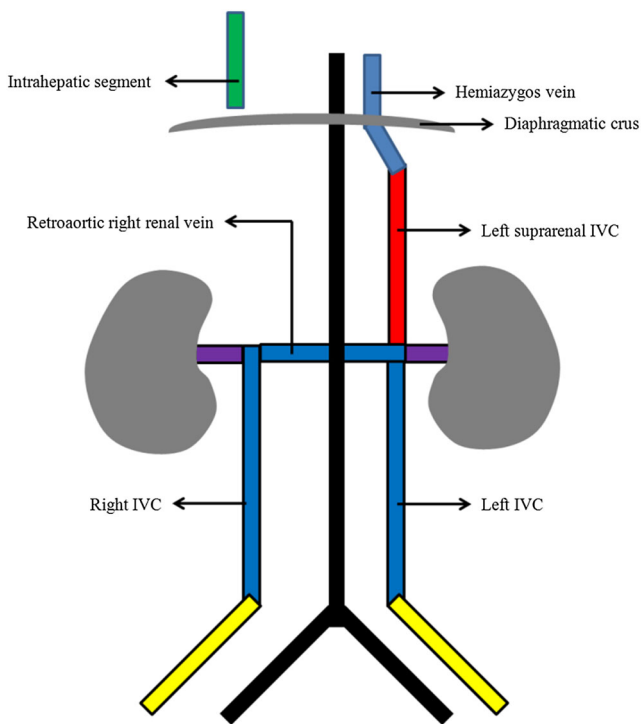


Fig. 14. Schematic of a double IVC interruption with hemiazygos continuation and a retroaortic right renal vein.

Conclusion

Congenital anomalies of the IVC are diverse and relatively common, resulting from abnormal development or regression during the complex process of embryogenesis.

Although most patients with IVC anomalies are asymptomatic, the anomalies can be significant in daily practice. Familiarity with the variety of IVC anomalies seen on imaging is vital for correctly diagnosing and managing patients in daily practice.

Compliance with ethical standards

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Conflict of interest The authors declare that they have no conflicts of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors. For this type of study, formal consent was not required.

Informed consent Statements of informed consent were not applicable because the manuscript contains no patient data.

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