The curious case of the disappearing IVC: A case report and review of the aetiology of Inferior Vena Cava Agenesis

Michael Paddock[1*], Nicola Robson[2]

1. Academic Unit of Radiology, University of Sheffield, Royal Hallamshire Hospital, Sheffield, United Kingdom

2. Department of Radiology, Poole Hospital NHS Foundation Trust, Poole, Dorset, United Kingdom

* **Correspondence:** Dr Michael Paddock, Academic Unit of Radiology, University of Sheffield, Floor C, Royal Hallamshire Hospital, Glossop Road, Sheffield, S10 2JF, United Kingdom
(*michael.paddock@doctors.org.uk*)

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ABSTRACT

We report the case of a previously well 18-year-old male who presented to the Emergency Department with lower limb pain. An ultrasound demonstrated extensive left sided deep vein thrombosis and computed tomography demonstrated inferior vena cava agenesis, leading to the diagnosis of inferior vena cava agenesis associated deep vein thrombosis. The aetiology of inferior vena cava agenesis is explored in depth.

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CASE REPORT

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An 18-year-old man presented to the Emergency Department (ED) complaining of a swollen, painful left knee after sustaining a 'twisting' injury of the left lower limb. The symptoms worsened on repeated activity.

Past history is relevant for mild learning difficulties and operations for a 'blocked bowel' when he was younger, for which the patient and his mother were unable to elaborate. However, later review of the medical notes revealed an episode of abdominal distension, grunting and acidosis on day 4 of life after a full term birth. An unsuccessful anal dilation and bowel decompression preceded the formation of a descending loop colostomy. This was reversed at 16 months but an anastomotic leak gave rise to peritonitis necessitating a further defunctioning colostomy, eventually closed at 4 years of age. Additionally, he had a pyloromyotomy for pyloric stenosis at 2 months of age.

There was no history of risk factors for venous thromboembolism (VTE) or deep vein thrombosis (DVT) and no family history of VTE or clotting diatheses. He denied smoking, taking alcohol or illegal drugs, and was otherwise well.

Initial examination revealed normal vital signs and significant swelling to the left calf and the posterior aspect of the left thigh with no associated superficial venous swelling, effusion or instability of the left knee. Peripheral pulses were palpable but left foot capillary refill time was prolonged at 3 to 4 seconds and the limb was cool to the touch. Neurological examination was normal. Abdominal examination revealed numerous well-healed surgical scars in a soft, non-tender abdomen with normal bowel sounds and no inguinal lymphadenopathy.

Blood tests revealed a neutrophilic leucocytosis and a significantly elevated d-dimer level (1850 ng/ml, NR 0 - 500 ng/ml). Platelet count, international normalised ratio, prothrombin time, activated partial thromboplastin time, urea, electrolytes, liver function tests and bone profile were all within the normal range.

An urgent ultrasound (US) of the left leg was performed where compression venography revealed evidence of extensive DVT. Incompletely occluding thrombus was seen in the left external iliac, common femoral vein and superficial femoral vein with some venous flow in the popliteal vein. The left common iliac vein was poorly visualised but contained

thrombus (Fig. 1). The posterior tibial and peroneal veins appeared clear. The inferior vena cava (IVC), although not well seen, had detectable venous flow in its distal portion. A chest radiograph (Fig. 2) demonstrated a normal heart size and no lung lesions with the caveat that a pulmonary embolus (PE) could not be excluded on the basis of these findings.

He was managed with elevation of the limb and anticoagulation with a low molecular weight heparin. He was not DVT thrombolysed given that distal neurovascular status was intact.

A CT (computed tomography) scan was organised and demonstrated that the intra-abdominal IVC was smaller (Fig. 3) than usual, and numerous venous collaterals in the abdomen and pelvis were visualised. The two common iliac veins contained thrombus and formed a small right-sided IVC. The right-sided iliac vein crossed the midline dividing and draining deep structures with collaterals noted between the psoas and iliacus muscle. A large collateral vein with thrombus extended from this iliac vessel in the pelvis, to drain the right kidney (Fig. 4), connecting into the IVC that directly drained the left kidney. On the left, a prominent gonadal vein was seen to connect to a lumbar vein forming a focal bulbous region with a vein passing from here, anterior to the aorta, to the suprarenal portion of the IVC, also draining the left kidney (Fig. 5). Collaterals with thrombus were demonstrated draining into this bulbous left sided vein in a similar manner to that which a congenital left sided IVC would, however their multiple nature and more lateral lie was noted (Fig. 6). Additionally, they arose from a small vein which appeared to connect directly to the left external iliac system and were therefore considered less likely to represent a double IVC anomaly, as opposed to the opening up of venous channels due to previous thrombosis: either is possible. Due to poor opacification it was difficult to follow the extent of clot, but there was suspicion of IVC thrombus in its infrarenal portion. Of note, a bony sacral defect was also demonstrated (Fig. 7).

The haematology team recommended a thrombophilia screen and immediate commencement of vitamin K antagonist therapy, initially for 6 months, with further review as an outpatient. The patient was made aware that he may have to continue lifelong anticoagulation and was discharged home. The outpatient haematology review revealed no evidence of inherited thrombophilia; a weakly positive lupus anticoagulant result in isolation was not thought to be of clinical significance. The patient received 6 months of oral anticoagulation therapy in total and was subsequently discharged from follow up.

DISCUSSION

Aetiology

Anomalies of the IVC are estimated in 0.5% of the general population [1]. DVT is associated with high morbidity and mortality and has an estimated prevalence rate of 1 per 1000 [2]. A lower prevalence rate exists in younger patient groups with an incidence as low as 1 per 10 000 [3]. The aetiology of DVT is associated with congenital risk factors (thrombophilias, autoimmune conditions) and acquired risk factors (oral contraceptive use, prolonged immobilisation, neoplastic disease): one or more of these risk factors may be found in up to 80% of patients with confirmed DVT [4]. IVC anomalies may be found in up to 5% of this younger patient population with confirmed DVT [5-7], which is greater than the expected rate: up to 5% observed versus 0.5% expected [1].

There is no firm consensus of the etiology of IVC agenesis (IVCA) with the literature presenting two schools of thought: congenital absence [3, 4] or early IVC thrombosis in the perinatal period [8, 9]. Congenital IVCA suggests defective embryonic development of all 3 renal IVC segments (infra-, renal and supra-) at week six to eight of gestation [4, 10], resulting in progressive segmental hypoplasia of the renal IVC [11]. Recent literature has demonstrated a link between confirmed perinatal IVC thrombosis and infrarenal IVC absence suggesting that thrombosis of the IVC leads to subsequent hypoplasia and 'absence' of the IVC [8, 9].

The CT images of our patient confirmed a rudimentary IVC, with no definable infrarenal IVC. The bulbous vein described is rather like a left IVC and has the usual connections to the right IVC. One could postulate that the left para-aortic collateral, which is single at the bifurcation, could be a small left IVC which thrombosed at some stage and may represent a possible hypoplastic remnant that may have re-cannulated, or, that it may represent a true congenital double IVC with a small left sided vessel, possibly associated with partial sacral agenesis (Fig. 7). There is one case report that describes sacral agenesis with an anomalous inferior and superior vena cava in conjunction with Goldenhar (oculoauriculovertebral) syndrome [12].

IVC anomalies may present with non-specific, vague symptoms, such as lower back [5, 10] or abdominal pain [3, 5], or may be typically asymptomatic [4]. The expected rate described above may also be an underestimation given the asymptomology as it may only be discovered as an incidental finding on imaging or at surgery. IVC abnormalities have been associated with other congenital abnormalities, mainly in those organs (spleen, liver, heart and lung) whose embryological development occurs at the same time of the IVC [4, 13]. Additionally, patients without associated congenital anomalies are typically asymptomatic [4]. A Brazilian case report has highlighted a potential association between partial IVCA and congenital abnormalities of the gut [14]. Our patient presented with features consistent with bowel obstruction in the neonatal period, the cause of which was speculated to include a congenital aetiology. In the absence of other risk factors, extensive intra-abdominal surgery in our patient's infancy may have been a risk factor for IVCA, subsequently leading to inferior vena cava agenesis associated deep vein thrombosis (IVCA-aDVT). It currently remains unknown whether catheter related procedures in the neonatal period carry a similar risk of thrombosis and further review of the medical notes did not allude to these procedures being performed.

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IVCA-aDVT has been described as a separate clinical entity in the literature [1, 5, 6, 15]. A retrospective analysis indicated that the incidence of IVCA-aDVT was greater in males: in a review of 72 documented cases, nearly 82% of IVCA-aDVT patients were male [5]. Affected patients are typically in their second to fourth decade of life [16] and a prospective study stated that occurrence of DVT affected those aged less than 30 years [11]. Iqbal et al, hypothesises that inadequate flow in collateral vessels results in chronic venous hypertension and venous stasis, promoting the formation of further venous collaterals which may 'precipitate thrombosis' [3]. Lambert et al, proposes that the inability of the venous collateral system to cope with the demands of increasing blood flow, results in venous stasis and a subsequent propensity for clotting, resulting in the formation of DVT [5]. It has been demonstrated in the literature that IVCA is an independent risk factor for DVT by this proposed mechanism [6, 8, 17] resulting from subsequent changes in 'venous flow velocity' [1]. Our patient may have become symptomatic when the collateral venous system was unable to maintain venous homeostasis, possibly secondary to blockage from new clot formation from resultant vessel endothelial instability, or changes in venous flow velocity following the twisting injury at the knee.

Concurrent PE is not a frequent finding with IVCA-aDVT given that a true IVC does not exist. Any emboli would have to travel through the collateral networks or the azygos/hemiazygos system to reach the pulmonary circulation [5], explaining the low frequency of lung involvement. However, one case report has highlighted this mechanism as a rare cause for PE [4]. Moreover, a prophylactic IVC filter would not be recommended given the low instance of PE in these patients.

Imaging findings

US, CT and Magnetic Resonance Imaging (MRI) have supplanted direct contrast venography of the IVC for diagnosis. US is a relatively good and cheap first line method for assessment of the IVC, however it is operator dependent and is limited by body habitus and bowel gas. In this case, visualisation was poor and the small infrarenal IVC could not be demonstrated on US: CT or MRI better assess anomalies of the IVC for this reason. CT confers the added advantage of demonstrating additional pathology such as pelvic masses and collateral circulation. However, CT imaging does 'not contribute to diagnosing IVCA if it is not specifically sought' [5]. Multiplanar reconstruction allows for excellent imaging on the whole but the poor opacification in this patient limited this technique. In retrospect, a pedal vein injection of contrast may have improved visualisation. MRI is replacing CT as the choice of imaging modality and is thought to be more accurate in detecting thrombosis and IVC anomalies [18].

Imaging the IVC may not always be able to delineate the true of aetiology of IVCA in patients. However, imaging should be interpreted in the context of the history and clinical presentation, which may give credence to one of the proposed aetiologies presented above. This may have ramifications for long-term patient management, for example, in a young child as part of a clinical syndrome or other congenital abnormalities, or an adult presenting with recurrent DVT.

Treatment and prognosis

Prolonged vitamin K antagonist therapy appears to be the mainstay of management with outpatient haematology input to monitor progress and manage the risks of long-term anticoagulation therapy. Thrombophilia and/or autoimmune screening should be performed to exclude an underlying clinical condition or syndrome with prothrombotic tendencies. Elastic stocking and leg elevation are useful anti-VTE measures in addition to limiting further acquired thrombotic risk factors. Treatment with low molecular weight heparin may be used as a safe alternative in patients with contraindications for long-term oral anticoagulation therapy [7].

Catheter directed thrombolytic therapy, although sometimes used in young patients, is problematic in those with abnormal anatomy. Surgical management is rarely indicated but should be considered if there is coexisting vascular impairment and depending on the extent of the DVT. Zhou et al, describe successful polytetrafluoroethylene graft placement in a symptomatic patient with infrarenal IVC absence and collateral formation resulting in pelvic congestion and lower extremity swelling. The graft connected the common femoral vein to the suprarenal IVC [19]. However, there is currently no evidence based best practice guidance regarding the long-term management of IVCA-aDVT, rather, current accepted DVT management as applied to IVCA-aDVT.

Patients with IVCA are at risk of long-standing asymptomatic clot formation and accumulation with a resultant high risk of DVT and recurrence. Early identification of these patients may allow for earlier intervention such that later risk in adulthood is reduced, however, there currently exists no data regarding long-term morbidity and mortality [9].

Differential diagnoses

The differential diagnosis as discussed above would lie mainly between a congenital absence of the infrarenal IVC or IVC thrombosis in the perinatal period. It is possible that a small left IVC was thrombosed and became recannulated or that the patient may have had a double IVC with a small left sided vessel. The IVC embryogenesis involves anastomoses between three paired embryonic veins. This is a complex process and anomalies are numerous. Thrombosis in the perinatal period could lead to persistence of channels that would otherwise have regressed. Various anomalies and illustrations of these are described in Tables 2 and 3.

TEACHING POINT

A young male presenting with a more proximal deep vein thrombosis in the absence of acquired venous thromboembolism risk factors should raise the suspicion of inferior vena cava agenesis and warrant further investigation.

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FIGURES

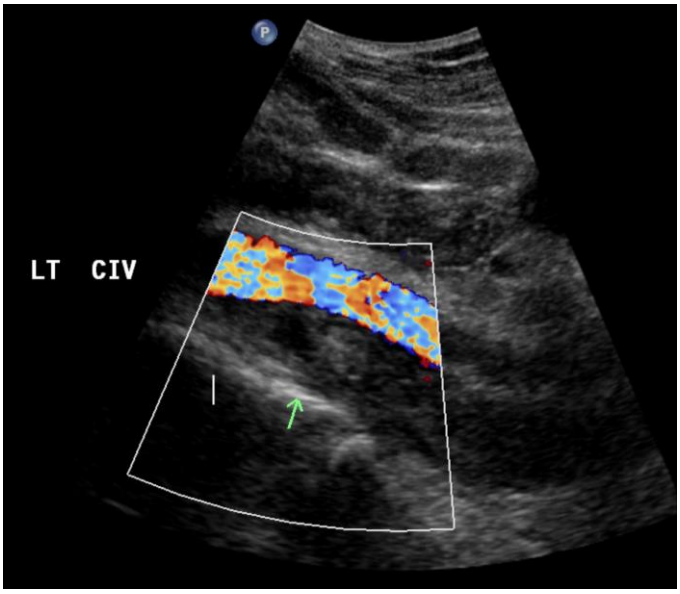

Figure 1: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Sagittal ultrasound image shows thrombus in the left common iliac vein (green arrow) with good blood flow in the artery overlying. TECHNIQUE: Ultrasonography was performed using a Philips IU22 (Philips Healthcare UK) ultrasound machine with linear array and curvilinear 5 - 2 megahertz (MHz) probes.

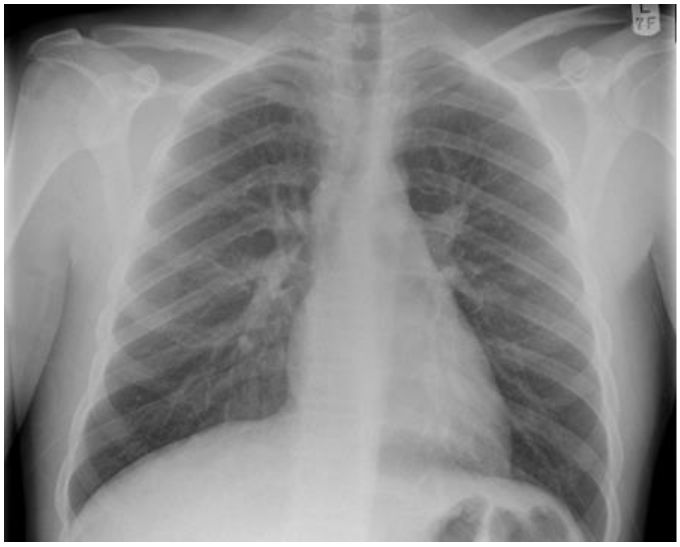

Figure 2: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Normal posterior-anterior chest radiograph appearances.

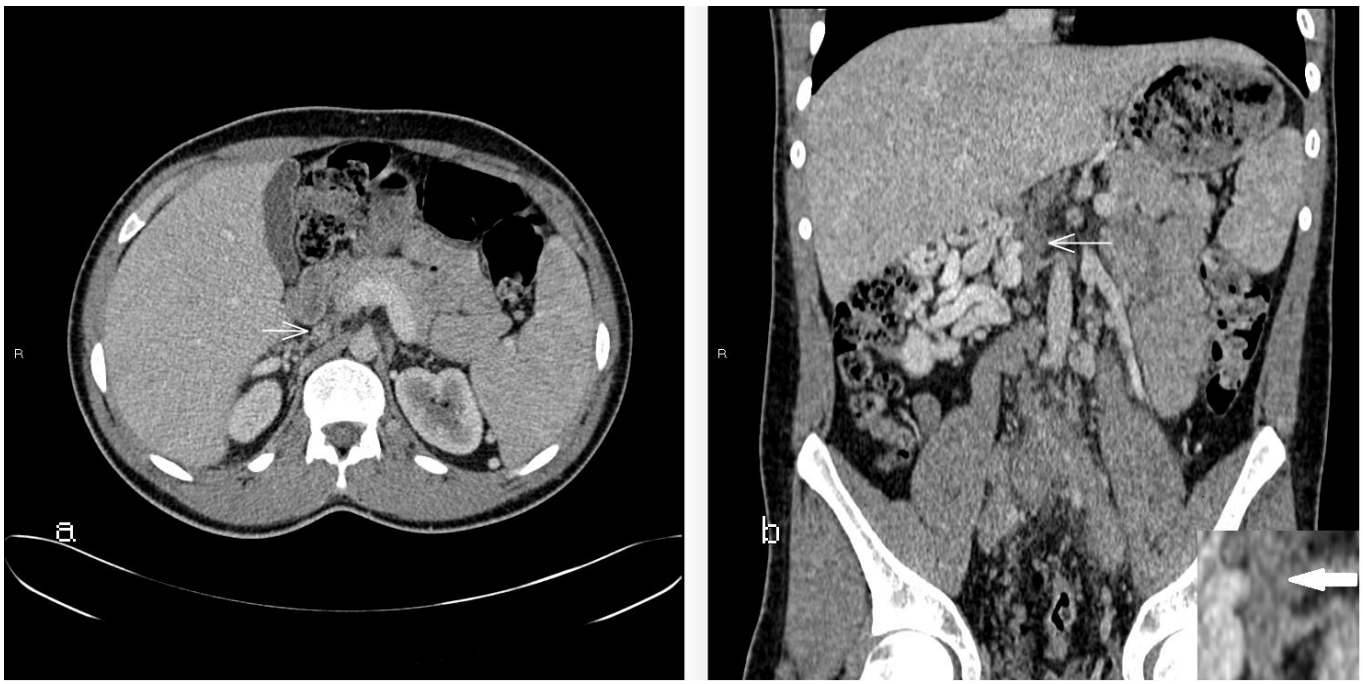

Figure 3: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Venous phase axial (a) and coronal (b) enhanced computed tomography of the abdomen demonstrating the small right sided suprarenal IVC cava (white arrows; solid white arrow in magnification (b)). TECHNIQUE: The patient was imaged on a Siemens Sensation 16 CT Scanner (Siemens Medical, Forchheim Germany) scanned helically with axial reconstruction at 1.5mms, matrix size 512x512, with 120kVp and modulated mAs varying from 70 to 205. An initial scan was acquired after administration of non ionic Iopamidol (Niopam 300mg Iodine/ml Bracco UK Limited) IV Contrast triggered from the aorta with 25 second delayed imaging performed from pelvis to lung apices with a second study of the pelvis visually triggered over the IVC as initial opacification of pelvic veins was poor. A pump injection was used with a speed of 4 mls/sec. The legs were not imaged as ultrasound had provided diagnostic information regarding the presence of thrombus on the left side.

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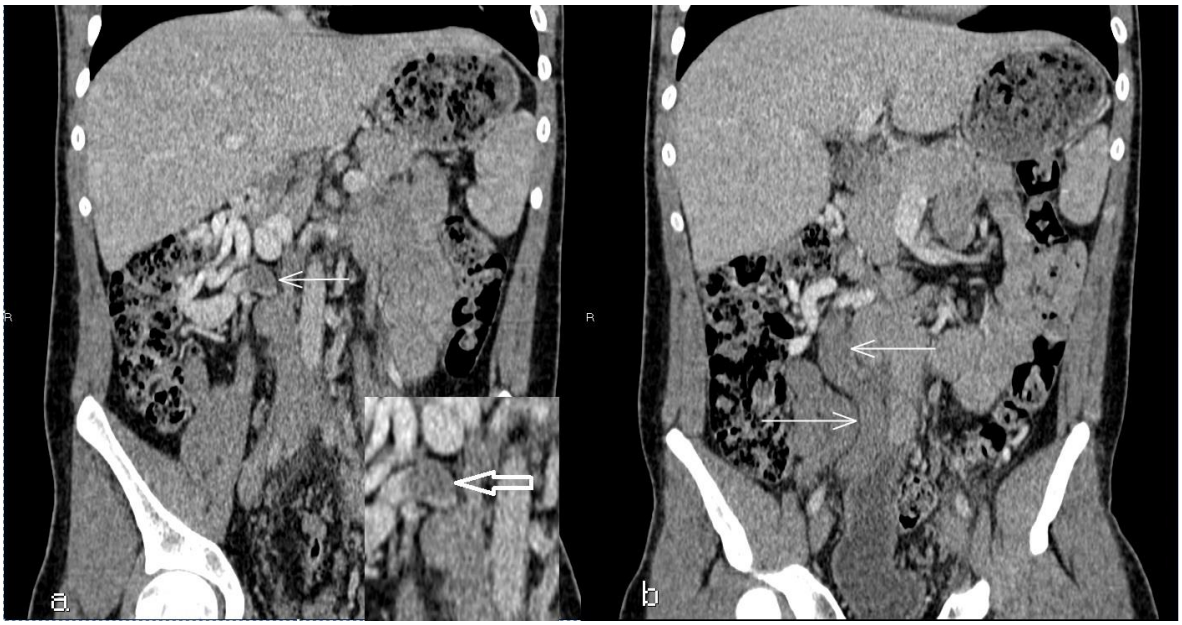

Figure 4: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Coronal views of the abdomen at different levels (a posterior to b) demonstrating a large collateral vessel (white arrows; hollow white arrow in magnification (a)) containing thrombus, draining the right kidney. TECHNIQUE: The patient was imaged on a Siemens Sensation 16 CT Scanner (Siemens Medical, Forchheim Germany) scanned helically with axial reconstruction at 1.5mms, matrix size 512x512, with 120kVp and modulated mAs varying from 70 to 205. An initial scan was acquired after administration of non ionic Iopamidol (Niopam 300mg Iodine/ml Bracco UK Limited) IV Contrast triggered from the aorta with 25 second delayed imaging performed from pelvis to lung apices with a second study of the pelvis visually triggered over the IVC as initial opacification of pelvic veins was poor. A pump injection was used with a speed of 4 mls/sec. The legs were not imaged as ultrasound had provided diagnostic information regarding the presence of thrombus on the left side.

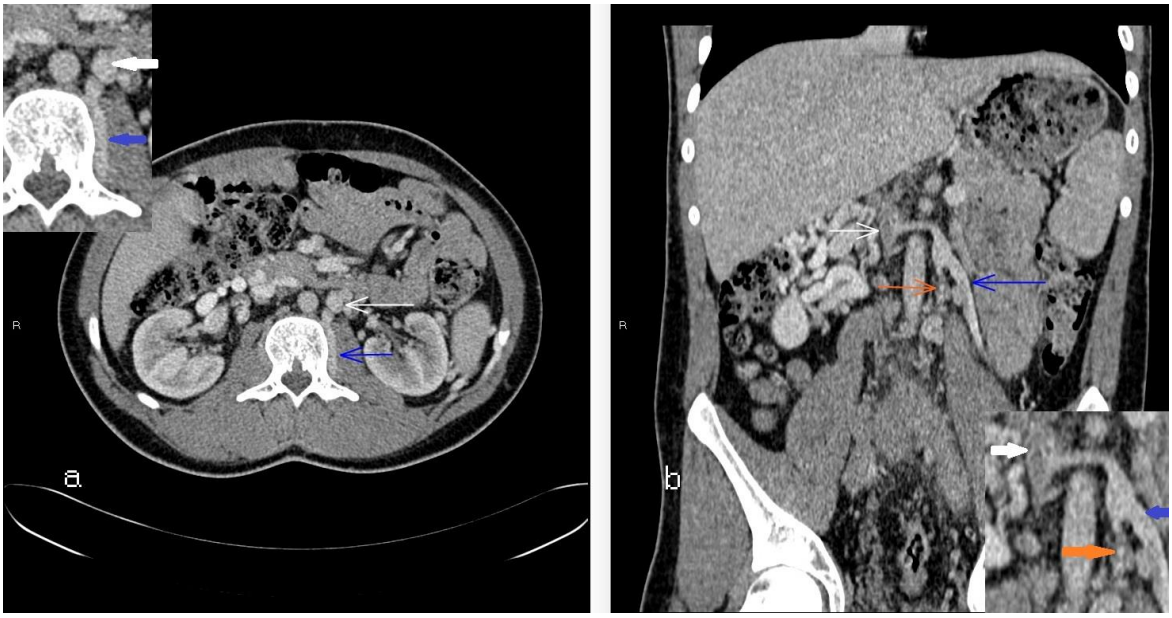

Figure 5: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Computed tomography axial scan (a) of the abdomen showing a lumbar vein (blue arrow in main image (a); solid blue arrow in magnification (a)) forming bulbous left 'inferior vena cava' (white arrow in main image; solid white arrow in magnification (a)). Coronal image (b) demonstrating a left gonadal vein (blue arrow in main image (b); solid blue arrow in magnification (b)) with left sided collaterals (orange arrow in main image (b); solid orange arrow in magnification (b)) draining into the bulbous left 'inferior vena cava' which crosses the aorta to join the suprarenal portion of the right sided inferior vena cava (white arrow in main image (b); solid white arrow in magnification (b)). TECHNIQUE: The patient was imaged on a Siemens Sensation 16 CT Scanner (Siemens Medical, Forchheim Germany) scanned helically with axial reconstruction at 1.5mms, matrix size 512x512, with 120kVp and modulated mAs varying from 70 to 205. An initial scan was acquired after administration of non ionic Iopamidol (Niopam 300mg Iodine/ml Bracco UK Limited) IV Contrast triggered from the aorta with 25 second delayed imaging performed from pelvis to lung apices with a second study of the pelvis visually triggered over the IVC as initial opacification of pelvic veins was poor. A pump injection was used with a speed of 4 mls/sec. The legs were not imaged as ultrasound had provided diagnostic information regarding the presence of thrombus on the left side.

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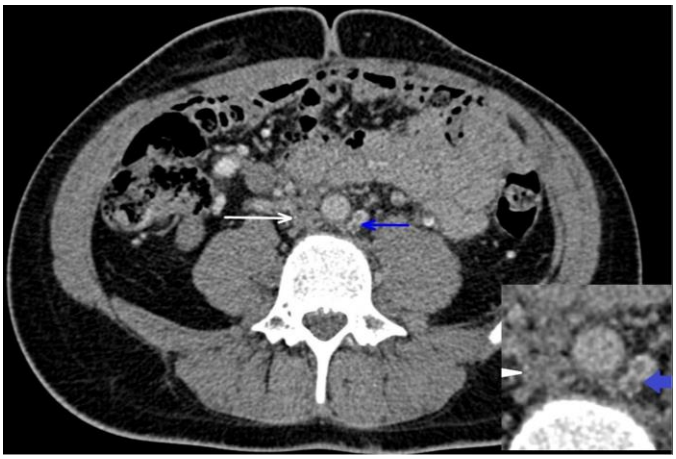

Figure 6: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: Computed tomography axial scan of the abdomen demonstrating the right sided inferior vena cava (white arrow; solid white arrow in magnification) and left sided collaterals containing thrombus (blue arrow; blue solid arrow in magnification). TECHNIQUE: The patient was imaged on a Siemens Sensation 16 CT Scanner (Siemens Medical, Forchheim Germany) scanned helically with axial reconstruction at 1.5mms, matrix size 512x512, with 120kVp and modulated mAs varying from 70 to 205. An initial scan was acquired after administration of non ionic Iopamidol (Niopam 300mg Iodine/ml Bracco UK Limited) IV Contrast triggered from the aorta with 25 second delayed imaging performed from pelvis to lung apices with a second study of the pelvis visually triggered over the IVC as initial opacification of pelvic veins was poor. A pump injection was used with a speed of 4 mls/sec. The legs were not imaged as ultrasound had provided diagnostic information regarding the presence of thrombus on the left side.

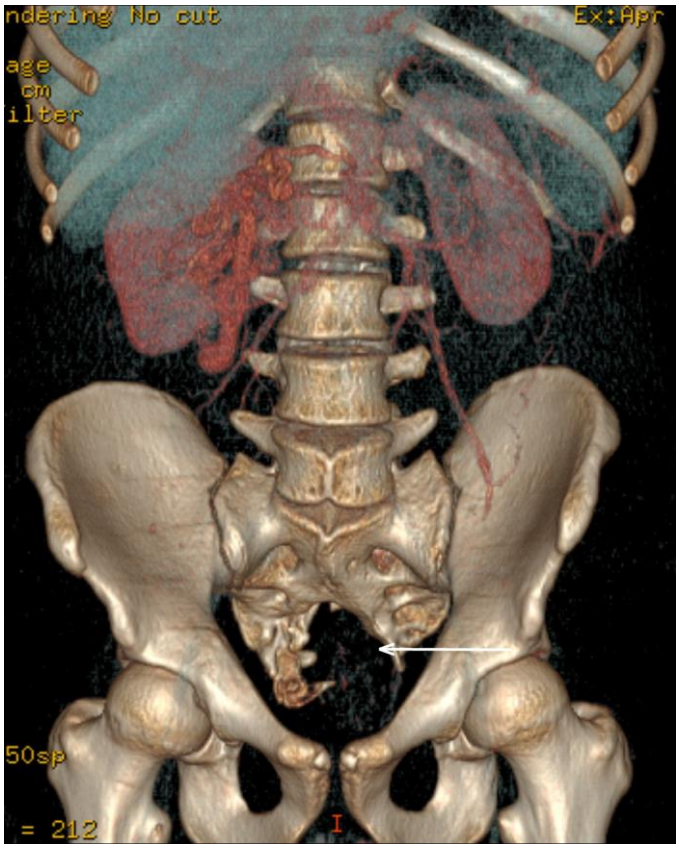

Figure 7: 18-year-old male with extensive DVT and infrarenal IVC agenesis. FINDINGS: A three-dimensional volumetric reconstruction demonstrating the left sided sacral bony defect (white arrow). TECHNIQUE: The patient was imaged on a Siemens Sensation 16 CT Scanner (Siemens Medical, Forchheim Germany) scanned helically with axial reconstruction at 1.5mms, matrix size 512x512, with 120kVp and modulated mAs varying from 70 to 205. An initial scan was acquired after administration of non ionic Iopamidol (Niopam 300mg Iodine/ml Bracco UK Limited) IV Contrast triggered from the aorta with 25 second delayed imaging performed from pelvis to lung apices with a second study of the pelvis visually triggered over the IVC as initial opacification of pelvic veins was poor. A pump injection was used with a speed of 4 mls/sec. The legs were not imaged as ultrasound had provided diagnostic information regarding the presence of thrombus on the left side.

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Aetiology	Inferior vena cava agenesis associated deep vein thrombosis; may present as part of a clinical syndrome with congenital abnormalities
Incidence	Inferior vena cava anomalies may be found in up to 5% of younger patients with confirmed DVT (5% observed vs. 0.5% expected)
Gender ratio	Male:female = 4:1
Age predilection	Second to fourth decade of life
Risk factors	Inferior vena cava agenesis, venous collateral formation
Treatment	Accepted DVT management; exclude thrombophilia
Prognosis	Good, as responds to management; risk of recurrence in prothrombotic state
Findings on imaging	Hypoplastic remnants or complete absence of the infrarenal IVC; venous collateral formation; venous thrombus. Operator dependant views of IVC on US; CT or MRI better assess IVC anomalies and extent of venous thrombus

Table 1: Summary table for inferior vena cava agenesis associated deep vein thrombosis.

Diagnosis	Cause of anomaly & incidence in population	Differential diagnosis	CT & MRI findings
Left IVC	Persistence of the left supracardinal vein and regression of the right supracardinal vein. 0.2–0.5%	Left para-aortic lymphadenopathy.	Contrast enhanced CT or MRI demonstrates an isolated left IVC that joins the left renal vein and crosses anterior to the aorta to join the normal right IVC.
Double IVC	Persistence of both supracardinal veins. 1–3%	Left para-aortic lymphadenopathy.	Imaging as for the left IVC but occasionally the left IVC crosses at a level below the renal vein: there can be a difference in size between the 2 IVCs. Associated anomalies seen on imaging include right double IVC with retro-aortic right renal vein and double IVC with hemiazygos continuation on the left.
Absence of infrarenal IVC	Intrauterine or perinatal thrombosis of the IVC. Very rare.	Enlarged collateral vessels simulate a paraspinal mass.	Contrast enhanced CT/MRI demonstrate absence of the infrarenal IVC with prominent ascending lumbar veins that drain into the azygos-hemiazygos system.
Retro aortic and circumaortic left renal vein	Persistence of the posterior supracardinal-supracardinal anastomosis with ventral arch regression. 1.7–8.7%	Left para-aortic lymphadenopathy.	Contrast enhanced CT/MRI demonstrate a single left renal vein which passes behind the aorta to join the right sided IVC or circumaortic venous ring with one vein coursing anterior and the other posterior to the aorta.
Azygos or hemiazygos continuation of IVC	Failure of the right subcardinal-hepatic vein anastomosis to form with atrophy of right subcardinal vein. 0.6%		Contrast enhanced CT/MRI demonstrates azygos vein continuation of the infrarenal IVC, and, if left IVC, as hemiazygos vein. The renal segment receives blood from both kidneys and passes posterior to the crura to enter the thorax as the azygos vein that joins the SVC in the normal place. The hepatic segment drains directly into the right atrium.
Retrocaval ureter	Infrarenal IVC develops from the right posterior cardinal vein that lies anterior and lateral to the ureter.		Contrast enhanced CT/MRI demonstrates part of the right ureter trapped posterior and medial to the IVC. Significant compression could result in hydronephrosis.

Table 2: Summary table for variants and anomalies of the IVC with corresponding imaging appearances on CT/MRI.

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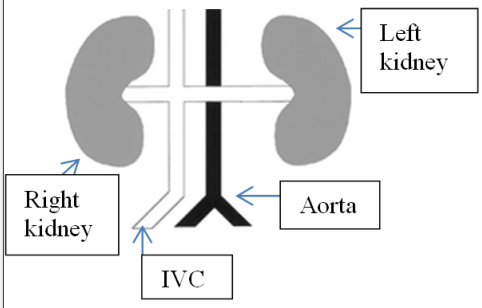
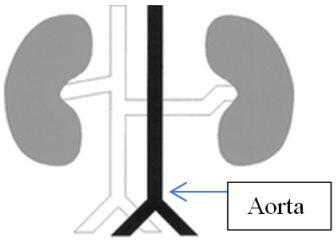
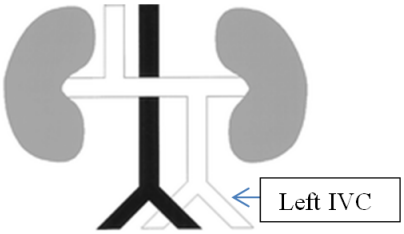
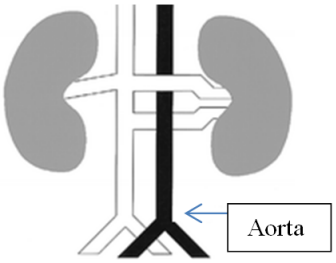
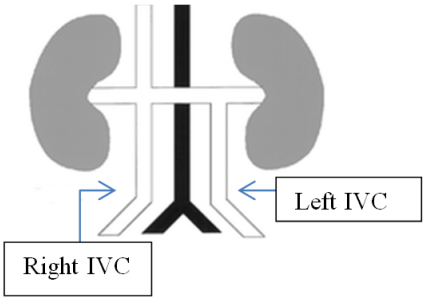
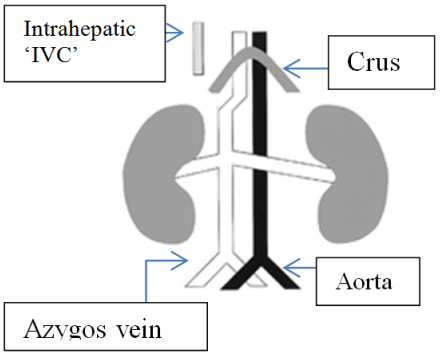
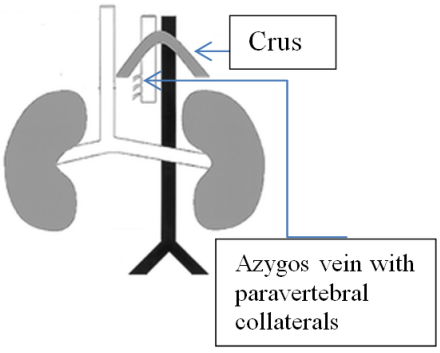
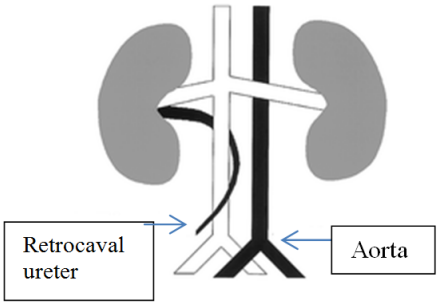
Diagnosis		Diagnosis	
Normal		**Retroaortic left renal vein**	
Left IVC		**Circumaortic left renal vein**	
Double IVC		**Azygos continuation of the IVC**	
Absence of infrarenal IVC		**Circumcaval ureter**	

Table 3: Summary table with illustrations of variants and anomalies of the IVC (Adapted from Radiographics 2000;20:639-652 [20].)

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ABBREVIATIONS

CT = computed tomography
DVT = deep vein thrombosis
ED = Emergency Department
IVC = inferior vena cava
IVCA = inferior vena cava agenesis
IVCA-aDVT = inferior vena cava agenesis associated deep vein thrombosis
MRI = magnetic resonance imaging
NR = normal range
PE = pulmonary embolus
US = ultrasound
VTE = venous thromboembolism

KEYWORDS

Inferior vena cava agenesis; deep vein thrombosis; inferior vena cava agenesis associated deep vein thrombosis; inferior vena cava anomalies; venous thromboembolism; IVC

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