

Extensive deep venous thrombosis in a young male with absent infrarenal inferior vena cava

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ABSTRACT

A previously well, independent 20-year-old man presented with a 4-day history of progressive left lower limb pain with associated phlegmasia cerulea dolens. Duplex venous ultrasound examination and computed tomography venogram revealed extensive deep vein thrombus from the left popliteal vein to abnormal venous vasculature proximally. Notably, no infrarenal inferior vena cava was detected, with distal venous return channeled through lumbar and visceral collateral channels into the azygous system. Treatment included systemic anticoagulation, catheter-directed thrombolysis, and prolonged therapeutic anticoagulation. In the absence of other risk factors, anatomical abnormalities should be considered in young, well patients presenting with lower limb venous thrombosis. (*J Vasc Surg Cases Innov Tech* 2022;8:146-50.)

Keywords: Deep vein thrombosis; Inferior vena cava; Congenital anomaly; Vascular malformations; 3D reconstruction

Anomalies of the inferior vena cava (IVC) are rare conditions affecting less than 1% of the population, with agenesis of the infrarenal IVC being an uncommon subset.¹ The most common presentation is extensive lower limb (LL) deep venous thrombosis (DVT) in young, well patients without pre-existing risk factors. Up to 5% of DVT in this demographic is attributable to anatomic variations.²

We present a case of IVC agenesis leading to extensive DVT in a well 20-year-old man who consented for this publication. We highlight the treatment pathways and the need to consider lifestyle factors for management.

CASE REPORT

A 20-year-old man with no previous medical history presented to the emergency department with a 4-day history of progressive, atraumatic, left-sided lower abdominal and LL pain.

Initial left flank pain progressed into the lower abdomen and left leg, exacerbated by weight-bearing, especially to the left hip. No pain at rest was reported. There were no further distinguishing features.

No risk factors were elicited from the history for venous thromboembolism: no recent travel, surgery, immobilization, previous or family history of venous thromboembolism. He was a nonsmoker and had no regular medications, history of recreational drug use, or occupational risk factors. The patient led an

active lifestyle, with regular participation in Australian Rules Football.

On examination, there was phlegmasia cerulea dolens of the left LL extending to the thigh. The skin was taut, especially over the lateral thigh, but compartments were soft with no pain on passive stretching. The right LL was unaffected. All LL pulses were palpable, and there were no neurological signs. Abdominal examination was unremarkable.

Full blood examination and coagulation profile showed no abnormality (D-dimer was not performed). Thrombophilia screen later showed heterozygosity for Factor V Leiden mutation. Targeted ultrasound examination of the left leg revealed occlusive thrombus extending from the left popliteal vein to the external iliac vein beyond the view of the ultrasound image. This was further characterized by a computed tomography venogram (Figs 1 and 2), demonstrating extensive clot burden distal to abnormal venous vasculature. An infrahepatic segment of the IVC was absent with a venous cuff below the hepatic portion of the IVC; caval drainage continued into the azygous system, grossly dilated with multiple lumbar and visceral collaterals. Distal to this, extensive thrombus burden was found in the left common iliac vein extending into the internal and external branches, along with thrombus in the right internal iliac vein.

The patient was initially managed with continuous heparin infusion, full length leg compression, and elevation. Subsequent catheter-directed thrombolysis was performed. Left popliteal vein puncture with an 8F introducer sheath was performed, followed by passage of a 5F Bern catheter and Glidewire to the venous cuff. Cavogram demonstrated well-developed collaterals draining into the azygous system and confirmed the absence of the infrarenal IVC. An 8F AngioJet catheter was advanced to the venous cuff with urokinase power pulse performed with 60-minute waiting time. Three runs of the AngioJet were performed with resultant partial recanalization. A 20-cm thrombolysis catheter was placed with the tip in the venous cuff, and urokinase thrombolysis was performed over 24 hours. Follow-up venogram demonstrated patent iliac veins with residual

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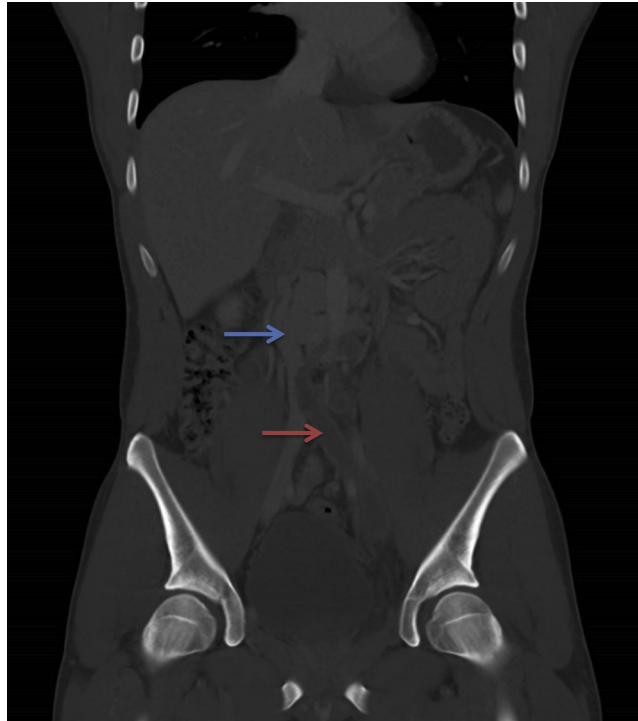


Fig 1. Coronal slice of the portal venous phase of the computed tomography (CT) venogram of the patient on admission to hospital before thrombectomy showing a clot in the left common iliac (red arrow) and venous bulb at a level above bifurcation of iliac veins (blue arrow).

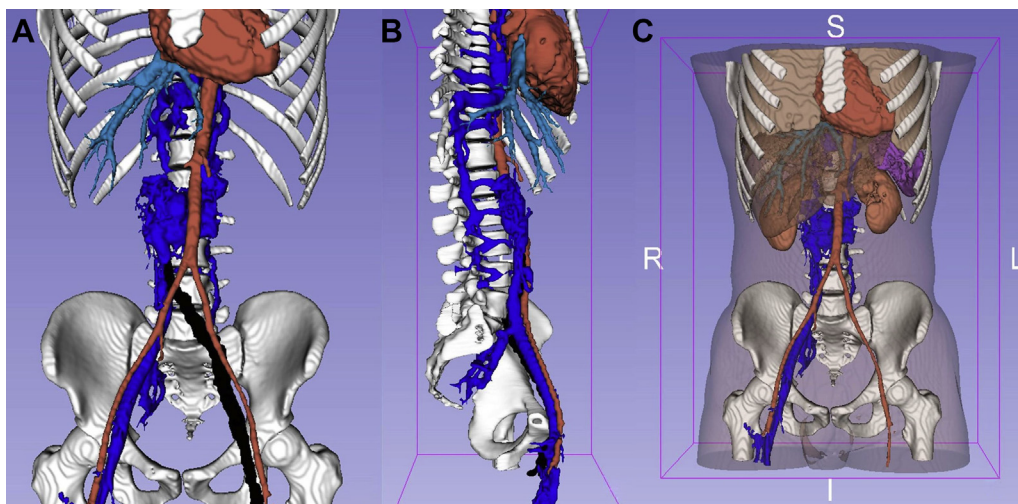


Fig 2. Three-dimensional reconstruction of the patient's anatomy with the inferior vena cava (IVC) in light blue, arteries in red, femoral and azygous veins in dark blue, and left femoral and iliac vein clot in black. Some smaller collateral veins have been removed for clarity. from left to right: **(A)** anteroposterior (AP) view of the vasculature, **(B)** view from the right with the skeleton cropped, and **(C)** AP view with organs in situ.

nonocclusive thrombus (Fig 3). Irregular stenosis along the iliac vein was noted; subsequent passes of the AngioJet and venoplasty with a 12-mm balloon resulted in significantly improved vessel caliber and flow on venogram. Heparin infusion was ceased and the patient was discharged with apixaban 5 mg twice daily and hematology follow-up.

Follow-up. At 3 months of follow-up, the patient had no further symptoms, and was tolerating apixaban. LL swelling was minimal after thrombolysis with compression stockings. Apixaban 5 mg twice daily was continued until the following review and LL compression was reduced to an as-needed basis for symptom control.

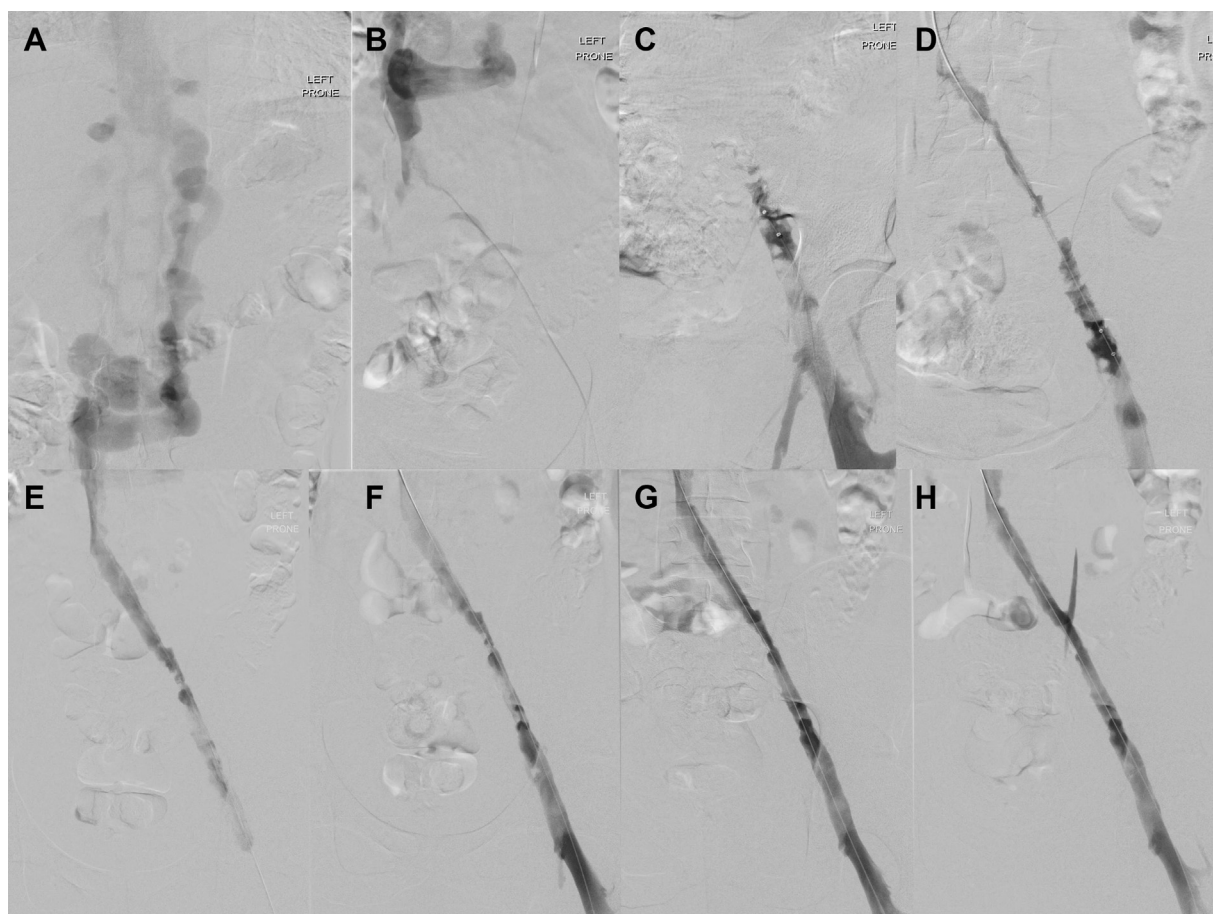


Fig 3. Images from angiography. Initial angiography showing the patient's thoracic (A) and lumbar (B) venous anatomy along with filling defects in the left iliac veins (C, D). Check venogram after 24 hours of thrombolysis showing residual filling defects along left iliac veins (E, F), and final images from completion venogram after angioplasty with a 12-mm balloon (G, H).

After 6 months, the patient continued to be asymptomatic and had expressed wishes to continue playing contact sports. After discussion and counseling on the risks involved with contact sports, a shared decision was made with the patient and family to reduce the apixaban dose to 2.5 mg twice daily to balance bleeding and thromboembolism risks in the setting of permanent risk factors.³ The patient was counseled to present to the emergency department immediately after any significant head impacts.

The hematology department will continue to review and personalize the treatment regime to best balance the risk-benefit profile of management.

DISCUSSION

Risk factors for DVT include genetic (eg, thrombophilia), congenital malformations, and acquired factors such as recent surgery, infections, and malignancy.⁴⁻⁶ The main predisposing factor in this patient was an absent infrarenal IVC—a rare venous malformation with prevalence estimated at 0.3%–0.5% of the population.^{3,7} However,

it may be responsible for up to 5% of idiopathic DVTs in young patients without other identifiable risk factors.^{7,8}

The infrarenal IVC develops from three paired veins at 4–8 weeks of gestation: the posterior cardinal, subcardinal, and supracardinal veins.⁸⁻¹⁰ These form the IVC below the retrohepatic portion, along with the azygous and hemiazygous systems and the iliac veins. The absence of the infrarenal portion of the IVC is among the rarest of the anomalies described and is caused by failure of the posterior cardinal and supracardinal veins with preservation of the suprarenal segment.¹¹ In these patients, blood from LLs is shunted through multiple collaterals—including the azygous and hemiazygous systems—emptying into the SVC. It has also been suggested that thrombosis of the IVC during development could lead to this condition.^{12,13} Anatomical drainage for patients with absent IVC predisposes to thrombosis, as distal venous shunting through collaterals results in venous stasis.^{14,15} Many cases subsequently present with DVT, with

common precipitants being major physical exertion, anabolic steroids, and immobilization.^{16,17}

Patients with this condition are generally male, under 40 years of age, and without other identifiable risk factors for thrombosis, presenting with DVT.^{9,18,19} Thrombus burden can extend into the pelvis, leading to abdominal and lumbar pain; subsequent clot extension into the ILLs may result in edema, venous insufficiency, and eventually ulceration.^{15,20}

The mainstay of treatment is medical therapy.^{10,16} Oral anticoagulation has been used successfully with adjunctive lifestyle modifications including compression stockings, cessation of smoking, and avoiding long periods of immobilization.²¹ These effective measures have been almost universally described for this patient population; however, duration of anticoagulation therapy remains controversial.^{9,15} Close clinical follow-up is required for these patients to monitor bleeding and thrombosis. In this case, this was of particular importance due to ongoing participation in contact sports; thus, an adapted anticoagulation plan was used after discussion with the patient. Few reports exist of surgical bypass for this condition,^{19,22,23} with the first being reported by Dougherty in 1996.²⁴ In the acute setting, thrombolysis has been demonstrated to successfully remove clot burden, with multiple modalities described.²⁵⁻²⁷

In this case, catheter-directed thrombolysis was used to acutely improve vessel patency in conjunction with systemic heparin infusion. Other endovascular interventions are also available including mechanical clot retrieval and ultrasound-driven techniques.^{26,28-30}

CONCLUSIONS

We demonstrate a successful case of catheter-directed thrombolysis in the treatment of extensive pelvic DVT in a patient with segmental infrarenal IVC agenesis. Along with consideration of acquired and genetic risk factors, vascular malformation should be a differential when investigating young patients with DVT. Acute treatment may include systemic and directed thrombolysis. Although long-term anticoagulation is still a topic for debate, there is a role for personalized treatment strategies to balance the risks of bleeding with thromboembolism.

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