

# Duplication of inferior vena cava and coagulation mutations with left-sided iliofemoral venous thrombosis

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## ABSTRACT

Duplication of the inferior vena cava (IVC) with coagulation mutations in the form of factor V Leiden and *MTHFR* mutations represents an unusual subset of patients. We are reporting a case of a 43-year-old man who presented with left iliofemoral deep venous thrombosis diagnosed on duplex ultrasound scan. At the time of catheter-directed thrombolysis with prophylactic IVC filter placement, a duplicated IVC system was observed. After thrombolysis, a stenotic lesion in the left common iliac vein and IVC was stented. IVC filters were retrieved after 6 weeks. On thrombophilia profile testing at 3 months, he was also found to have factor V Leiden and *MTHFR* mutations. After 12 months of follow-up, the patient is asymptomatic with a patent ilio caval venous system and is receiving lifelong anticoagulation. (J Vasc Surg Cases and Innovative Techniques 2019;5:26-30.)

**Keywords:** Duplicated IVC; Catheter-directed thrombolysis; Deep venous thrombosis

There are various types of inferior vena cava (IVC) anomalies, among which duplication is a more common one with a prevalence of 0.2% to 3.0%.<sup>1</sup> However, duplication of the IVC associated with coagulation mutation is a highly under-reported entity that increases the risk of deep venous thrombosis (DVT) significantly. We are reporting a case in which duplication of the IVC was complicated by left lower limb iliofemoral DVT and the patient was subsequently diagnosed to have coagulation mutations as well. Consent to publish this case report was obtained from the patient.

## CASE REPORT

A 43-year-old man presented with sudden-onset pain and swelling over the left lower limb for 1 day. There was no history of trauma, infection, or any other provoking factor for DVT. There was no history suggestive of pulmonary thromboembolism. On examination, the patient had a grossly swollen and tense left lower limb with significant calf tenderness. Arterial examination findings were normal. Duplex ultrasound assessment revealed that the left external iliac vein, common femoral vein, superficial femoral vein, and popliteal vein were dilated and filled with

acute thrombus; the thrombus was also seen extending into the great saphenous vein. The right-sided deep venous system was normal. Routine blood investigation results were normal. Intravenous anticoagulation was started immediately. In view of unabated pain and tense swelling that did not resolve with 24 hours of anticoagulation and involvement of both the superficial and deep venous systems of the left lower limb, it was decided to subject the patient to catheter-directed thrombolysis.

Initially, a Denali IVC filter (Bard Peripheral Vascular, Tempe, Ariz) was placed in the infrarenal part of the IVC after cavography was performed through the right internal jugular vein (routine protocol at our institution before catheter-directed pharmacomechanical thrombolysis). The left popliteal vein was punctured under ultrasound guidance. A guidewire with catheter was advanced into the common iliac vein and IVC. To our surprise, the wire did not traverse through the filter; instead, it moved in a separate track parallel to the IVC with filter. Contrast-enhanced venography confirmed this segment to be the left-sided IVC. As thrombolysis was planned and per our protocol, an IVC filter was deployed in the left-sided IVC through the right internal jugular vein approach, and the right-sided IVC filter was kept in situ to prevent post-thrombolysis bleeding from the filter insertion site of the IVC (Fig 1). Mechanical thrombolysis was performed using an 8F guiding catheter, and catheter-directed thrombolysis was performed using a multi-hole infusion catheter with alteplase. Alteplase (1 mg/h) and unfractionated heparin were infused through the catheter and sheath, respectively. Serum fibrinogen level and activated partial thromboplastin time were monitored every 4 hours. After overnight infusion for 12 hours, check venography was performed, which showed good clearance of the thrombus load with minimal thrombus in the iliac vein and a long-segment residual stenosis in the left IVC, common iliac vein, and external iliac vein. The minimal amount of thrombus found was aspirated through a guiding catheter followed by venoplasty and stenting of the left ilio caval system with an 18- × 100-mm Wallstent (Boston

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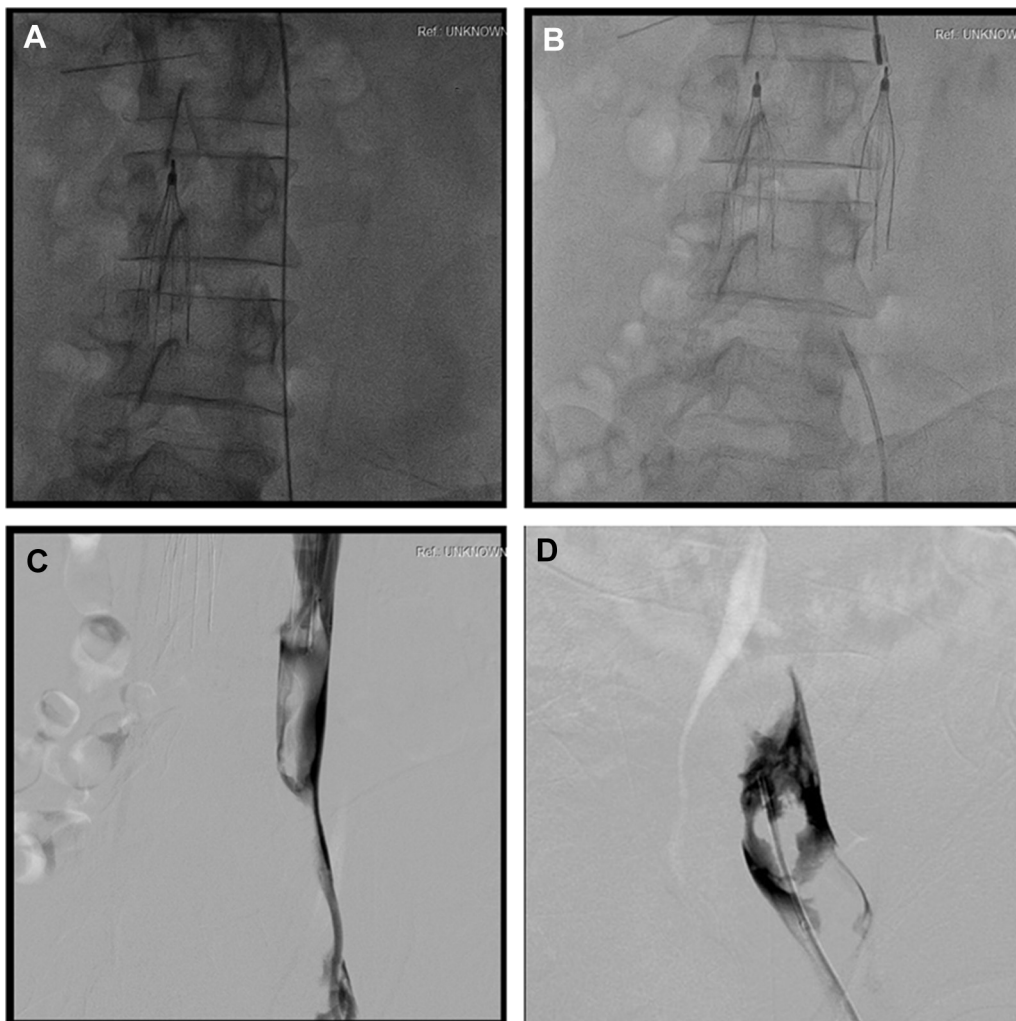
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**Fig 1.** Inferior vena cava (IVC) filter placement and thrombolysis. **A**, Filter in right IVC and guidewire traversing through left IVC. **B**, Filters in both IVCs. **C**, Thrombus in IVC below the filter. **D**, Thrombus in the left iliac vein.

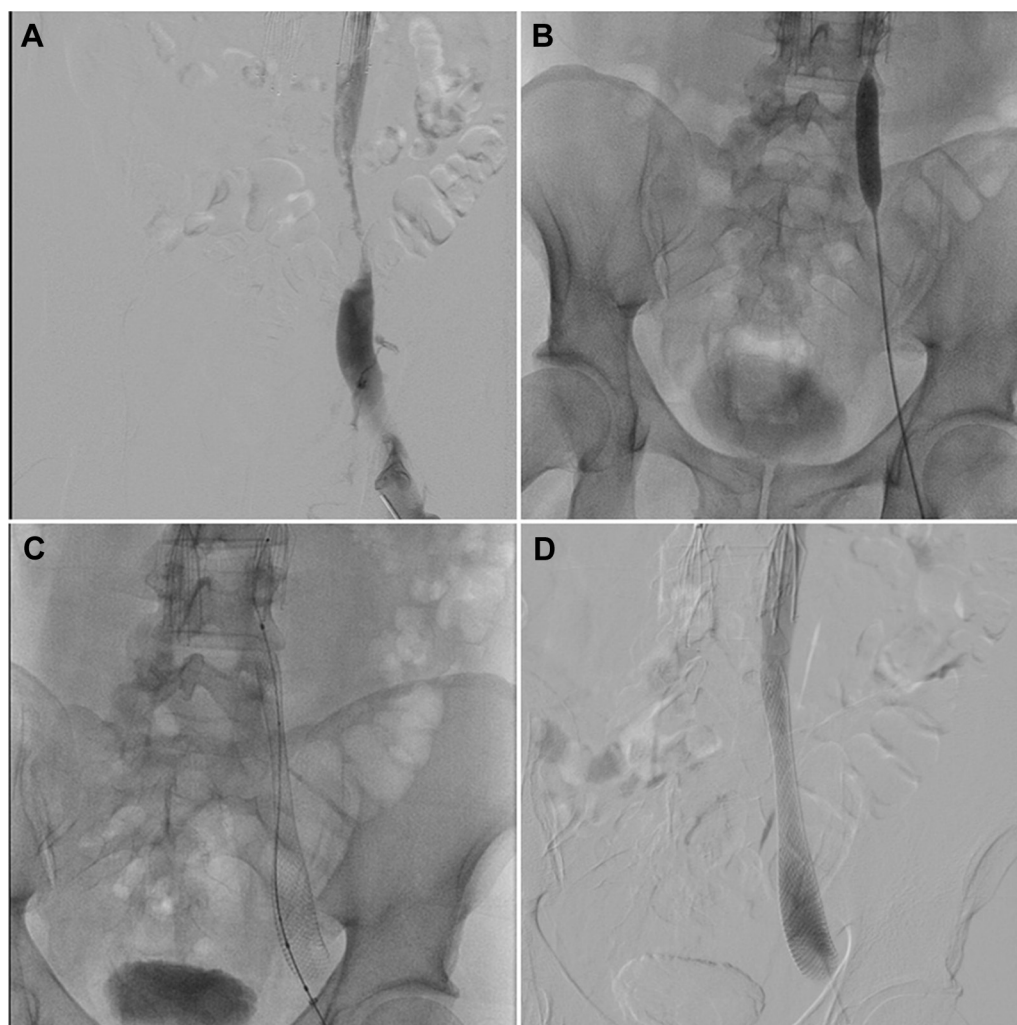
Scientific, Marlborough, Mass). The size of the stent was chosen by measuring the proximal IVC under fluoroscopy. After stenting, good flow of contrast material was noted (Fig 2). The patient had dramatic improvement in limb swelling and pain. The patient received enoxaparin 60 mg twice a day for 5 days, aspirin 75 mg (lifelong), and graduated compression stockings in the left lower limb. Later, enoxaparin was converted to dabigatran. Computed tomography venography at 1 month showed widely patent IVCs and filters (Fig 3). Both IVC filters were removed after 6 weeks (Fig 4).

After 3 months, D-dimer levels were found to be normal and anticoagulation was stopped for 2 weeks to ascertain the thrombophilia profile, which showed factor V Leiden mutation and *MTHFR* mutation. Anticoagulation with dabigatran was restarted and continued lifelong. In follow-up at 6 months and 12 months, duplex ultrasound scans showed good phasic flows in the deep venous system of the left lower limb with no clinical findings suggestive of post-thrombotic syndrome.

## DISCUSSION

The IVC develops from subcardinal, supracardinal, and posterior cardinal veins. Both posterior cardinal veins fuse to form the iliac veins and their confluence. The right supracardinal vein persists and develops into the infrarenal IVC, and the left supracardinal vein regresses normally. Right subcardinal-supracardinal anastomosis develops into the renal part of the IVC, and the right subcardinal veins along with the hepatic vein develop into the rest of the IVC. Failure of regression of the left supracardinal vein leads to duplication of the IVC.<sup>2</sup>

The incidence of IVC anomalies is estimated at 0.07% to 8.7% of the population; however, the incidence of IVC anomalies discovered in individuals presenting with a DVT is 5.1% to 6.7%. Patients presenting with DVT and IVC anomalies are younger at presentation compared with the persons with DVT



**Fig 2.** Post-thrombolysis venography and stenting. **A**, Residual stenosis in left common iliac vein. **B**, Balloon dilation of stenotic lesion. **C**, Wallstent being deployed. **D**, After stent deployment, good flow of contrast material could be appreciated.

without anomalies.<sup>3-5</sup> Few isolated case reports described the association of congenital coagulation mutations and hypoplasia of the IVC.<sup>6</sup> Few studies consider duplicated IVC to be the cause of DVT, perhaps because of retrograde stasis.<sup>7</sup> Duplication of the IVC may be associated with recurrent venous thromboembolism if the anatomic variation goes undiagnosed.<sup>8</sup> The risk of venous thromboembolism might be further multiplied if it is associated with coagulation mutations. The absolute increase in risk is still an unknown entity.

Phlebography is considered the “gold standard” in diagnosis of duplicated IVC, but this is invasive.<sup>9</sup> It is generally performed when fluoroscopy-guided catheter-directed thrombolysis is planned. In general, duplication of the IVC is an incidental finding on computed tomography

scan done for some other indication. In this case, the patient was diagnosed with a duplicated IVC on phlebography.

Treatment of a duplicated IVC depends on the clinical presentation. Asymptomatic duplication of the IVC may be observed.<sup>7</sup> The patients presenting with DVT may receive anticoagulation or catheter-directed thrombolysis. There are only a couple of reports in which duplicated IVC with DVT was treated by catheter-directed thrombolysis.<sup>9</sup> In this case, catheter-directed thrombolysis was performed for DVT in view of a painful tense limb with superficial venous thrombosis and DVT.

There are some isolated reports of permanent double IVC filter placement in patients with congenital duplication of the IVC complicated by recurrent



**Fig 3.** Computed tomography venography image showing patent inferior vena cava (IVC) and filters bilaterally. Left renal vein is draining in left-sided IVC.

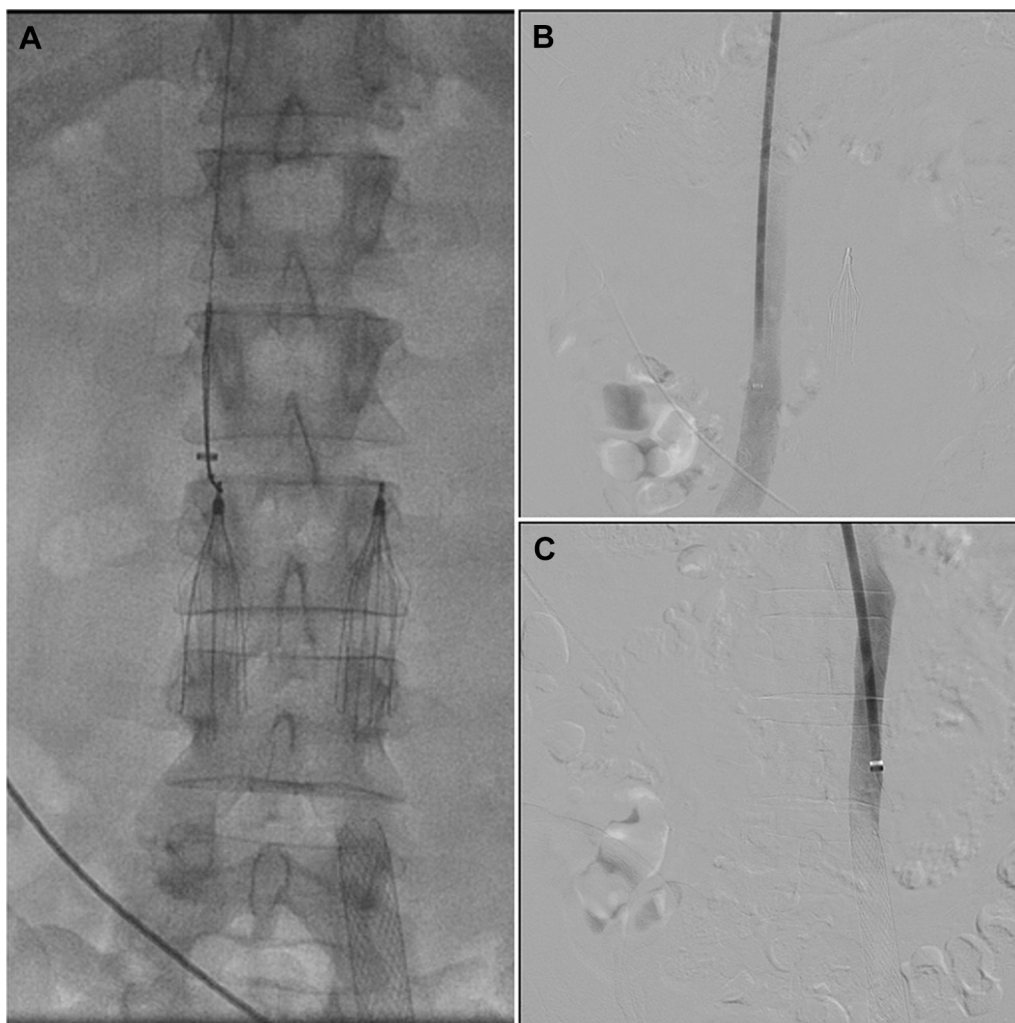
pulmonary thromboembolism.<sup>10</sup> In our institution, there is a routine protocol of IVC filter insertion before proceeding to pharmacomechanical thrombolysis.

Iliocaval venous stenting is indicated in patients with nonthrombotic iliac vein lesions, in patients with chronic post-thrombotic iliocaval disease affecting the quality of life, and as an adjunctive therapy for residual thrombosis after thrombolysis in patients with acute DVT.<sup>11</sup> In our case, the patient had residual left iliac vein thrombosis after thrombolysis. Iliac vein stenting was performed to avoid the post-thrombotic syndrome, and anticoagulation was given in the postoperative phase for 6 months. Now, the patient is being followed up with annual duplex ultrasound scan or interim

duplex ultrasound scan in the presence of signs or symptoms of DVT to look for any new-onset stenosis or thrombosis.

### CONCLUSIONS

Venous thromboembolism is a rare complication of duplicated IVC; however, the risk is significantly increased by coagulation mutations. Catheter-directed thrombolysis was found to be a safe and effective means of management of DVT in such a duplicated system and unmasked a stenotic lesion in the iliac vein and left-sided DVT. Further evaluation will be required to assess the association of congenital coagulation mutations with caval anomalies and to accurately quantify the risk of venous thromboembolism.



**Fig 4.** Inferior vena cava (IVC) filter removal. **A**, Both IVC filters in situ. **B**, Right-sided IVC filter removed; good flow of contrast material can be seen. **C**, Left-sided IVC can be visualized cleared of thrombus.

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