EJVES Vascular Forum (2021) 52, 5—10

CASE REPORT

Endovascular IVC Reconstruction in an 18 Year Old Patient with Subtotal IVC Atresia

Matthew L. Hung, Dennis Kwon, Deepak Sudheendra *

Division of Interventional Radiology, Department of Radiology, Hospital of the University of Pennsylvania, Philadelphia, PA, USA

Introduction: Inferior vena cava (IVC) atresia is an uncommon venous anomaly that is an under recognised cause of unprovoked acute deep venous thrombosis (DVT) in young adults. The purpose of this case report is to highlight endovascular IVC reconstruction as a feasible treatment option, particularly in challenging cases when other therapeutic modalities have failed.

Report: This is the report of an 18 year old patient with near complete IVC atresia and a longstanding history of exertional nausea of unknown aetiology, who presented with extensive acute DVT. He was treated successfully by endovascular IVC reconstruction after failing initial anticoagulation and thrombolysis. Symptom resolution and venous patency were maintained at 2.5 year follow up.

Discussion: IVC atresia is an important aetiology to consider in a young patient presenting with unprovoked DVT. Endovascular stenting can restore venous patency and is feasible even when there is near complete IVC atresia. This case was uniquely challenging in the length of atretic IVC that was reconstructed and also highlights an atypical clinical presentation of IVC atresia.

Article history: Received 4 January 2021, Revised 23 April 2021, Accepted 1 June 2021,

Keywords: Deep venous thrombosis, Endovascular intervention, IVC atresia, IVC reconstruction, Post thrombotic syndrome, IVC agenesis

INTRODUCTION

Inferior vena cava (IVC) atresia is a rare venous anomaly, occurring in 0.15—1% of the population.[1] Although patients with IVC atresia may be asymptomatic, some may become symptomatic in childhood or adulthood, most commonly presenting with unilateral or bilateral iliac deep venous thrombosis (DVT). Other patients may present with chronic venous insufficiency[1] or exhibit atypical symptoms, such as exertional nausea.[2] While there is no consensus on the optimal management of these patients, treatment strategies include anticoagulation, catheter directed thrombolysis, surgical bypass grafts, and endovascular reconstruction.[3—5]

This is the report of a technically challenging case of near complete IVC atresia in a patient with a long history of exertional nausea of unknown aetiology who presented with acute bilateral lower extremity DVT. The patient was treated successfully by endovascular IVC reconstruction after failed thrombolysis with complete resolution of symptoms and continued patency.

REPORT

A 16 year old male with a history of hereditary haemorrhagic telangiectasia presented initially to an outpatient clinic with nine months of progressive exertional nausea and dry heaving. Symptoms were most severe after intense exercise. The physical exam was unremarkable and no superficial abdominal collaterals were seen. Cardiology and allergy and immunology consultants attributed the symptoms to motion sickness. Stress echocardiogram and interrogated labs were unrevealing. The patient was given antihistamines with poor relief of symptoms.

Two years later while playing basketball, the patient represented to an outside hospital with sudden onset of nausea, back pain, and bilateral calf pain and swelling. Computed tomography (CT) venography demonstrated occlusive DVT bilaterally from the common iliac to the popliteal veins. The infrahepatic IVC was not visualised and replaced with collateral veins. Hypercoagulability workup was negative and the patient did not have a history of DVT. The patient underwent catheter directed thrombolysis and mechanical thrombectomy with improved symptoms and was discharged on anticoagulants. However, he was readmitted within 24 hours with recurrent bilateral iliofemoral vein thrombosis and worsening lower extremity pain

This case was presented at the 2018 Vascular Interventional Advances (VIVA) Conference, Las Vegas, NV, 5—8 November 2018.

* Corresponding author. Department of Radiology, Hospital of the University of Pennsylvania, 3400 Spruce Street, Philadelphia, PA 19104, USA.
E-mail address: dr.sudi@gmail.com (Deepak Sudheendra).

https://doi.org/10.1016/j.ejvsvf.2021.06.001

and oedema. He was transferred to the present authors' institution for further management.

Pre-intervention CT venogram confirmed extensive lower extremity DVT, partial absence of the intrahepatic IVC, complete absence of infrahepatic IVC, and dilated thrombosed paraspinal collateral veins draining into enlarged azygos and hemiazygous veins (Fig. 1). The patient was taken to the interventional radiology suite daily for three consecutive days.

Mechanical thrombectomy and thrombolysis

On the first day, bilateral lower extremity venography was performed via posterior tibial vein access, demonstrating a clot burden from the bilateral posterior tibial veins to the common iliac veins (CIVs) (Fig. 2A). The IVC was not visualised from the confluence of the CIVs and no significant abdominopelvic collaterals were identified (Fig. 2B). Mechanical thrombectomy with Angiojet (Boston Scientific, Marlborough, MA, USA) was performed bilaterally in the iliac, femoral, popliteal, and posterior tibial veins with marginal success. Cragg-McNamara infusion catheters (Medtronic, Minneapolis, MN, USA) were placed bilaterally, terminating at the confluence of the CIVs, and tPA was infused at 0.5 mg/h in each catheter for 24 hours.

On the second day, repeat venography demonstrated near complete resolution of thrombus throughout both lower extremities and the outflow azygos vein (Fig. 3A). Within 10 minutes of terminating tPA, there was re-thrombosis of the azygos and CIVs (Fig. 3B). Thus, infusion catheters were again placed and tPA infusion was restarted. Given the rapid re-thrombosis, the patient agreed to an attempt at IVC reconstruction to restore venous outflow.

Inferior vena cava recanalisation and stenting

On the third day under general anaesthesia and with the patient in the supine position, venography of the proximal intrahepatic IVC was performed via a right internal jugular vein access, demonstrating complete occlusion of the infrahepatic and distal intrahepatic IVC (Fig. 4A). Venography was performed at the confluence of the CIVs via bilateral common femoral vein access, revealing a small remnant of the IVC (Fig. 4B). Recanalisation of the IVC was performed through this diminutive stub in a caudal to cranial direction using a variety of sharp recanalisation techniques, including a 16 gauge Colapinto needle (Cook Medical, Bloomington, In, USA), back end of a 0.035 Amplatz Super Stiff guidewire (Boston Scientific), and 9 and 12 French vascular sheath dilators as bougies. The atretic IVC was traversed from both the right internal jugular vein and common femoral vein accesses using a 0.035 Roadrunner wire (Cook Medical) and 0.035 Quick-Cross catheter (Spectranetics, Colorado Springs, CO, USA). The right renal vein was catheterised and venography was performed, confirming that the recanalised vessel was the IVC (Fig. 4C). A snare was used to gain body floss access and venoplasty of the atretic IVC was performed from the confluence of the CIVs to the infrahepatic IVC with a 10 mm balloon. Intravascular ultrasound (IVUS) was performed from the right atrium down through the IVC and bilateral common and external iliac

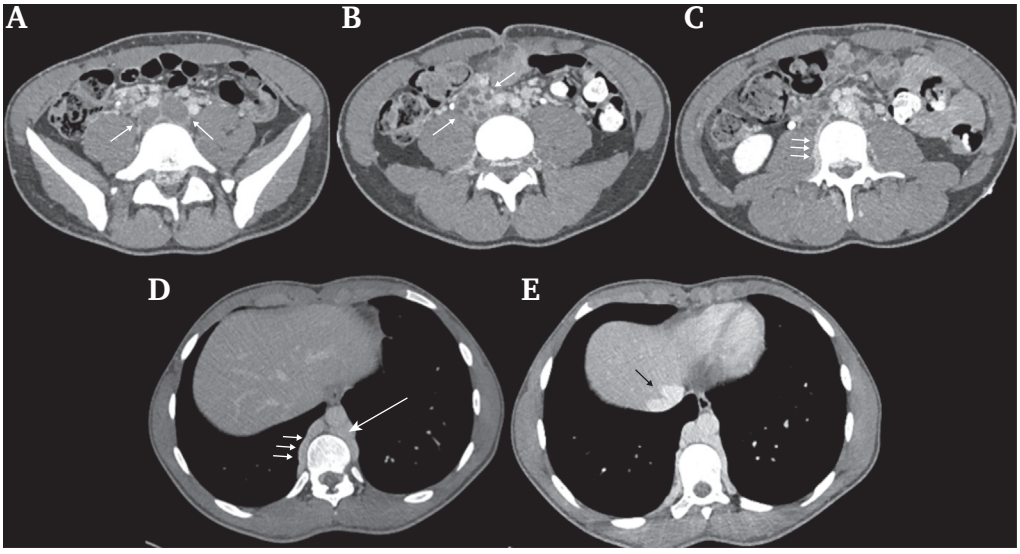

Figure 1. Pre-procedural axial CT venogram images proceeding from a caudal to cranial direction. (A) The bilateral common iliac veins (arrows) are dilated by acute thrombus. (B) At the expected location of the confluence of the common iliac veins, there are enlarged, thrombosed paraspinal venous collaterals (arrows). (C) These collaterals eventually drain into the azygos system (arrows). (D) Superiorly, the azygos vein is patent, but distended (small arrows). The hemiazygous vein is also engorged (large arrow). (E) The IVC is only identified at the superior aspect of the liver, at the confluence of the hepatic veins (arrow).

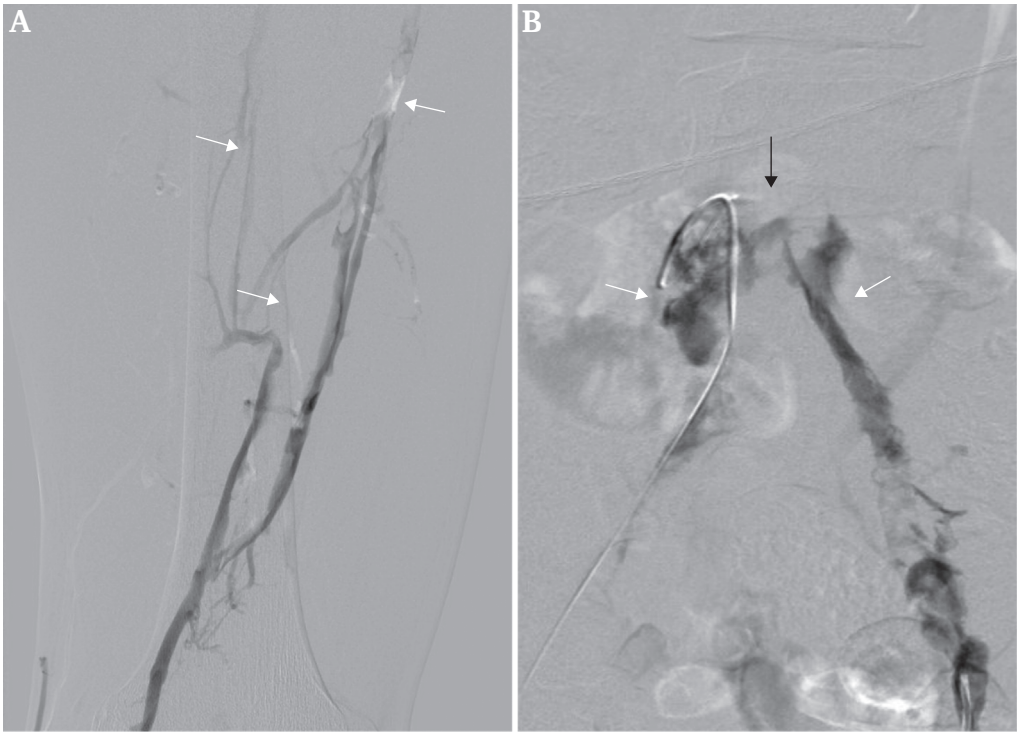

Figure 2. Digital subtraction angiography images from Day 1 thrombolysis and thrombectomy. (A) Initial venography from the left posterior tibial vein demonstrates clot through the femoral veins (arrows). (B) Extensive clot burden is also seen in the bilateral common iliac veins (white arrows). The IVC is absent at its expected location (black arrow).

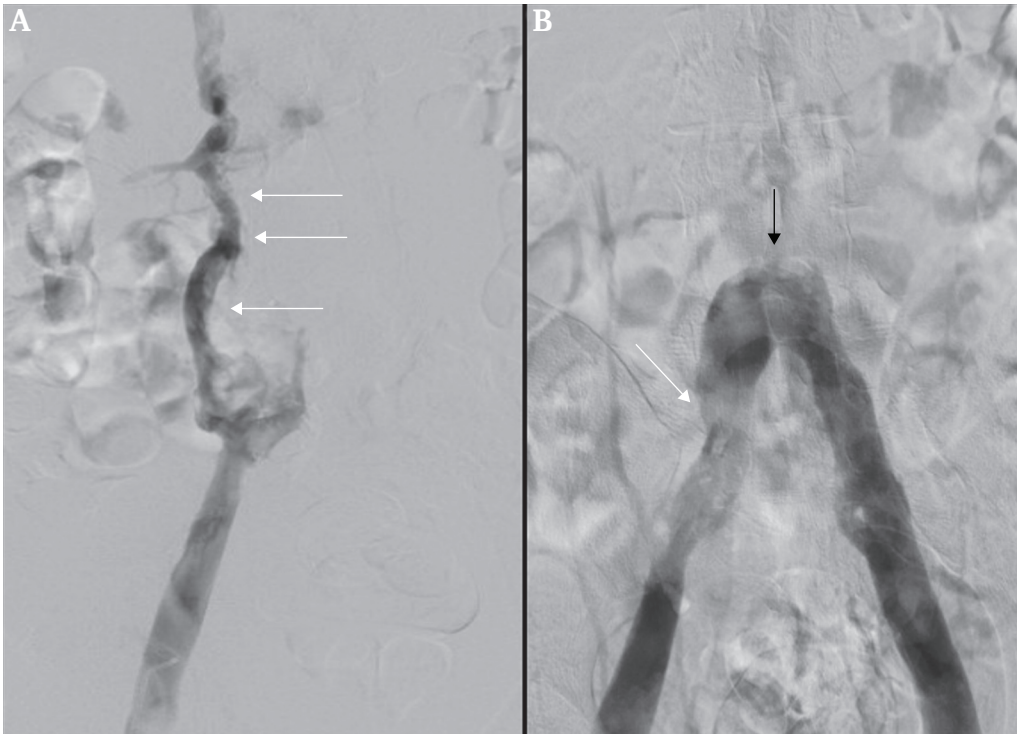

Figure 3. Digital subtraction angiography images from Day 2 lysis check. (A) Following 24 hours of catheter directed thrombolysis, there is near complete resolution of thrombus, and a large outflow azygos vein (arrows) is identified. (B) However, within 10 minutes of terminating the tissue plasminogen activator infusion, the azygos vein is no longer seen on repeat venography. Re-thrombosis also occurred in the right common iliac vein (white arrow). The IVC remained absent at its expected location (black arrow).

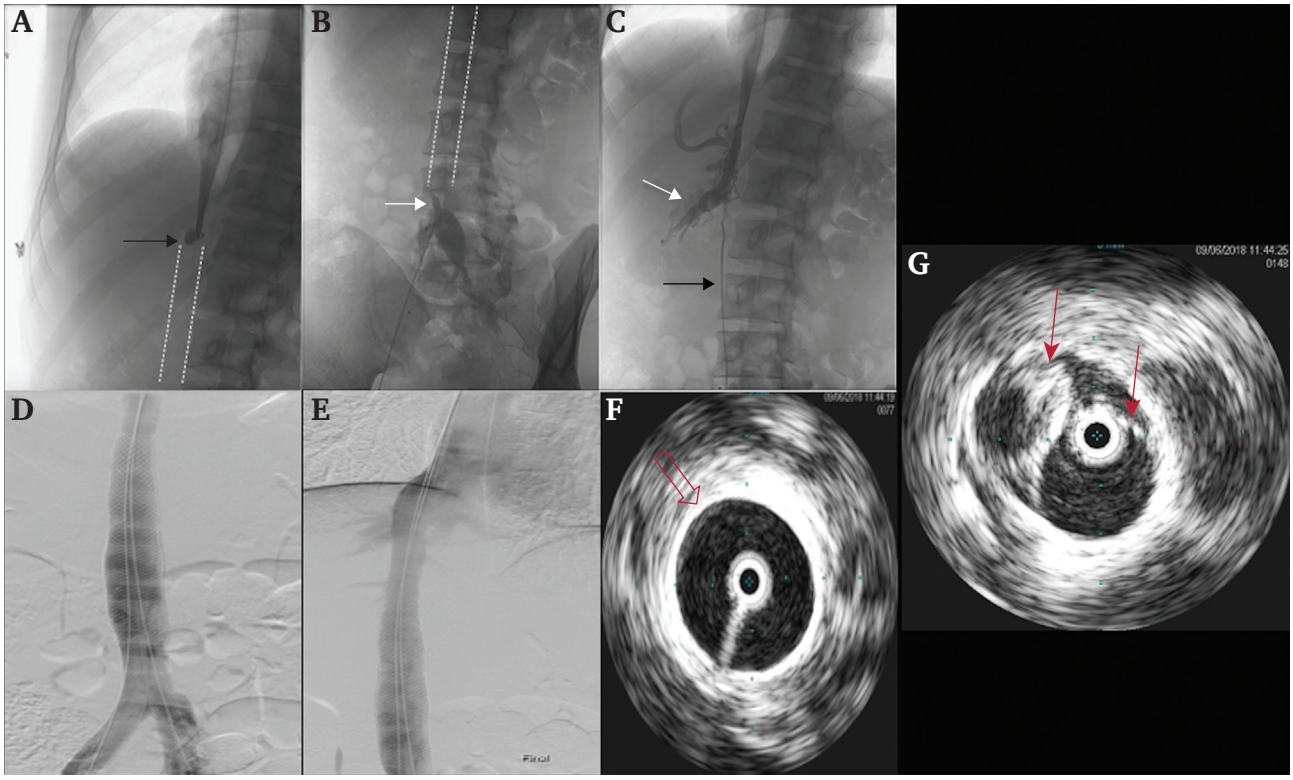

Figure 4. IVC reconstruction on Day 3. (A, B) Fluoroscopic images demonstrate abrupt termination of the proximal IVC at the level of the hepatic veins (black arrow). A small remnant of the IVC (white arrow) is now identified at the confluence of the common iliac veins. The dashed lines outline the expected course of the atretic IVC and the length to be reconstructed. (C) Using several recanalisation techniques and after traversing the atretic IVC (black arrow), venography of the right renal vein (white arrow) confirmed that the recanalised vessel was the IVC. (D, E) After extensive venoplasty, intravascular ultrasound evaluation, and deployment of Wallstents in the IVC and common iliac veins, the final venogram demonstrates a patent, fully reconstructed IVC. (F, G) Intravascular ultrasound demonstrates a recanalised IVC with a patent IVC stent (open red arrow) and well apposed kissing iliac stents (solid red arrows) at the inferior aspect.

veins, revealing extensive synechiae within the IVC and bilateral common and external iliac veins. Venoplasty with 16 mm and 24 mm balloons was performed in the IVC followed by repeat venography and IVUS that showed a reduction in synechiae and venous collaterals with significant recoil throughout the atretic IVC. Two overlapping 24 mm × 70 mm Wallstents (Boston Scientific) were placed in the recanalised IVC, starting in the intrahepatic IVC down to the iliac confluence. These were dilated to 24 mm and repeat venography and IVUS was performed. Two kissing 18 mm × 90 mm Wallstents were placed starting at the caudal aspect of the inferior 24 mm Wallstent and ending in each CIV. The IVC end of the 18 mm Wallstents was dilated to 14 mm and the common iliac end was dilated to 16 mm (Fig. 4D and E). Final venography and IVUS demonstrated a widely patent reconstructed IVC with good wall apposition of the stents (Fig. 4F and G).

The patient was discharged on post-operative day four on enoxaparin 80 mg SC BID (1 mg/kg), and clopidogrel 75 mg PO daily for one month. At the one month clinic visit, CT venography demonstrated a patent reconstructed IVC and bilateral iliac veins (Fig. 5, Supplemental Video 1). The patient was transitioned to lifelong apixaban 5 mg PO BID and aspirin 81 mg PO daily. He reported complete resolution of exertional nausea and denied lower extremity pain or swelling at the one and seven month post-procedural clinic visits and remains asymptomatic at 2.5 years of follow up. The patient will be seen annually going forward.

DISCUSSION

IVC atresia is an uncommon entity and is often asymptomatic in patients with well developed azygos/hemiazygos continuation.[1] However, it is increasingly recognised as an important aetiology to consider when a young patient presents with unprovoked iliofemoral DVT, especially if bilateral.[6] Ruggeri *et al*. estimated IVC atresia to be present in 5% of cases of DVT in patients younger than 30 years.[7] Furthermore, these patients may initially present with atypical symptoms. The present patient had exertional nausea probably because of poor venous return during strenuous activity, which was also observed in another case report.[2]

IVC atresia, IVC agenesis, and congenital absence of the IVC have been used interchangeably within the literature. Although it is nearly impossible to differentiate these conditions in the absence of prior imaging, they are distinct. IVC agenesis implies failed development of the subcardinal,

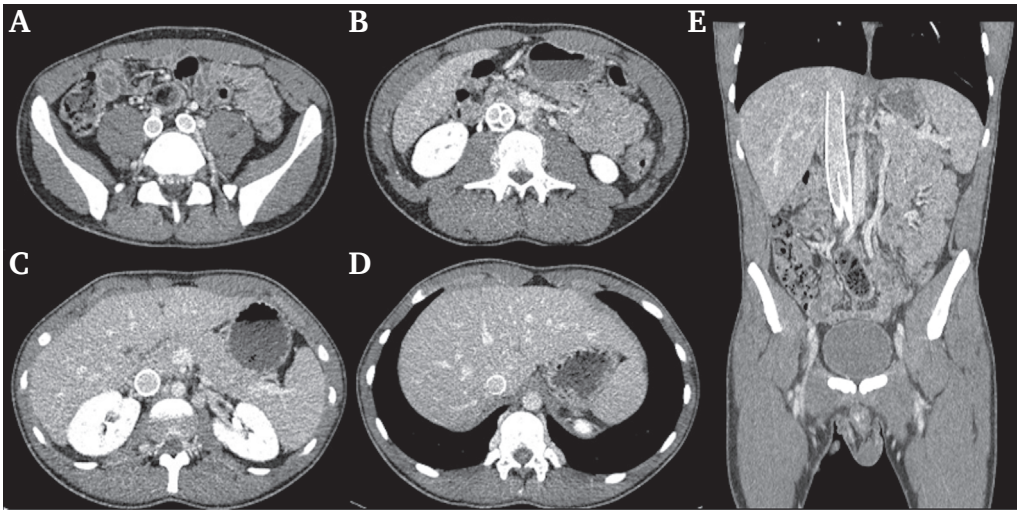

Figure 5. Post-procedural CT venogram images at one month. Representative axial images (A—D) and coronal image (E) at the iliac veins (A), distal IVC (B), renal IVC (C), and intrahepatic IVC (D) demonstrate patency of the kissing 18 mm Wallstents and 24 mm Wallstent, which are free of thrombus.

supracardinal, or posterior cardinal veins. IVC atresia, on the other hand, results from perinatal insult or thrombosis that leads to fibrosis and closure of the IVC. Many patients with an apparently absent IVC probably have IVC atresia. Mabud *et al.* recently reported a series of 17 patients with IVC atresia and noted a history of hereditable thrombophilia in 52.9% of patients.[8] There was a history of neonatal central lines, abdominal malignancy and/or neonatal surgery in 62.5% of the remaining patients. The distinction is also a technical consideration, as the creation of a "neocava" in IVC agenesis is more challenging than IVC reconstruction in IVC atresia, and potentially unfeasible.

Because of the rarity of IVC atresia, there is no consensus on the optimal management of these patients. A recent two centre study of 18 paediatric patients with IVC atresia and DVT managed with long term therapeutic anticoagulation, with or without thrombolysis, reported a re-thrombosis rate of 17%.[9]

Catheter directed thrombolysis of iliofemoral DVT in IVC atresia appears to be effective with high reported technical success rates, defined as near complete resolution of thrombus with observed venous outflow through collaterals, and no observed cases of re-thrombosis in a small series.[3] However, in the present patient both mechanical thrombectomy and thrombolysis initially failed, which led to exploration of endovascular stenting as a treatment option. Endovascular stenting of the atretic IVC has been reported to have high technical success rates in a few cases,[2,5,8,10] although re-intervention rates are as high as 42.9%.[8] The technical approach in the present patient was similar to previously reported cases, but was uniquely challenging in the length of the atretic IVC that was reconstructed. Whereas prior cases usually had atresia of either the suprarenal or infrarenal IVC, the present patient had near complete IVC atresia with the most superior intrahepatic IVC as the only patent segment.

In conclusion, this is the report of a technically challenging case of endovascular IVC reconstruction in a patient with near complete IVC atresia. Further studies are needed to determine optimal patient selection and long term outcomes after endovascular stenting.

FUNDING

None.

CONFLICT OF INTEREST

Dr Sudheendra is a consultant for Boston Scientific, Vesper Medical, and Sirtex; Dr Hung and Dr Kwon report no conflicts of interest.

APPENDIX A. SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ejvsvf.2021.06.001.

REFERENCES

1 Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. *Eur J Radiol* 2007;**62**:257—66.

2 Schiffman MH, Cornman-Homonoff J, Sos TA. Percutaneous orthotopic IVC construction in a pediatric patient with symptomatic IVC agenesis. *Cardiovasc Intervent Radiol* 2019;**42**:308—12.

3 Broholm R, Jørgensen M, Just S, Jensen LP, Bækgaard N. Acute iliofemoral venous thrombosis in patients with atresia of the

inferior vena cava can be treated successfully with catheter-directed thrombolysis. *J Vasc Interv Radiol* 2011;**22**:801—5.
4 Sagban TA, Grotemeyer D, Balzer KM, Tekath B, Pillny M, Grabitz K, et al. Surgical treatment for agenesis of the vena cava: a single-centre experience in 15 cases. *Eur J Vasc Endovasc Surg* 2010;**40**:241—5.
5 Haskal ZJ, Potosky DR, Twaddell WS. Percutaneous endovascular creation of an inferior vena cava in a patient with caval agenesis, Budd-Chiari syndrome, and iliofemorocaval thrombosis. *J Vasc Interv Radiol* 2014;**25**:63—9.
6 Tufano A, López-Jiménez L, Bikdeli B, Garcia-Bragado F, Mazzolai L, Amitrano M, et al. Inferior vena cava agenesis in patients with lower limb deep vein thrombosis in the RIETE registry. When and why to suspect. *Int J Cardiol* 2020;**305**:115—9.
7 Ruggeri M, Tosetto A, Castaman G, Rodeghiero F. Congenital absence of the inferior vena cava: a rare risk factor for idiopathic deep-vein thrombosis. *Lancet* 2001;**357**:441.
8 Mabud TS, Sailer AM, Swee JKY, Tamboli M, Arendt VA, Jeon G, et al. Inferior vena cava atresia: characterisation of risk factors, treatment, and outcomes. *Cardiovasc Intervent Radiol* 2020;**43**:37—45.
9 Tarango C, Kumar R, Patel M, Blackmore A, Warren P, Palumbo JS. Inferior vena cava atresia predisposing to acute lower extremity deep vein thrombosis in children: a descriptive dual-center study. *Pediatr Blood Cancer* 2018;**65**:e26785.
10 Murphy EH, Johns B, Varney E, Raju S. Endovascular management of chronic total occlusions of the inferior vena cava and iliac veins. *J Vasc Surg Venous Lymphat Disord* 2017;**5**:47—59.