Extravascular reconstruction of a congenitally absent inferior vena cava

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ABSTRACT

Congenital absence of the inferior vena cava is an uncommon venous anomaly with treatment algorithms consisting of predominately medical management. We present a case of a 36-year-old man with venous ulcers who had failed conservative treatment for recurrent venous ulcers. From a catheter directed approach, we were able to develop an extravascular retroperitoneal space and perform an iliocaval reconstruction with Wallstents. At 1-year postoperatively, his leg pain and edema had resolved, and had achieved resolution of his venous ulceration. (J Vasc Surg Cases and Innovative Techniques 2020;6:681-5.)

Keywords: Inferior vena cava reconstruction extravascular reconstruction; Congenital absence of inferior vena cava; Iliocaval obstruction

Congenital absence of the inferior vena cava (IVC) and its venous tributaries is an uncommon but long recognized anomaly that was first described by Abernethy in 1793.[1] Given the relative rarity of true caval agenesis, a paucity of literature exists on its management in the adult population. Historically, treatment algorithms consist of medical management only, with anticoagulation and compression; thrombolysis was reserved for select cases with extensive acute thrombus burden and severe symptoms. We present a case of congenital absence of the IVC and a somewhat novel application of previously described endovascular reconstruction techniques.

CASE REPORT

A 36-year-old man initially presented to an outside hospital with a 2-day history of severe left thigh and leg pain with swelling. His symptoms acutely worsened the day of presentation to the point that he was unable to ambulate without assistance. He was nonsmoker, led an active lifestyle, and denied any history of trauma, long distance travel, immobilization, or systemic symptoms, and had no personal or family history of hypercoagulability. His medical history was significant for an

appendectomy and radiofrequency ablation of the left great saphenous vein with stab phlebectomy 2 years prior at another hospital for "heaviness" and symptomatic painful varicosities. On venous duplex imaging, he was found to have acute thrombosis of the left femoral and common femoral veins. Because he was sensorimotor intact, he was initially managed with therapeutic heparin, resulting in some improvement, then transitioned to apixaban and was discharged home on hospital day 2. A full hypercoagulable workup was negative.

Several months later, despite anticoagulation and compliance with graduated compression stocking wear, he presented to our clinic with complaints of pain is in his entire left leg that was worse in the evenings and exacerbated by standing. He had extensive varicosities on his left anterior thigh and pelvis, popliteal fossa, and anterior and posterior calf, as well as isolated varicosities of the posterior right calf. Bilateral moderate stasis dermatitis was present and he had a chronic left medial malleolus venous stasis ulcer that had required an injection for bleeding, several weeks prior.

A computed tomography venogram (Fig 1, *A* and *C*) revealed a patent suprahepatic and intrahepatic vena cava but an absent infrahepatic vena cava and common iliac system. The bilateral external iliac veins and left common femoral vein were thrombosed and extensive bilateral retroperitoneal collaterals were found to be draining patent renal veins while dilated anterior abdominal wall and pelvic collaterals were draining into a prominent azygous system.

Because he failed best medical management with persistent active venous ulceration (CEAP 6), we elected to take the patient for a venogram and iliocaval reconstruction. Under general anesthesia, we accessed the right internal jugular vein and a wire and catheter was parked in the infrahepatic IVC. Diagnostic venography revealed a total occlusion of the IVC just distal to the hepatic vein with no distal reconstitution (Fig 1, *B*). We then accessed the patent bilateral femoral veins and a bilateral venogram was performed though short access sheaths revealing a flush occlusion of the common femoral veins with venous drainage through bilateral tortuous pelvic collaterals (Fig 1, *D*). Initially, endovenous crossing was pursued in the event that

Fig 1. Preoperative computed tomography angiography and intraoperative fluoroscopy. There is complete absence of the infrahepatic inferior vena cava (IVC) and common iliac veins. (**A** and **B**) The IVC terminates just inferior to the liver (*white*) with tortuous retroperitoneal venous collateral networks (*Grey*) draining the patent renal veins. A prominent azygous system is present (*black*). (**C** and **D**) Distally, there is complete absence of the common iliac veins with venous outflow reconstituting at the common femoral veins (*white*). Prominent pelvic collaterals are seen and account lower extremity venous return (*black*).

this case was actually atresia or early thrombosis. However, in the setting of absent caval remnants or scar on preoperative imaging or intraoperative venography and several unsuccessful attempts to endovenously cross, we further supported our original diagnosis of a true caval agenesis and proceeded with an extravascular reconstruction. We established a stable endoconstruct for treatment, as detailed in Fig 2, followed by creation of a potential space in the retroperitoneum, which was then lined with Wallstents (Boston Scientific; Marlborough, Mass) (Fig 3). In the event of extravasation into the retroperitoneum,

covered stents were available; however, they are not typically required in the setting of a low-flow venous system.

After an approximately 6-hour duration, we demonstrated on completion venography successful inline venous drainage from both legs into the right atrium (Fig 4, *A*). Postoperatively, the patient had 1-2 weeks of mild back discomfort managed with acetaminophen and nonsteroidal anti-inflammatory drugs. He was discharged home on postoperative day 2 with a plan for 1-year of clopidogrel and lifelong therapeutic anticoagulation on apixaban. At the 30-day follow-up, a computed tomography
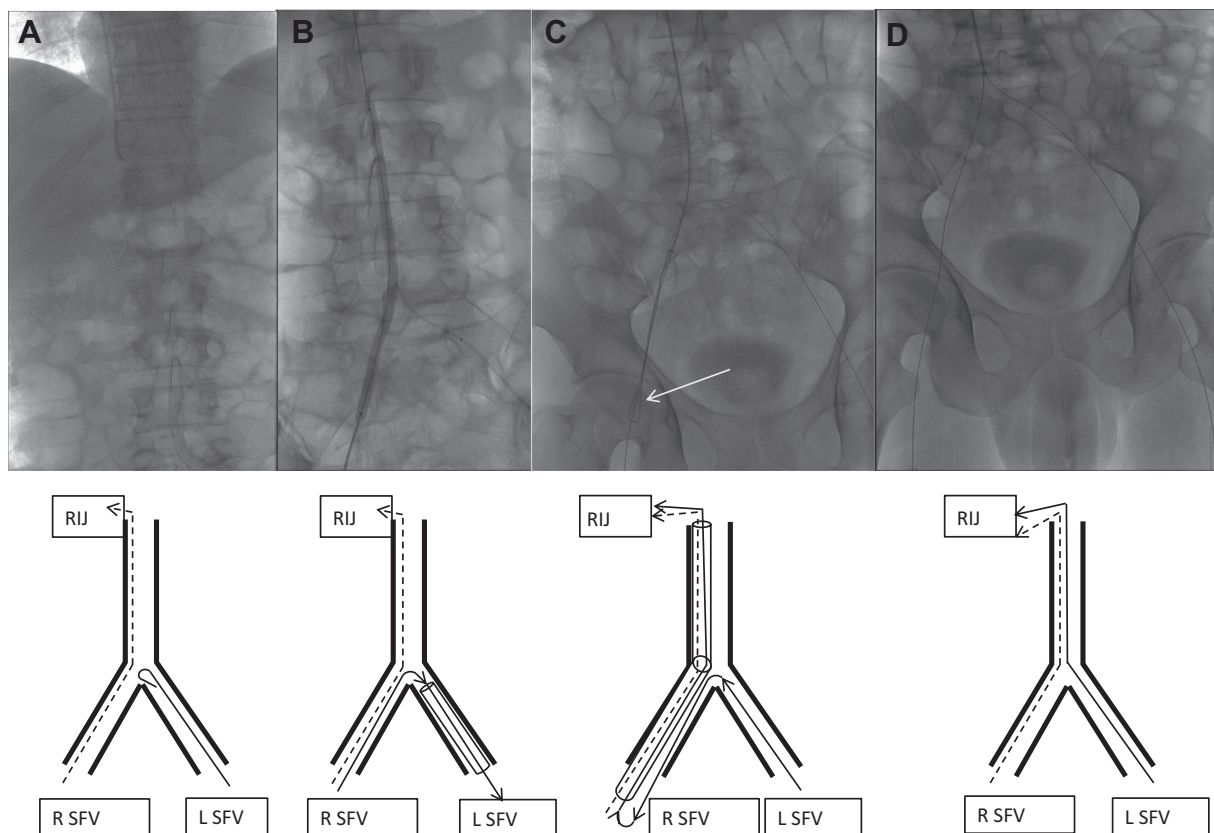
Fig 2. Overview of crossing technique. **A,** Using floppy and stiff angled Glidewires, Glidewire Advantage and a NaviCross Catheter (Terumo, Somerset, NJ), we were able to cross the occluded right (*R*) common femoral vein, external iliac vein and common iliac vein to enter the infrahepatic retroperitoneal space. During retroperitoneal crossing, care was taken not to push against resistance combined with multiple gantry angles to ensure the peritoneal space was not violated. After successfully crossing the retroperitoneum, that wire was then snared from the R internal jugular vein (*R IJV*) access and externalized to create a R femoral vein (*R FV*) to R IJV (R FV → R IJV) rail. We attempted the same procedure from the left (*L*) groin access, but were unable to enter a common retroperitoneal anatomic plane. **B,** A common anatomic plane was found at the approximate level of the inferior vena cava (IVC) bifurcation. Using a buddy-wire from the R FV, we were able to cannulate a catheter from the L FV access and externalize our wire to create a R FV to L FV (R FV → L FV) rail. **C,** A catheter spanning the R IJV → R FV rail was cannulated with the groin end of the RFV → L FV rail and fed through the body to exit the R IJV access. The catheter and R groin end of the R FV → L FV rail was then externalized (arrow marks looped wire as it is externalized). **D,** A parallel rail system from the bilateral FV to the RIJV was established. *SFV,* Superficial femoral vein.

venogram demonstrated patency of the IVC stent construct and decompression of all previously dilated venous collaterals, with unchanged renal drainage by the azygous system (Fig 4 and *C*) Clinically, his leg pain had resolved, with near complete resolution of his venous ulceration. At the 1-year follow-up he continues to have radiographic patency and reports complete resolution of his presenting symptoms and no further venous ulcerations. Further follow-up will be yearly by clinical symptoms only. For the content of this article, the patient agreed to allow the authors to publish their case details and images.

DISCUSSION

Congenital absence of the IVC is a rare venous malformation and may be responsible for 5% of all idiopathic deep vein thrombosis (DVT) in young (<30 years old) healthy adults with absent hypercoagulable risk factors.[2,3] In complete caval absence, extensive accessory venous drainage pathways result in IVC anomalies remaining clinically silent until the third to fourth decades of life, typically with acute DVT or varicosities.[2] Congenital absence of the IVC is also associated with atrophy of the right renal system (in rare cases the left) and has been implicated as cause of pelvic congestion syndrome.[4,5]

Further, well-defined collaterals may explain why many cases have historically been managed analogously to acute DVT with anticoagulation and compression.[1,4,6] Along those lines, several authors describe successful treatment of acute iliofemoral DVT in the setting of caval absence with catheter directed pharmacomechanical thrombolysis and thrombectomy with excellent 2- to 5-year outcomes.[1,7-9] Surgical management is rare, and
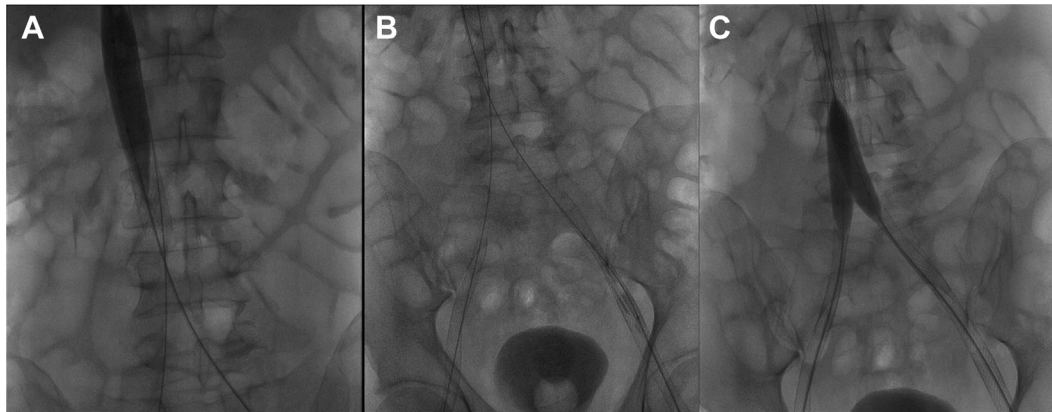
Fig 3. A, Serial angioplasty with double-barreled 5-, 8-, and 12-mm balloons was performed to create a potential space in the retroperitoneum. After confirming a patent channel with venous inflow and outflow, we began deploying 22-mm Wallstents to establish a new inferior vena cava (IVC) just distal to the hepatic vein. **B,** Wall stents were then placed in the bilateral iliac veins in a tapered fashion, 14 mm inferiorly and 16 mm superiorly. **C,** Two 18-mm double-barreled kissing stents were deployed into the previously placed IVC stents from the bilateral femoral accesses to complete the iliocaval reconstruction.
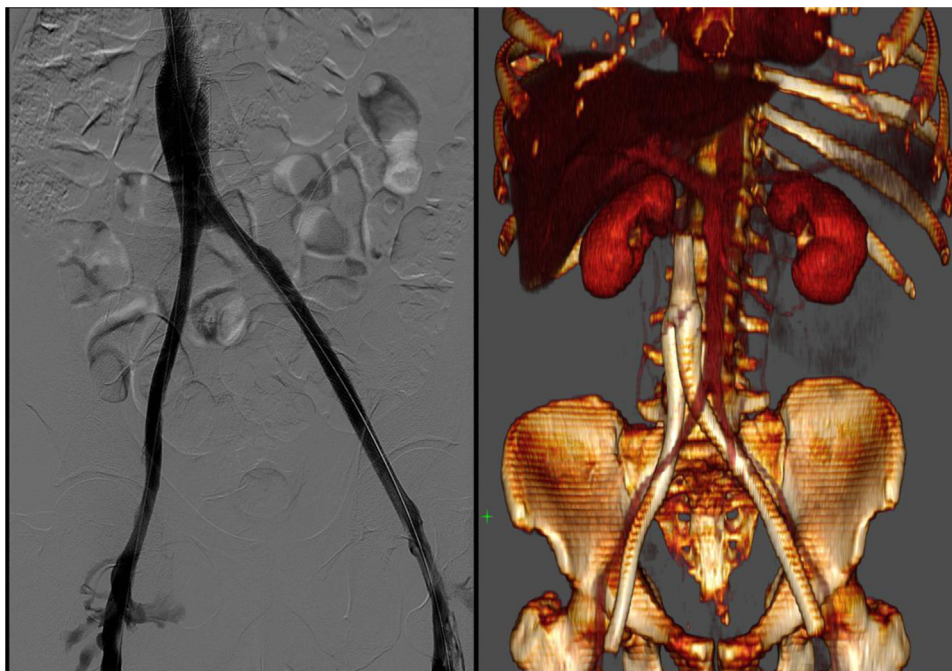

Fig 4. Completion intraoperative angiography and computed tomography angiography reconstruction post operatively. There is patent inferior vena cava (IVC) stent construct and decompression of the azygous system and venous collaterals seen on preoperative imaging.

only one case of open surgical management of caval absence by Dougherty et al[10] has been described recently.

Little is described in the literature regarding management of patients with caval agenesis who have failed best medical therapy or who have persistent venous ulceration. Beyond the single case of open bypass noted, management options are few with little data to support one option over another. When planning this case, we

looked toward the experience of others in the management of caval thrombosis, typically secondary to malignancy, hypercoagulability, and IVC filters. Chick et al[11] in a recent series of 120 patients with symptomatic iliocaval thrombosis due to IVC filters recanalized the IVC with endovascular stenting and achieved excellent 2-year primary (87.2%) and primary-assisted (90.3%) patency rates. This is consistent with others who have reported >80%-90% technical success rates and 2- to 5-year primary

and primary-assisted patency rates of approximately 85%-90%, and secondary patency rates of >85%.[12-16] Not unsurprisingly, iliocaval recanalization for symptomatic venous insufficiency was found to result in clinical improvement in 88.9% of patients and associated with decreased CEAP score.[12]

In conclusion, we have presented a somewhat novel case of extravascular iliocaval reconstruction for complete infrahepatic caval absence in the setting of a young man with venous ulceration who had failed best medical management. This case highlights the need for increased clinical awareness for an anatomic variant that is likely under detected in the general population and should be considered in the evaluation of any young patient who presents with proximal DVT, in the absence of known risk factors. To the best of our knowledge, this represents one of the first technically successful cases of complete minimally invasive reconstruction for congenitally absent IVC and expands on significant work from other institutions in recanalizing IVC thrombosis.

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