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CASE REPORT

Percutaneous Orthotopic IVC Construction in a Pediatric Patient with Symptomatic IVC Agenesis

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Abstract Inferior vena cava agenesis is an uncommon condition usually attributed to embryologic dysgenesis. When symptomatic, unprovoked deep venous thrombosis and/or lower extremity venous congestion are the most frequent manifestations. Its rarity has precluded consensus regarding appropriate management. Symptomatic chronic venous congestion requires surgical construction of auxiliary venous pathways, which may involve substantial morbidity, prolonged recovery and extensive scarring. We report successful minimally invasive management via percutaneous endovascular orthotopic inferior vena cava construction in a pediatric patient, thereby obviating the need for surgery and its associated morbidity.
Level of Evidence Level IV, case study.

Keywords IVCA (IVC agenesis) · Neocava · Interrupted IVC · Percutaneous IVC construction

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Introduction

Inferior vena cava agenesis (IVCA) occurs in 0.15–0.3% of the population, most commonly manifesting as unprovoked lower extremity deep venous thrombosis (DVT) or congestion [1, 2]. There is no consensus on management, with recommendations including compression stockings, chronic anticoagulation and/or surgery. We report a case of symptomatic suprarenal IVCA in a pediatric patient treated via percutaneous orthotopic construction of the absent segment. Follow-up for nearly 3 years has demonstrated continued patency and clinical improvement.

Case Report

A 14-year-old male was referred with 4 years of chronic nausea. Symptoms occurred daily and were most severe in the mornings and following strenuous exertion. The patient maintained adequate weight gain with regular ondansetron administration but had self-limited his activity and missed over 40 days of school in the past year. A review of systems was otherwise negative, and both physical examination and laboratory evaluation were unremarkable. Brain MRI, fluoroscopic upper gastrointestinal series and esophagogastroduodenoscopy were all unrevealing.

A contrast-enhanced abdominopelvic MRI was interpreted as demonstrating a complex right renal arteriovenous malformation; direct venography clarified the diagnosis to be absence of an 8-cm segment of suprarenal inferior vena cava (IVC) (Fig. 1). Collateral flow via retroperitoneal, ascending lumbar and paravertebral vessels resulted in the formation of large varices.

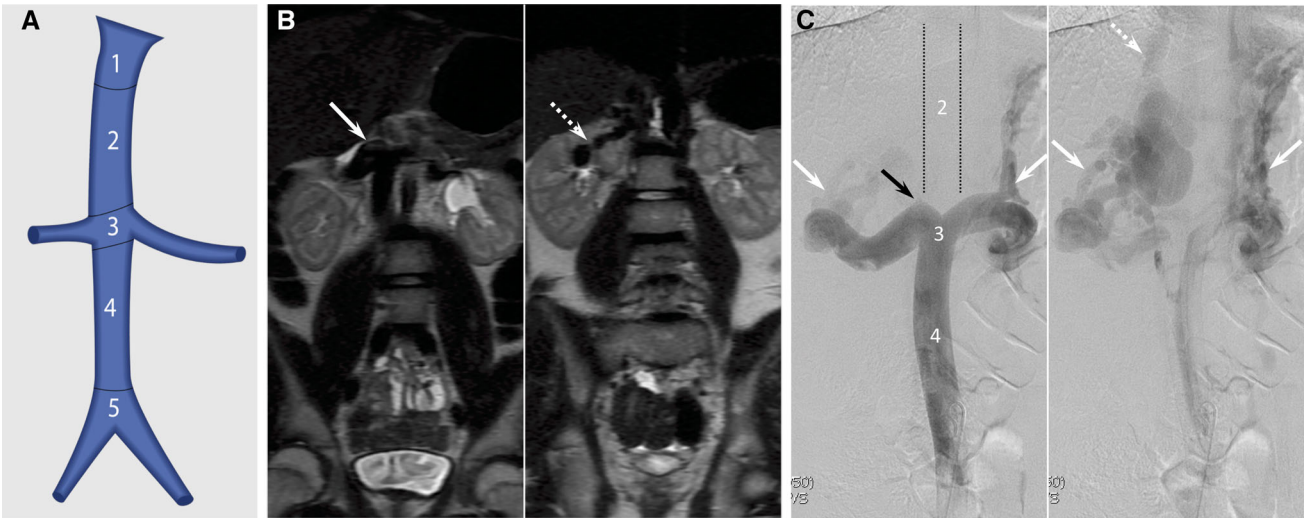

Fig. 1 **A** IVC segmental anatomy indicating the named adult segments and embryologic origins: *1*—hepatic IVC from the vitelline vein; *2*—suprarenal IVC from the right subcardinal vein; *3*—renal IVC/renal veins from the right supracardinal/subcardinal anastomosis; *4*—infrarenal IVC from the right supracardinal vein; *5*—common iliac veins from the posterior cardinal vein. **B** Coronal single-shot fast spin echo MR images demonstrating the absence of the suprarenal IVC (solid arrow) and prominent collateral vessels (dashed arrow). **C** Digital subtraction angiogram (DSA) images of the atretic IVC at earlier (left) and later (right) timepoints demonstrating the unremarkable renal (*3*) and infrarenal (*4*) segments, as well as complete absence of the suprarenal (*2*) segment (dotted lines indicate expected location). A small outpouching (black arrow) arising from the superior aspect of IVC/right renal vein junction represents a remnant of the atretic suprarenal IVC. Markedly enlarged and tortuous collateral vessels (solid arrows) show progressive opacification, some of which appear to communicate with the suprahepatic remnant (dashed arrow)

The patient was referred to a subspecialty center for surgery where he was informed that correction would require a left hepatectomy, placement of a prosthetic bypass graft and creation of temporary bilateral lower extremity arteriovenous fistulae, the latter of which would require surgical closure 3–9 months later. Given the substantial morbidity and prolonged recovery this would entail, the patient and his parents opted instead for percutaneous management.

The procedure was performed under general anesthesia by two board-certified interventional radiologists. The patient was positioned supine and access obtained via the right internal jugular (IJ) and right common femoral veins with 8-French and 11-French sheaths, respectively. A 6-French × 80 cm Berenstein occlusion balloon catheter (Boston Scientific, Natick, Massachusetts) was advanced via the right IJ sheath through the heart into the short segment of native suprahepatic IVC to act as a target during bridging puncture.

Through the femoral access site, a 0.035 Bentson wire and a 5-F Rubicon catheter (Boston Scientific) were advanced to the most cranial extent of the IVC and into the remnant outpouching. Applying gentle force to the wire caused it to buckle through the vessel wall into the retroperitoneal fat. Once extravascular, the relatively soft atraumatic wire was advanced under fluoroscopic guidance to the level of the suprahepatic remnant. The previously introduced right IJ balloon occlusion catheter was then inflated with contrast and a cone beam CT (CBCT) performed to confirm wire position (Fig. 2). The Rubicon catheter was then advanced over the wire into the retroperitoneum, the Bentson wire exchanged for a 0.035 Amplatz wire and this stiffer wire used to guide a sheath into the retroperitoneum. Once the wire, catheter and sheath were all positioned with tips adjacent to the suprahepatic remnant, the catheter was exchanged for a 16-gauge × 50.5 cm Colapinto needle (Cook Medical, Bloomington, Indiana).

Under fluoroscopic visualization, the needle was thrust through the wall of the suprahepatic remnant into the vessel lumen; contact between the needle and inflated balloon provided tactile and visual confirmation of position (Fig. 3A). A 0.035 Glidewire Advantage wire (Terumo Medical, Shibuya, Tokyo, Japan) was passed through the needle across the newly created communication and up into the right IJ vein, establishing "through and through" access. The needle was exchanged for a radiopaque marker catheter (Cook Medical), and the Glidewire for an Amplatz wire. The latter was used to deploy a 20 mm × 8 cm Wallstent (Boston Scientific), which was angioplastied first with a 12 mm × 80 mm Mustang balloon dilatation catheter (Boston Scientific) and then with a 20 mm × 60 mm Atlas Gold dilatation catheter (BARD Peripheral Vascular, Tempe, Arizona) (Fig. 3B). In its final position, the stent extended from the infrarenal IVC over the bilateral renal vein ostia and into the suprahepatic IVC remnant.

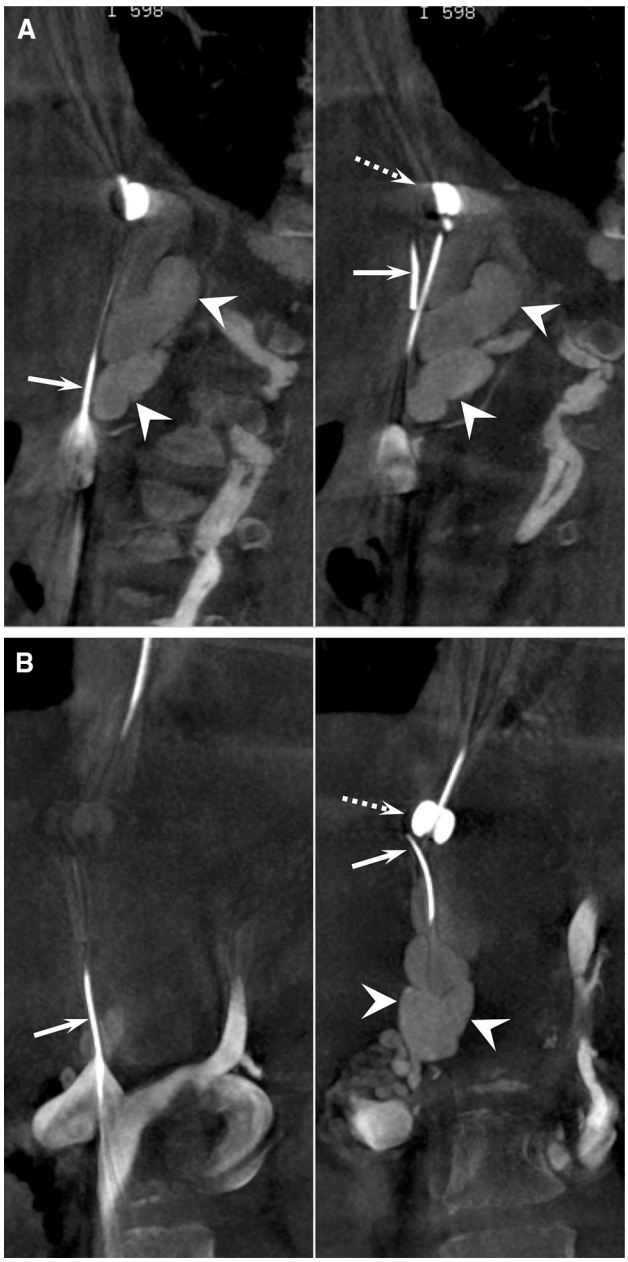

Fig. 2 Sagittal (**A**) and coronal (**B**) reconstruction CBCT images obtained prior to bridging puncture. The distal end of the wire (solid arrow) is seen doubled over immediate adjacent to the inflated balloon occlusion catheter (dashed arrow), and the relationship of the wire to the dilated retroperitoneal venous collaterals (arrowheads) is depicted

Imaging confirmed brisk anterograde flow through the stent with complete decompression of the varicose collaterals and preserved inflow from the renal veins. Sheaths were removed and hemostasis achieved via manual compression. The patient was discharged that evening on daily enteric-coated aspirin 81 mg with instructions to maintain adequate hydration and avoid contact sports; ondansetron 4 mg three times daily was also continued on an as-needed basis. He subsequently experienced symptomatic

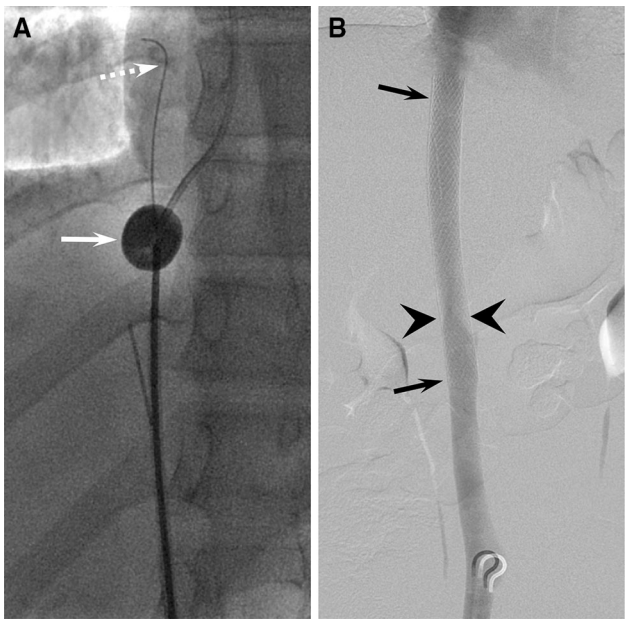

Fig. 3 **A** Fluoroscopic image obtained immediately after bridging puncture. The Glidewire Advantage wire is seen bypassing the contrast-opacified occlusion balloon (solid arrow) with wire tip in the right atrium (dashed arrow) en route to the right IJ vein. **B** DSA image demonstrating the final appearance of the reconstructed IVC. There was preferential anterograde flow through the stent (arrows) without extravasation or collateral opacification. Bilateral renal vein inflow is present (arrowheads)

improvement and returned to school 2 weeks post-procedure. Surveillance ultrasound and contrast-enhanced MRI obtained at 34 months post-procedure documented continued stent patency and variceal decompression (Fig. 4). He continues to experience intermittent bouts of nausea of reduced severity and frequency, but has demonstrated appropriate growth and development.

Discussion

Chronic nausea has not been reported in patients with IVCA, but the concordance between symptomatic exacerbation and augmentation of lower extremity venous return, as well as the patient's overall improvement following intervention, suggests that the anomaly was contributory. Though purely speculative, we postulate that gastric venous hypertension may have altered gastric hormonal secretion leading to feelings of nausea, and/or that the engorged retroperitoneal vessels directly irritated the celiac plexus and/or splanchnic nerves.That the IVCA was an entirely incidental finding must be acknowledged, although the authors note that the presence of markedly distended varices placed the patient at risk of acute life-threatening hemorrhage from relatively mild trauma, supporting the validity of intervention [3].

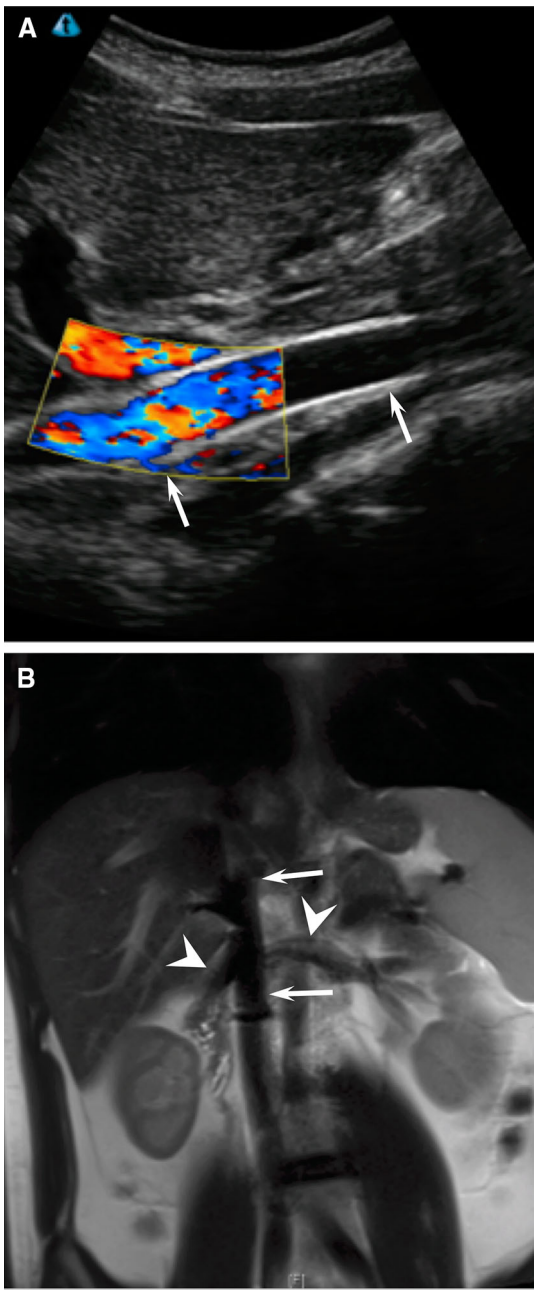

Fig. 4 **A** Sagittal Doppler US and **B** coronal T2-weighted MR images obtained at 34 weeks post-procedure demonstrate wide stent patency with brisk anterograde flow (arrows). Flow voids are also noted in both native renal veins (arrowheads)

Treatment of chronic venous congestion generally requires construction of auxiliary pathways of venous return. Surgical options include open placement of prosthetic bypass grafts accompanied by creation of temporary lower extremity arteriovenous fistulae to improve patency rates [4–6]. In the largest series on surgical intervention for IVCA consisting of 15 patients, Sagban et al. reported a secondary graft patency rate of 83% at 41 months [6]. A percutaneous endovascular approach mitigates the morbidity that accompanies surgery and has been employed for treatment of chronic IVC occlusion. Gilbert et al. described the creation of a non-orthotopic retroperitoneal pathway using covered stents in a patient with IVC thrombosis [7]. Haskal et al. reported the first creation of a "neocava" in a 29-year-old woman who developed iliocaval thrombosis and Budd–Chiari syndrome in the setting of IVCA with azygous continuation; expanded polytetrafluoroethylene (ePTFE)-covered stents were deployed across the 8-cm defect and the patient discharged on lifelong anticoagulation [8]. Cronan et al. described a 15-year-old patient with bilateral lower extremity DVT in the setting of infrarenal IVC agenesis who was managed via endovascular stenting of the collateral channels and catheter-directed thrombolysis [9]. More recently, Cooper et al. reported endovascular iliocaval reconstruction with stent grafts in a 65-year-old man who developed symptoms of lower extremity congestion after undergoing near-total IVC resection for malignant involvement [10].

In the current case, a bare metal stent (BMS) was utilized because (1) it was thought to be less thrombogenic than a covered graft, obviating the need for lifelong anticoagulation; (2) it was necessary to stent across the renal veins, as the only available landing zone was the renal/infrarenal IVC; and (3) BMS were available with diameters up to 24.0 mm, whereas the largest covered stent diameter was 14.5 mm. To achieve adequate pressure reduction, stent size should approximate that of the native adult IVC, which is 20 mm [11]. Consequently, only an uncovered stent provided sufficient flow. Near-complete occlusion of the stent wall interstices by retroperitoneal fat was sufficient to prevent extravasation, but had it occurred, occlusion balloons and covered stents were immediately available.

A particular challenge in the utilization of a BMS in this setting is the shortening which occurs during stent expansion, given that the stent was deployed in the retroperitoneum as opposed to within a preexisting channel. Before deployment, the stent was positioned such that the superior end was located in the right atrium; as it expanded, the superior end migrated caudally into the target location. Using this technique, no gap was left at either end of the stent.

In conclusion, we report a case of percutaneous orthotopic IVC construction in a pediatric patient with IVCA. Further studies are necessary to establish rates of technical and clinical success and of long-term patency.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Consent for Publication For this type of study, consent for publication is not required.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. For this type of study, formal consent is not required. Institutional Review Board (IRB) approval is not required for case reports at our institution.

Informed Consent For this type of study, informed consent is not required.

References

1. Gayer G, Luboshitz J, Hertz M, et al. Congenital anomalies of the inferior vena cava revealed on CT in patients with deep vein thrombosis. AJR Am J Roentgenol. 2003;180(3):729–32.
2. Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. Eur J Radiol. 2007;62(2):257–66.
3. Balzer KM, Pillny M, Luther B, Grabitz K, Sandmann W. Spontaneous rupture of collateral venous aneurysm in a patient with agenesis of the inferior vena cava: a case report. J Vasc Surg. 2002;36(5):1053–7.
4. Zhou W, Rosenberg W, Lumsden A, Li J. Successful surgical management of pelvic congestion and lower extremity swelling owing to absence of infrarenal inferior vena cava. Vascular. 2005;13(6):358–61.
5. Tofigh AM, Coscas R, Koskas F, Kieffer E. Surgical management of deep venous insufficiency caused by congenital absence of the infrarenal inferior vena cava. Vasc Endovasc Surg. 2008;42(1):58–61.
6. Sagban TA, Grotemeyer D, Balzer KM, et al. Surgical treatment for agenesis of the vena cava: a single-centre experience in 15 cases. Eur J Vasc Endovasc Surg. 2010;40(2):241–5.
7. Gilbert PJ, Sanders KW, Bourgeois AC, Semaan A, Guimaraes MS. Percutaneous iliocaval construction to treat symptomatic chronic lower extremity venous insufficiency. J Vasc Interv Radiol. 2016;27(8):1168–9.
8. 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(1):63–9.
9. Cronan JC, Gill AE, Gera A, Hawkins CM. Endovascular treatment of inferior vena cava agenesis using the azygos vein. J Vasc Interv Radiol. 2017;28(9):1270–1.
10. Cooper KJ, Chick JFB, Khaja MS, et al. Total endovascular iliocaval reconstruction using polytetrafluoroethylene stent–graft placement for the treatment of inferior vena cava resection. Cardiovasc Interv Radiol. 2018;41(7):1116–20.
11. Raju S. Treatment of iliac-caval outflow obstruction. Semin Vasc Surg. 2015;28(1):47–53.