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Treatment of Inferior Vena Cava Obstruction Following Pediatric Liver Transplantation: Novel Use of a Customized Endovascular Stent

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Liver transplantation is the treatment of choice for pediatric patients suffering with complications from end-stage liver disease. Although the perioperative management of patients undergoing liver transplantation has improved significantly, post-transplantation risks of graft failure, infection, bleeding, and bowel obstruction persist.[1] Technical variant allografts from living and deceased donors account for 50% of allografts transplanted in children aged ≤5 years and are associated with an increased risk of biliary and vascular complications.[2-4] Early recognition and rescue of allografts from perioperative complications are critical to realizing the best post-transplantation patient outcomes.[5] Hepatic venous outflow obstruction, although less common than portal vein or hepatic artery thrombosis, may adversely impact allograft health and function.

Anatomic obstruction of the inferior vena cava (IVC) and/or hepatic vein may be caused by vascular torsion or compression (early) or by fibrosis at the surgical anastomotic site (medium to late) after liver transplantation.[6-9] IVC obstruction leads to venous hypertension proximal to the lesion, which typically presents with a combination of signs and symptoms including ascites, lower extremity edema, hepatomegaly, pleural effusion, and graft dysfunction.[9] Compliant or dynamic lesions, such as vascular torsion or compression, may be associated with fluctuating symptoms owing to variability in the degree of vascular obstruction, potentially contributing to delayed or missed recognition. Treatment of post–liver transplantation IVC anastomotic complications may be pursued via surgical or transcatheter approaches. In this report, we discuss the impact of pediatric post–liver transplantation IVC stenosis, detail available treatment options, and describe a novel approach to transcatheter relief of IVC obstruction that precludes the risk of obstruction to hepatic venous egress.

Case Description

A 14-year-old girl with complications of portal hypertension due to cystic fibrosis–related end-stage liver disease underwent a deceased donor reduced–left lobe liver transplantation with typical bicaval and portal venous reconstruction. Following a routine early post-transplantation convalescence, she developed progressive abdominal ascites, and presented 2 months after initial hospital discharge with respiratory distress, elevated liver enzymes, and moderate ascites. She underwent paracentesis with drainage of 1200 mL of serosanguinous fluid. Analysis of liver biopsy specimens demonstrated mild acute cellular rejection. IVC venography performed in interventional radiology demonstrated a complex area of venous stenosis, felt to possibly involve the hepatic venous confluence. A pressure gradient was not assessed. Balloon venoplasty was performed, resulting in complete elimination of the lesion at low atmospheric pressure (1-2 atmospheres), suggesting a compliant nature of the stenosis (eg, vascular torsion). Over the ensuing 4 months, abdominal ascites recurred, and 2 additional paracentesis procedures were performed, with removal of 1500 mL of serous fluid at each procedure. Invasive catheter-based evaluation was reperformed at the third paracentesis procedure, which raised a concern for right atrial hypertension, prompting referral to the cardiac catheterization laboratory for complete assessment of right heart hemodynamics.

On presentation for the right heart catheterization, severe ascites was noted. Invasive evaluation was notable for normal right atrial and pulmonary artery pressure but confirmed the presence of IVC stenosis, with an 8-mmHg mean pressure gradient across the IVC obstruction and nonphasic pressure tracings in the IVC and hepatic veins (**Figure 1**, A). Venography demonstrated a complex vascular stenosis at the level of the IVC just proximal to its entrance to the right atrium, with involvement of the hepatic venous connection to the IVC (**Figure 2** and **Videos 1** and **2**; available at www.jpeds.com). The stenosis measured 3.5 mm at its narrowest point. There was evidence of severe contrast stasis in the distal IVC with collateralization from the infrahepatic IVC to the azygous venous system. Balloon venoplasty of the IVC lesion again supported the presence of a highly compliant vascular lesion, with elimination of the balloon waist at low pressure. The procedure was completed without further intervention.

A multidisciplinary discussion ensued, focusing on the potential approaches to relieving chronic IVC obstruction in the setting of a compliant vascular lesion and known caval and hepatic venous hypertension. Ultimately, the team elected to proceed with transcatheter relief of IVC stenosis in the cardiac catheterization laboratory, using a patient specific highly customized balloon-expandable vascular stent.

IVC Inferior vena cava

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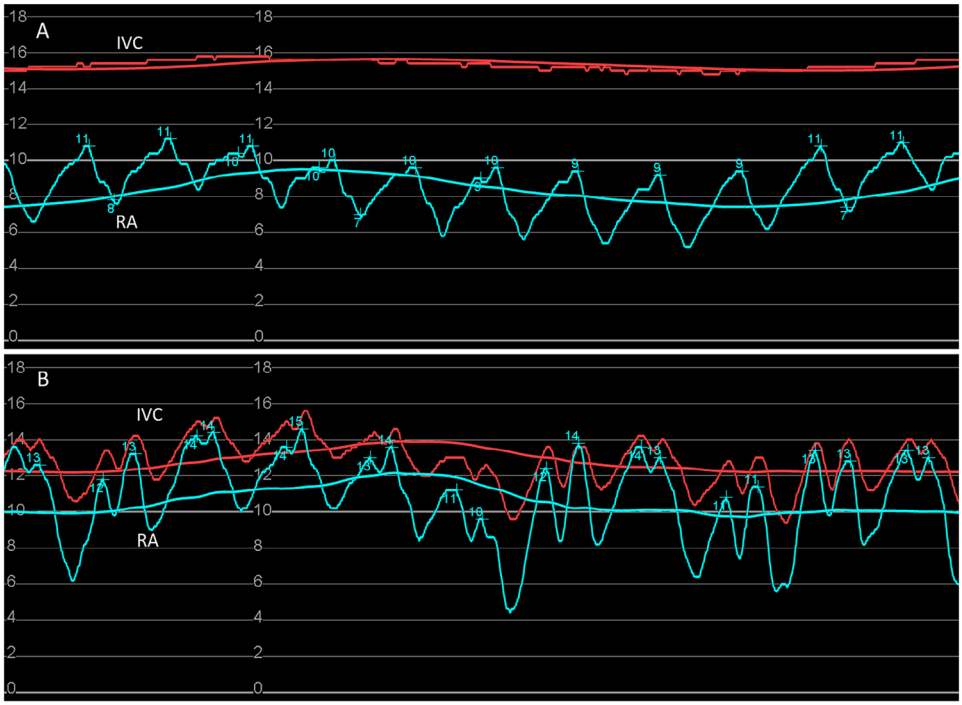

Figure 1. Venous wave forms acquired during right heart catheterization before and after IVC stent implantation. **A,** Before implantation, the IVC wave form is substantially elevated with a nonphasic flow pattern present, consistent with severe caval obstruction. **B,** After stent therapy, normal phasic flow is returned to the IVC tracing, with a trivial residual gradient to the right atrium (RA).

The patient returned to the cardiac catheterization laboratory, and femoral and internal jugular venous access was obtained. Following biplane venography in multiple angles, wire position across the lesion was obtained from the femoral venous approach. A 36-mm stent (Mega LD; ev3 Endovascular, Plymouth, Minnesota) was hand-crimped onto a 10-mm × 4-cm balloon (Z-MED II; NuMED, Hopkinton, New York) and advanced to the IVC lesion via a long sheath. The stent was implanted with complete balloon/stent waist resolution at low pressure (**Figure 3**, A and B). The stent was deployed across the entire IVC stenosis, intentionally spanning the hepatic venous entrance into the IVC, to fully and ideally relieve the long-segment IVC stenotic lesion.

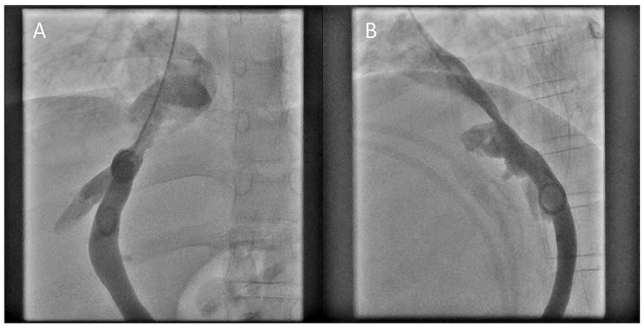

Figure 2. IVC venogram in **A,** frontal and **B,** lateral projections before transcatheter intervention. There is evidence of complex and long-segment stenosis of the IVC with involvement of the hepatic venous anastomosis.

We next proceeded to "customize" the stent in vivo, to facilitate unobstructed hepatic venous egress and match the stent to the patient's venous anatomy. The cranial portion of the stent extended past the stenotic area into a region of the IVC immediately proximal to the right atrium, where the cava was significantly dilated (poststenotic dilatation). In this region, the stent was flared with a 14-mm Z-MED II balloon (**Figure 3**, C and D). Then, to begin the process of eliminating the stent side struts that were "jailing" (or crossing) the hepatic venous egress to the IVC, we advanced a 0.014-inch coronary guide wire through a side hole in the open-cell stent and positioned in a distal hepatic vein. The stent struts (at the orifice of the hepatic vein entrance to the IVC) were then serially dilated from 2.5 mm to 14 mm in diameter, using a series of angioplasty balloons (**Figure 3**, E and F). In so doing, we created a customized "side hole" in the stent, thereby generating an entirely unobstructed hepatic venous egress into the stented IVC, without the presence of stent material jailing the hepatic venous confluence.

After the creation of a customized side hole in the stent, the IVC portion of the stent was redilated (because the side hole creation had crushed a portion of the IVC stent posteriorly) and then further dilated to 12 mm, generating a completely unobstructed IVC (**Figure 3**, G and H). The final customized stent is shown in **Figure 3**, I and J.