Transjugular Intrahepatic Portosystemic Shunt Creation in a Polycystic Liver Facilitated by Hybrid Cross-sectional/Angiographic Imaging

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Polycystic liver disease (PCLD) has long been considered to represent a contraindication to transjugular intrahepatic portosystemic shunt (TIPS) creation, primarily because of the risk of hemorrhage. Three-dimensional (3D) navigation within the enlarged and potentially disorienting parenchyma can now be performed during the procedure with the development of C-arm cone-beam computed tomography, which relies on the same equipment already used for angiography. Such a hybrid 3D reconstruction-enabled angiography system was used for safe image guidance of a TIPS procedure in a patient with PCLD. This technology has the potential to expedite any image-guided procedure that requires 3D navigation.

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Abbreviations: IVC = inferior vena cava, PCLD = polycystic liver disease, 3D = three-dimensional, TIPS = transjugular intrahepatic portosystemic shunt, 2D = two-dimensional

THE adoption of transjugular intrahepatic portosystemic shunt (TIPS) creation for treatment of complications of portal hypertension has been widespread. However, the patient population is heterogeneous, and anatomic and physiologic contraindications still remain (1). Although successful TIPS creation has been reported in three patients with hepatic fibrosis secondary to polycystic liver disease (PCLD) (2–4), the potential for technical failure, cyst rupture, and hemorrhage still causes PCLD to be considered at least a relative, if not absolute, contraindication to TIPS creation (1).

Several reports have described hybrid projectional and cross-sectional imaging methods to guide TIPS procedures, including the use of magnetic resonance (MR) imaging (5), three-dimensional (3D) ultrasound (US) (6), and intravascular US (7). All these methods require multiple distinct imaging instruments. The development of digital flat-panel x-ray detectors with high readout rates facilitates 3D tomographic reconstruction, resulting in a new class of 3D-enabled hybrid C-arm systems capable of producing conventional projectional angiographic images as well as computed tomography (CT)-like images with use of the same equipment. We used such a hybrid instrument to guide a TIPS procedure in a patient with a severely enlarged and distorted liver affected by PCLD and fibrosis.

CASE REPORT

Our institutional review board does not require approval for this type of retrospective case report.

A 62-year-old woman with an 18-year history of symptomatic polycystic kidney disease, PCLD, and hypertension presented with progressive abdominal distension, pain, and refractory hypertension. She was functionally anuric and had an operational Brescia-Cimino shunt. The patient underwent elective bilateral radical nephrectomies for relief of bulk symptoms and renovascular contribution to hypertension. Although her immediate postoperative course was uncomplicated, she experienced severe bilateral lower extremity and lower truncal pitting edema as well as massive ascites within 3 months.

MR imaging revealed near-occlusion of the inferior vena cava (IVC) and marked distortion of the hepatic veins caused by deforming cysts (Fig 1). Hepatic venography and IVC venography were performed, confirming these findings and revealing gra-
Gradient of 8 mm Hg between the hepatic vein and the right atrium and 7 mm Hg between the infrahepatic IVC and the right atrium. Self-expanding nitinol stents were deployed in the IVC stenosis and the middle hepatic vein, which was the only hepatic vein that could be recognized and cannulated. Within approximately 3 weeks, during which there were nine hemodialysis sessions, the peripheral edema completely resolved. However, massive ascites persisted, requiring therapeutic large-volume paracentesis of 8–9 L every 3 weeks. The ascites was presumed to be a result of progressive hepatic fibrosis with a possible component of small-vessel hepatic venous obstruction, but histopathologic studies were unavailable because the risk of hemorrhage from biopsy was considered prohibitive. On the basis of a consensus among a transplant surgeon, a nephrologist, an interventional radiologist, the patient, and the patient’s family, we performed portal decompression via creation of a TIPS. The patient’s Model for End-stage Liver Disease score was 20 as a result of renal failure, and the Child-Pugh-Turcotte score was 8.

The patient was administered general anesthesia for maximum control of respiration and motion. Projectional images were acquired with use of a flat-panel C-arm angiography instrument (Axiom Artis dBA; Siemens Medical Solutions, Forchheim, Germany). Cross-sectional images were reconstructed on a workstation (X-

Figure 1. Transverse axial T1-weighted MR images of the liver. (a) At the level of the confluence of hepatic veins, the IVC appears as a slit (arrow), and the only recognizable hepatic vein is a severely distorted middle vein (white arrowhead). The azygos system (black arrowhead) is hypertrophied, likely as a result of IVC obstruction. (b) The main portal vein is patent (arrow) but surrounded by cysts of different signal intensities, representing fluids with different concentrations of protein and blood. Intrahepatic collateral hepatic venous vessels (arrowheads) likely result from long-standing venous outflow obstruction.

Figure 2. A C-arm axial transverse CT image through the target main right portal vein (arrowheads) shows pericystic and aortic calcifications used as landmarks. This cross-sectional image was reconstructed with use of DynaCT software. The associated two-dimensional (2D) x-ray input projections were acquired with an Axiom Artis dBA flat-panel detector C-arm system.
Leonardo; Siemens) with use of 3D reconstruction software (DynaCT; Siemens). No intravenous contrast agent was administered. The cross-sectional images depicted the long and cyst-laden path between the hepatic vein and portal vein (Fig 2). Anatomic landmarks, including individual cysts and calcifications, were identified from CT and MR images previously obtained with use of contrast medium enhancement, and the relative position of the target right portal vein was established in relation to these landmarks.

Access was obtained into the preexisting middle hepatic vein stent with a Hawkins TIPS set (Angiodynamics, Queensbury, NY). The distal 2 cm of the 22-gauge needle was custom-shaped with a gentle 15° curve to allow fine steering. An initial pass was performed under fluoroscopic guidance, and 3D image reconstruction was repeated (Fig 3). On the basis of the 3D data obtained, the needle was then advanced more inferiorly, anteriorly, and medially, directly into the right main portal vein. The tract was dilated to 8 mm in diameter and lined with overlapping covered stents 10 cm and 9 cm long and 8 mm in diameter (Viatorr; W.L. Gore & Associates, Flagstaff, AZ). Digital subtraction angiography and 3D image reconstruction with and without contrast agent injection were performed, confirming successful creation of the shunt (Figs 4, 5) and revealing a small amount of intracystic hemorrhage without active extravasation. The portosystemic gradient measured between the splenic vein and right atrium was reduced from 13 mm Hg to 4 mm Hg.

Postoperatively, the patient showed stable vital signs and tolerated routine hemodialysis. Her hematocrit level increased 2% compared with measurements before the TIPS procedure. She was treated for spontaneous bacterial peritonitis in the first postoperative week, required one therapeutic paracentesis of 3 L in the first postoperative month, and requires low-dose lactulose administration for prevention of encephalopathy, but is now free of ascites at 6-month follow-up.

**DISCUSSION**

TIPS creation has essentially completely replaced surgical portosystemic shunt creation for treatment of complications of portal hypertension. The original consensus document published by the National Digestive Diseases Advisory Board in 1995 (8) outlined indications and contraindications, but many guidelines were based on skepticism and theoretical risks without scientific evidence. The updated guidelines from the American Association for the Study of Liver Diseases (1) show that some of the clinical contraindications continue to be accepted (eg, heart failure, severe pulmonary hypertension), but technical and anatomic contraindications such as Caroli disease, portal vein thrombosis, and PCLD need to be reconsidered, especially since the advent of covered stents. TIPS creation in the patient with polycystic kidney disease without hepatic cysts is technically routine (9), but the safe decompression of portal hypertension in patients with hepatic cysts depends largely on the

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**Figure 3.** Repeat cross-sectional reconstructed images after the first advancement of TIPS needle. (a) Axial transverse image at the same level as Figure 2 shows the tip of the needle lying approximately 2 cm posterolateral to the target right portal vein (arrow). (b) Coronal reconstruction along the needle path shows the guiding cannula within the preexisting middle hepatic vein stent and the 22-gauge needle traversing several large cysts toward the landmark calcifications. The needle was adjusted from this position to puncture the target.
ability to locate the portal vein amid the array of cysts (2–4).

The development of ascites after nephrectomy in patients with polycystic kidney disease has been reported in three patients and is thought to be caused by hepatic venous outflow obstruction from torsion and distortion of the hepatic veins and IVC from the repositioning of the liver (10,11). Patients may also experience hepatic fibrosis with or without cystic involvement of the liver, which can eventually lead to ascites. Stent implantation of the IVC in our patient relieved lower-extremity edema, but stent implantation of the dominant hepatic vein was ineffective in the treatment of the ascites, suggesting that macroscopic obstruction of venous outflow was not the sole cause of ascites. The relatively low portosystemic gradient measured on portal vein puncture (13 mm Hg) and the minimal varices observed raised further questions about the actual cause of the ascites, but we proceeded with the shunt procedure because predicting efficacy on the basis of the gradient before TIPS creation is difficult (12), and TIPS creation is potentially reversible. The patient’s positive clinical response to TIPS creation suggests that portal hypertension played a role.

Three-dimensional image reconstruction with use of a C-arm system generates an isotropic 3D data set from 2D x-ray input projections. For data acquisition, we used the 30-cm × 40-cm detector of the Axiom Artis dBA biplane system. Acquisition protocols that require a maximum of 20 seconds result in as many as 538 projections through a partial rotation of at least 200°. For optimal detector performance, this system relies on a dose-control system whereby the detector entrance dose is held constant by adjustment of the current time product of the x-ray tube and regulation of the tube voltage only if needed. In our case, 20-second runs were performed with the x-ray source voltage at 125 kVp and the detector entrance dose set at 1.2 μGy per projection.

The 2D x-ray projections are subsequently fed into a 3D tomographic image reconstruction algorithm modified to account for irregular but stable scan trajectories (13). To optimize 3D soft-tissue image quality, a sequence of correction algorithms is applied, including scatter correction, beam-hardening correction, truncation correction, and ring-artifact correction (14). The enhanced 3D reconstruction algorithm, marketed as DynaCT, also provides bone and vessel kernels for 3D reconstruction. Bone kernels are designed for tomographic reconstruction of soft-tissue details, whereas vessel kernels are optimized for contrast medium–enhanced vasculature. Soft-tissue structures such as cysts inside a liver are best depicted by bone kernels to avoid the inherent edge enhancement of vessel kernels. We reconstructed 3D data sets with an isotropic voxel size of approximately 0.8 mm. Voxels in neighboring slices may be averaged to achieve a larger effective slice width and thus a reduction in noise.

![Figure 4. Digital subtraction angiography performed on the same C-arm device used to acquire the 2D images used for 3D C-arm CT reconstruction. (a) Initial portal venogram shows sluggish antegrade flow into the distorted intrahepatic portal system and some retrograde flow into mesenteric vessels. Note that the length of the tract from the portal vein wall to the IVC measures more than 13 cm, and the needle throw from the hepatic vein wall to the portal vein wall measures more than 9 cm. (b) Portal venogram after placement of overlapping stents 10 cm and 9 cm long and 8 mm in diameter shows some preservation of intrahepatic portal flow and no extravasation. The portosystemic gradient is reduced from 13 mm Hg to 4 mm Hg.](image)
The use of a hybrid 2D/3D imaging instrument capable of 2D angiographic projection imaging and 3D reconstruction facilitated our long trans-cystic TIPS creation procedure and minimized blood loss. Considerable extra planning was required before this challenging procedure was attempted, and successful use of this new technology will likely remain very operator dependent. With use of accurate imaging and impermeable covered stents, TIPS creation in the patient with PCLD appears to be safe and effective.

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References

Figure 5. Repeat transverse axial C-arm CT image obtained after completion of the TIPS procedure several centimeters more cephalad than in Figures 2 and 3a. A small amount of intracystic hemorrhage is observed (arrowheads) involving a traversed cyst, which probably occurred during tract dilation. Delayed images (not shown) reveal no change, indicating no active extravasation. The stent anterior to the vertebral body is within the IVC.