Transjugular Intrahepatic Portosystemic Shunt in a Child with Budd-Chiari Syndrome: Technical Modification and Extended Follow-Up

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Abstract
An 8-year-old girl with Budd-Chiari syndrome presented with upper gastrointestinal hemorrhage and ascites. TIPS to decompress the portal hypertension was performed by puncturing the portal vein directly from the inferior vena cava. The shunt remains patent after 3 years without requiring secondary intervention.

Key words: Transjugular intrahepatic portosystemic shunt—Portal hypertension—Budd-Chiari syndrome

Transjugular intrahepatic portosystemic shunt (TIPS) is a side-to-side shunt that connects an intrahepatic branch of the portal vein (PV) to an hepatic vein with the aid of a metallic stent. TIPS has become an important alternative to surgically created shunts in the management of acute and chronic variceal hemorrhage in several series in adults [1]. Its use in children has been limited [2–4], due partly to concerns about technical feasibility [5].

We present a technical modification of TIPS consisting of connecting the inferior vena cava (IVC) directly to the PV instead of an hepatic vein. As far as we know, this is the first report in the literature using this technique in a child with Budd-Chiari syndrome.

Case Report
An 8-year-old girl, weighing 31 kg, presented with huge refractory ascites, severe gastrointestinal hemorrhage and severe impairment of her nutritional status. A percutaneous liver biopsy suggested Budd-Chiari syndrome. She was placed on the waiting list for liver transplantation. Her blood chemistry included serum albumin level of 4.1 g/dl (normal 3.5–5.0 g/dl); mean prothrombin time of 14.5 sec (control value 11.4–17.2 sec), platelet count of $130 \times 10^3$ (normal 200–400 $10^3$) and total serum bilirubin level of 5.2 mg/dl (normal 0.1–1.2 g/dl). Upper gastrointestinal endoscopy showed large esophageal varices with signs of recent bleeding. TIPS was indicated as a bridge to liver transplantation in order to control the variceal bleeding.

After informed consent was obtained the child was sent to the interventional radiology suite. The procedure was performed under general anesthesia. Superior mesenteric angiography was performed through the right femoral artery approach to identify the vascular anatomy and confirm patency of the PV. The right internal jugular vein was entered with a 5 Fr coaxial micropuncture introducer set (Cook, Bloomington, IN, USA). Inferior vena cavaography showed no reflux into hepatic veins and the retrohepatic portion of the inferior vena cava (IVC) was compressed by the caudate lobe (Fig. 1). No hepatic vein could be entered selectively (Fig. 2A), therefore, the PV was directly punctured from the IVC with a Rösch-Uchida System (Cook) (Fig. 2B) using arterial portography as a road map. An Amplatz torque guidewire (Medi-tech/Boston Scientific, Watertown, MA, USA) was introduced into the PV and advanced into the superior mesenteric vein. The portosystemic pressure gradient was 22 mmHg. The hepatic parenchymal tract was predilated with a 6-mm-diameter balloon, and an $8 \times 60$-mm Wallstent (Schneider, Minnetonka, MN, USA) was placed intrahepatically, connecting the PV to the IVC. The stent was dilated with an 8-mm balloon and portography was performed (Fig. 3). After stent placement, the portosystemic pressure gradient dropped to 5 mmHg. The procedure took about 5 hours. The child did well after the procedure with no complications.
Standard liver function tests normalized. Doppler ultrasound was performed to assess the patency of the shunt. The child was discharged 5 days after the procedure.

Doppler ultrasound assessment was performed every 3 months, showing patency of the shunt. Two follow-up angiograms did not reveal the need for any revision (Fig. 4). The child also received oral anticoagulation for 6 months after TIPS creation.

Three years after the procedure the child is doing well, liver function tests are normal, there is no encephalopathy and ascites disappeared. The child has normal growth with significant improvement in her nutritional status. She has not received a transplant yet.

Discussion

The main causes of portal hypertension in children are biliary atresia, congenital hepatic fibrosis, alpha 1-antitripsine deficiency, Budd-Chiari syndrome and portal vein thrombosis [6].

The use of TIPS in adults has been reported by many authors [7–9], but experience in children is scarce and there are only a few reports on the efficacy of TIPS in children [2–5]. A limiting factor to the widespread use of the procedure in children is the lack of adequate pediatric equipment. In the present case, the same instruments used for adults were employed without encountering technical difficulties. Hackworth et al. [4] reported technical difficulties when using the pediatric TIPS set (Cook), that was subsequently exchanged for a standard 5.2 Fr needle catheter (Rösch-Uchida System; Cook).

TIPS is an efficient procedure to control hemorrhage from gastric and/or esophageal varices as well as from hypertensive gastropathy [10–12]. TIPS has been used extensively as a bridge to liver transplantation [13, 14].

In children with Budd-Chiari syndrome, some technical details are very important when performing a TIPS. Substantial hypertrophy of the caudate lobe obscures the location of the right hepatic vein ostium. Before performing the trans caval puncture, it is necessary to stabilize the needle introducer where the IVC is surrounded by the caudate lobe, so that the needle tract to the portal vein branch remains intrahepatic, decreasing the risk of intraperi-