Transjugular Intrahepatic Portosystemic Shunt Performed in a 2-Year-Old Infant With Uncontrollable Intestinal Bleeding

By Takuya Kimura, Toshimichi Hasegawa, Takaharu Oue, Takaya Hoashi, Takamichi Murakami, Kenji Nakamura, and Akira Okada

Osaka, Japan

Although transjugular intrahepatic portosystemic shunt (TIPS) is widely accepted in adults, there have been few successful reports in infants. The authors describe a 2-year-old boy with postoperative biliary atresia who underwent TIPS for uncontrollable lower intestinal bleeding and achieved hemostasis. Massive melena developed, which was not controlled by conservative therapy and devascularization. A shunt was created between the right hepatic vein and the right portal vein with a 6-× 50-mm Wallstent. After TIPS, the porto-systemic pressure gradient decreased from 15 to 11 mm Hg, and collaterals markedly reduced. He has not had recurrent bleeding for over 7 months. TIPS may be a technically feasible and effective treatment to control intestinal bleeding in infants. However, further experience and long-term follow-up will be required.


INDEX WORDS: Transjugular intrahepatic portosystemic shunt, intestinal bleeding, variceal bleeding, portal hypertension, biliary atresia, infant.

Bleeding from the lower digestive tract associated with portal hypertension, such as intestinal, mesenteric, or stomal varices is difficult to diagnose and treat. Endoscopic treatment usually is difficult, and surgical procedures are not always effective. However, a newly developed transjugular intrahepatic portosystemic shunt (TIPS) is a less invasive procedure and is rapidly gaining acceptance in the adult population. In the pediatric age group, however, only few attempts at TIPS have been made, because the procedure is rather laborious, and suitable size of the stent has not yet been determined. Successful reports, especially in infants below 3 years of age, are very few. We describe a 2-year-old boy who underwent TIPS for uncontrollable lower intestinal variceal bleeding and achieved hemostasis.

CASE REPORT

A 2-year-old boy suddenly had massive melena and shock with a blood hemoglobin level of 6.0 g/dL. He had been doing well without jaundice after hepatic portojejunostomy for biliary atresia. Despite the aggressive conservative therapy including blood transfusion, the use of β-blocker, and octreotide, he continued to bleed. Because the portography showed possible bleeding from mesenteric varicies of the Roux-en-Y loop, he underwent devascularization. Initial hemostasis was achieved, but melena recurred when oral feeding was increased. TIPS was considered in an attempt to reduce the portal pressure.

The procedure was performed under general anesthesia. A Ring Pediatric Transjugular Intrahepatic Access Set (Cook, Bloomington, IN) was used. Initially, the hepatic artery was catheterized with a guide wire by a Seldinger technique. Because the portal vein runs parallel to the hepatic artery, insertion of an 18-gauge transjugular needle through the hepatic vein into the portal vein was attempted by following the guide wire in the hepatic artery under fluoroscopy. After several trials, the right posterior branch of the portal vein was penetrated by the needle guide (Fig 1A). The tract was dilated with a 6-mm-diameter angioplasty balloon and stented with a 50-mm Wallstent (Fig 1B).

RESULTS

After TIPS, portosystemic pressure gradient decreased from 15 to 11 mm Hg. Portography performed after the procedure showed that the collaterals were markedly reduced. The stent was thrombosed on the second day. This was detected by color Doppler examination. It was recanialized successfully by percutaneous transjugular thrombectomy. This time, the tract was dilated to 8 mm, with the porto-systemic pressure gradient of 11 mm Hg. After the procedure, oral feeding was increased gradually and total parenteral nutrition (TPN) was weaned on the 10th day after TIPS. Liver dysfunction or encephalopathy has not been observed except 1 episode of a transient elevation of serum ammonia and total bilirubin levels. Six months after TIPS, the patient gradually presented with venous dilatation of the abdominal wall, although he has not had any more bleeding. Because the stent was thrombosed again, it was recanialized by the same procedure. Since this episode the patency of the stent has been confirmed by color Doppler examination for this 1 month.
DISCUSSION

Although TIPS has been used widely for the treatment of portal hypertension in the adult age group, this procedure is rarely attempted in children.3-8 Problems that hamper trial of TIPS in small children are difficulty in obtaining a device of the adequate size and technical difficulty because of the small size of the portal vein. There should be some technical differences between pediatric patients and adults to perform TIPS. Pediatric groups need general anesthesia, custom-ordered device, shorter size of metallic stent, and technical modification to access the portal vein. Table 1 summarizes 5 infants, including the current case, below the age of 3 years reported previously.4,6-8 Primary diseases were biliary atresia in 3, cystic fibrosis in 1, and TPN-related cirrhosis in 1 case. All patients underwent TIPS under general anesthesia. The reported stent size was 5 to 8 mm in diameter and 20 to 50 mm in length in these cases. In the current case, we used custom-ordered device. Guidance of the portal vein at insertion was made with the carbon dioxide or the needle placed transcutaneously into the portal vein in case 1, 2, and 4, respectively. In the current case, we placed a guide wire in the hepatic artery using the Seldinger technique to identify the portal vein. This technique was done safely and may be useful to access the narrow portal vein especially in the small children. In adults, the aim of this procedure is to reduce the portosystemic pressure gradient to less than 12 mm Hg.5 In these reported cases with the age below 3 years, the gradient after TIPS ranged from 5 to 11 mm Hg.

TIPS may be a technically feasible and effective treatment to control intestinal variceal bleeding associated with portal hypertension even in an infant. However, further experience and long-term follow-up will be required.

Fig 1. (A) Portography after transjugular needle just entered the portal vein showing the numerous collaterals to the intestine. (B) Portography after the creation of the TIPS shunt shows hepatopetal flow with markedly reduction of the collaterals. (Closed triangle, portal vein and stent; open triangle, intestinal varicies.)
Table 1. Cases of TIPS in Patients Under 3 Years of Age

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Body Weight (kg)</th>
<th>Cause of Liver Disease</th>
<th>Indication (Site of Varix)</th>
<th>Stent Diameter × Length (mm)</th>
<th>Porto-Systemic Pressure Gradient (mmHg)</th>
<th>Before TIPS</th>
<th>After TIPS</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&lt;sup&gt;e&lt;/sup&gt;</td>
<td>10 mo</td>
<td>9</td>
<td>BA</td>
<td>Variceal bleeding (gastroesophagus)</td>
<td>5 × 40</td>
<td>17</td>
<td>5</td>
<td></td>
<td>Infection, death</td>
</tr>
<tr>
<td>2&lt;sup&gt;e&lt;/sup&gt;</td>
<td>15 mo</td>
<td>9.7</td>
<td>CF</td>
<td>Variceal bleeding (intestine)</td>
<td>8 × 24</td>
<td>26</td>
<td>8</td>
<td></td>
<td>Liver failure, death</td>
</tr>
<tr>
<td>3&lt;sup&gt;e&lt;/sup&gt;</td>
<td>2.5 yr</td>
<td>13</td>
<td>TPN-related cirrhosis</td>
<td>Variceal bleeding (stoma)</td>
<td>8 × 40</td>
<td>12</td>
<td>5</td>
<td></td>
<td>Alive, no bleeding</td>
</tr>
<tr>
<td>4&lt;sup&gt;e&lt;/sup&gt;</td>
<td>2.5 yr</td>
<td>13.9</td>
<td>BA</td>
<td>Variceal bleeding (undescribed)</td>
<td>5 × 20</td>
<td>24</td>
<td>11</td>
<td></td>
<td>Alive, no bleeding waiting for LTx</td>
</tr>
<tr>
<td>5&lt;sup&gt;e&lt;/sup&gt;</td>
<td>2 yr</td>
<td>12</td>
<td>BA</td>
<td>Variceal bleeding (intestine)</td>
<td>6(8) × 50</td>
<td>15</td>
<td>11</td>
<td></td>
<td>Alive, no bleeding</td>
</tr>
</tbody>
</table>

Abbreviations: BA, biliary atresia; CF, cystic fibrosis.
* Current case.

REFERENCES