

Transjugular Intrahepatic Portosystemic Shunt in Children with Cirrhosis of Unknown Origin: A Case Report and Literature Review

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1. Abstract

1.1. Objective: To investigate the safety and efficacy of transjugular intrahepatic portosystemic shunt (TIPS) in children with cirrhotic portal hypertension.

1.2. Methods: We retrospectively analyzed the clinical data of a child with cirrhosis and portal hypertension, and reviewed the literature.

1.3. Results: A 12-year-old boy with a height of 167 cm and a weight of 60 kg was admitted to hospital for hematemesis for 2 days, and his physical examination showed liver cirrhosis. Multiple visits to an external hospital in the past 6 months, with bone marrow and liver puncture undertaken for exclusion of leukemia and other lesions, suggested cirrhosis of unknown origin. Hematemesis and melena occurred many times in the past 6 months, bleeding stopped after medical treatment and he was admitted to the hospital for hematemesis 2 days ago. TIPS was performed after medical treatment, and the patient recovered and was discharged from hospital.

1.4. Conclusion: TIPS is safe and effective in children with liver cirrhosis and gastrointestinal hemorrhage.

2. Introduction

The common causes of portal hypertension in children are different from those in adults [1]. Although the etiology of liver disease is different in most children and adults, the manifestations of portal hypertension are similar, including variceal bleeding, ascites and hepatic encephalopathy [2]. At present, the treatment of portal hypertension in children is the same as that in adults. Comprehensive treatment includes internal medicine, endoscopy and endovascular treatment. Liver transplantation is often the ultimate goal of treatment of primary liver disease [3]. Transjugular Intrahepatic Portosystemic Shunt (TIPS) can serve as a bridge to transplantation in the management of portal hypertension complications, but extensive data on the feasibility and effectiveness of TIPS in children and adolescents are lacking. There are some single-center retrospective reports in foreign literature [4]. There are few reports in China of TIPS in children with cirrhosis and portal hypertension. We here report a case of cirrhotic portal hypertension in a child treated with TIPS, and review the literature to evaluate the technical success rate, patency rate, safety and efficacy of TIPS in pediatric patients.

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3. Case Presentation

3.1. Chief complaints and history of disease

A 12-year-old boy, 167 cm in height and 60 kg in weight, was admitted for hematemesis. Six months ago, the patient was scheduled to undergo circumcision, and routine preoperative examination revealed a decrease in the three lineages of blood cells, and further physical examination suggested cirrhosis complicated with splenomegaly and hypersplenism. In the past 6 months, several visits to another hospital, with bone marrow and liver puncture, ruled out leukemia and other lesions, suggesting cirrhosis of unknown origin. Hematemesis and melena occurred many times in the past 6 months, bleeding stopped after medical treatment and he was admitted to the hospital for hematemesis 2 days ago. After

hemostasis and rehydration, blood transfusion and drug therapy, hematemesis stopped and vital signs stabilized.

3.2. Laboratory tests

Hemoglobin: 49 g/L (male: 120–160 g/L), red blood cells: 1.52×10^{12} (child: 4.2×10^{12} – 5.2×10^{12} /L), white blood cell: 1.52×10^9 (4.0×10^9 – 10.0×10^9 /L), platelets: 41×10^9 (100×10^9 – 300×10^9 /L); prothrombin time (PT): 13.20 s; activated partial thromboplastin time (APTT): 26.2 s; serum albumin: 28.7 g/L (40–55 g/L), total bilirubin: 12.9 $\mu\text{mol/L}$ (0–55 g/L; 23 $\mu\text{mol/L}$), alanine aminotransferase: 15 U/L (7–40 U/L), aspartate aminotransferase: 22 U/L (13–35U/L).

3.3. Imaging examinations

Gastroscopy showed four thick varicose veins in the lower esophagus with obvious red sign, gastric varices, formation of larger venous bulbs, obvious red sign, and different degrees of congestion and erosion of the gastric mucosa (Figure 1).

Color Doppler ultrasound and contrast-enhanced Computed Tomography (CT) of the upper and middle abdomen showed no proximal inferior vena cava thinning, no abnormal blood flow velocity, liver deformation, intrahepatic portal vein thinning, thickening of the trunk, massive varicose veins in the esophagus and fundus, and enlarged spleen (Figure 2).

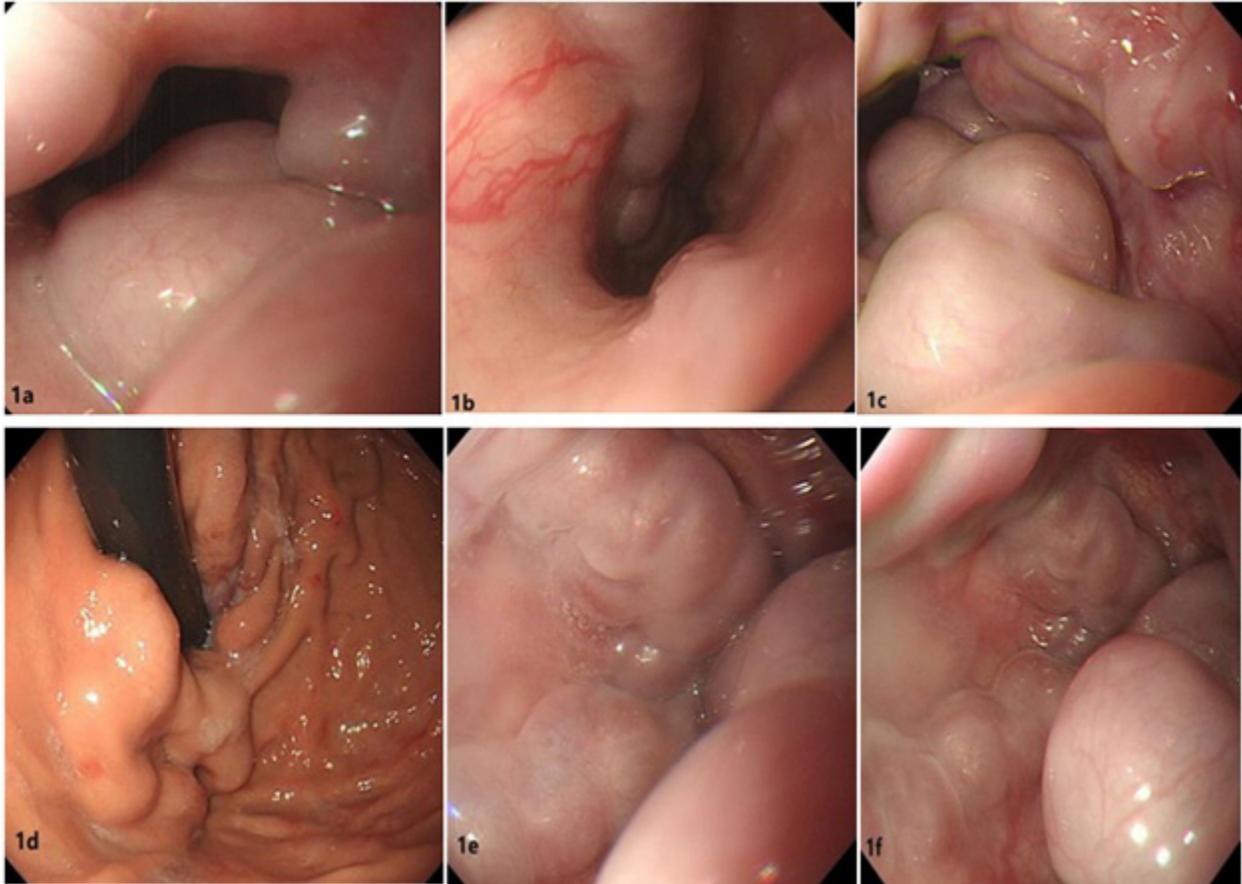


Figure 1: Preoperative endoscopic findings. a–c: Four varicose veins of the lower esophagus, red sign and too thick. d–f: Venous bulb formation of fundus variceal veins, red sign, and different degrees of congestion and erosion of the gastric mucosa.

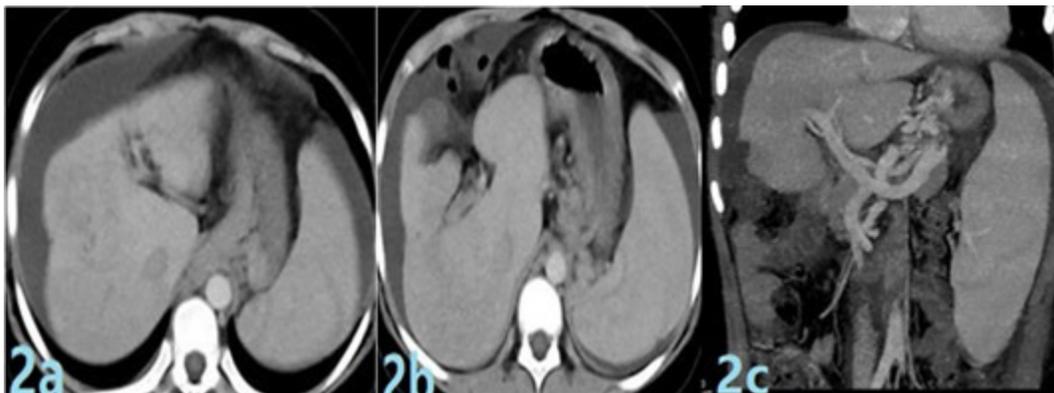


Figure 2: Preoperative upper abdominal CT findings. a and b: Abdominal contrast-enhanced CT transverse axial view: liver deformation, widened hepatic fissure, intrahepatic portal vein thinning, thickening of the main trunk, and small amount of ascites. c: Sagittal abdominal contrast-enhanced CT: main portal vein thickened and left gastric vein enlarged. Thick, massive varicose veins in the fundus and lower esophagus, enlarged spleen, and small amount of ascites.

3.4. Treatment and outcome

After drug treatment, bleeding stopped and emergency endoscopic treatment was performed. However, the esophagogastric varices were too thick to be ligated and endoscopic sclerotherapy was feasible. Because the child visited another hospital many times, the family members had reservations about endoscopic treatment and chose to refuse it. After repeated discussions, the family opted for TIPS first and liver transplantation later. During this period, the growth of the child was observed.

TIPS procedure. The right internal jugular vein was punctured under general anesthesia as described previously [5]. The RUPS-100 puncture system was introduced through a guidewire, into the right hepatic vein, and successfully punctured from the proximal end of the right hepatic vein to the left branch of the intrahepatic portal vein. All varicose veins were embolized with coils, and the intrahepatic puncture tract was dilated with a balloon. The covered stent was implanted in the shunt (Viatorr; 8 × 60 × 20 mm).

For post-stent implantation, follow-up portal vein angiography showed good intrahepatic portal vein perfusion, with portosystemic gradient reduced from 45 to 20 mmHg (Figure 3).

On postoperative days 1 and 5, color Doppler ultrasound showed that the shunt was unobstructed and the blood flow velocity was normal (Figure 4).

On postoperative day 5, the laboratory tests showed: hemoglobin: 78 g/L (male: 120–160 g/L), red blood cells: 1.62×10^{12} (child: 4.2×10^{12} – 5.2×10^{12} /L), white blood cells: 1.42×10^9 (4.0109 – 10.0×10^9 /L), platelets: 51×10^9 (100×10^9 – 300×10^9 /L); PT: 13.20 s; APTT: 26.2 s; serum albumin: 29.7 g/L (40–55 g/L), total bilirubin: 12.9 μ mol/L (0–23 μ mol/L), alanine aminotransferase: 15 U/L (7–40 U/L), aspartate aminotransferase: 22 U/L (13–35 U/L), blood ammonia 67 μ mol/L (0–60 μ mol/L). The general condition of the patient was good after the operation, and he was discharged from the hospital without any discomfort. The patient will be followed up at 1, 3 and 6 months, and 1–5 years.

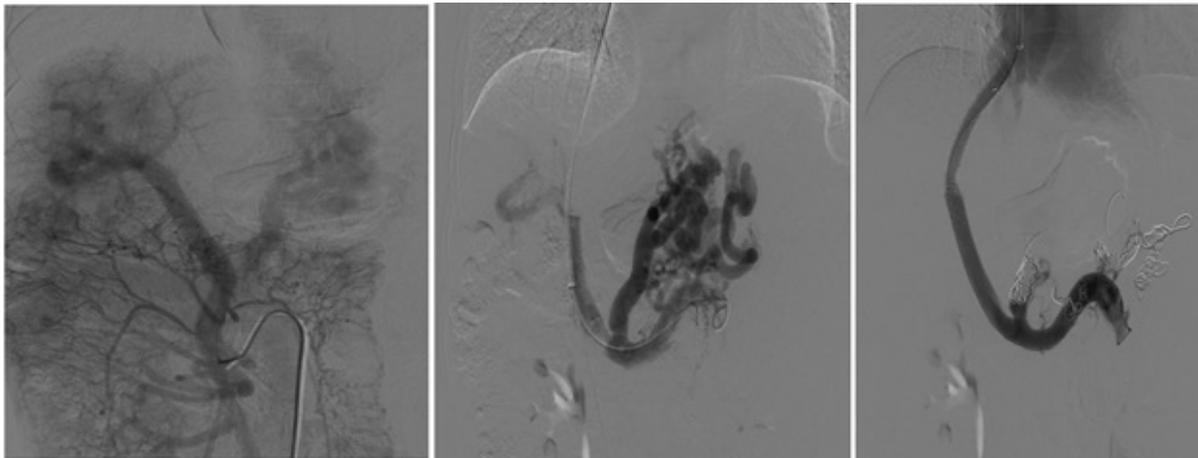


Figure 3: Intraoperative performance of TIPS. a: Indirect portal venography showed the whole course of the portal venous system. b: A catheter in splenic venography showed thickening of the main portal vein. The left gastric vein, gastric fundus vein and lower esophageal veins were thickened. c: Implanted membrane after stenting, collateral vessels were not re-imaged.

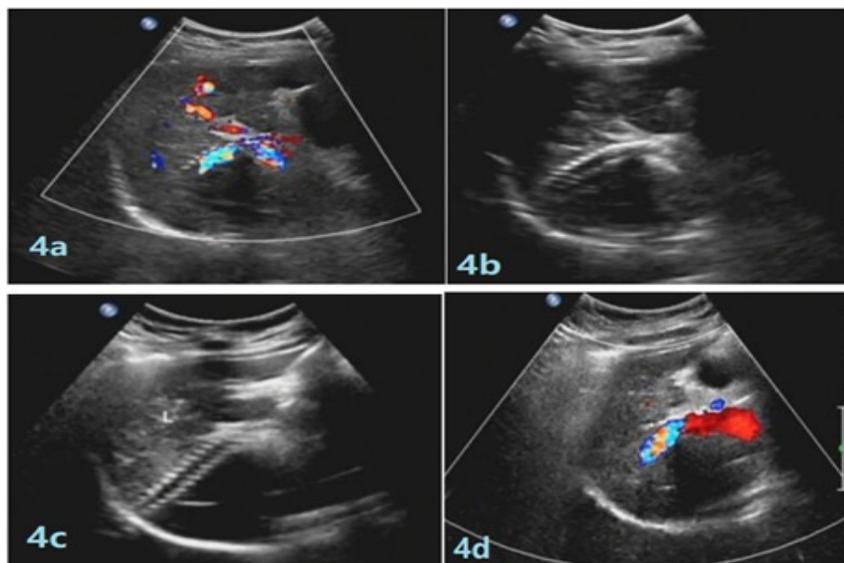


Figure 4: Ultrasonography of portal vein after TIPS. a: Re-examination of color Doppler ultrasound on postoperative day 1 showed normal blood flow velocity. b: The shunt was unobstructed. c: Re-examination of color Doppler ultrasound on postoperative day 5 showed that the shunt was unobstructed. d: The blood flow velocity was normal.

3.5. Postoperative follow-up

Color Doppler ultrasound at 1, 3, 6 and 12 months after the operation showed that the intrahepatic shunt channel was unobstructed, liver function tests were normal, blood routine showed that the cells of the three systems were low, and blood ammonia was about

70 $\mu\text{mol/L}$ (0–60 $\mu\text{mol/L}$). Endoscopy showed that the esophageal fundus varices gradually disappeared. Enhanced CT in the middle and upper abdomen showed that the shunt channel was unobstructed, and the esophageal fundus varices gradually atrophied (Figure 5). The patient was re-examined as planned, and no other related treatments were given.

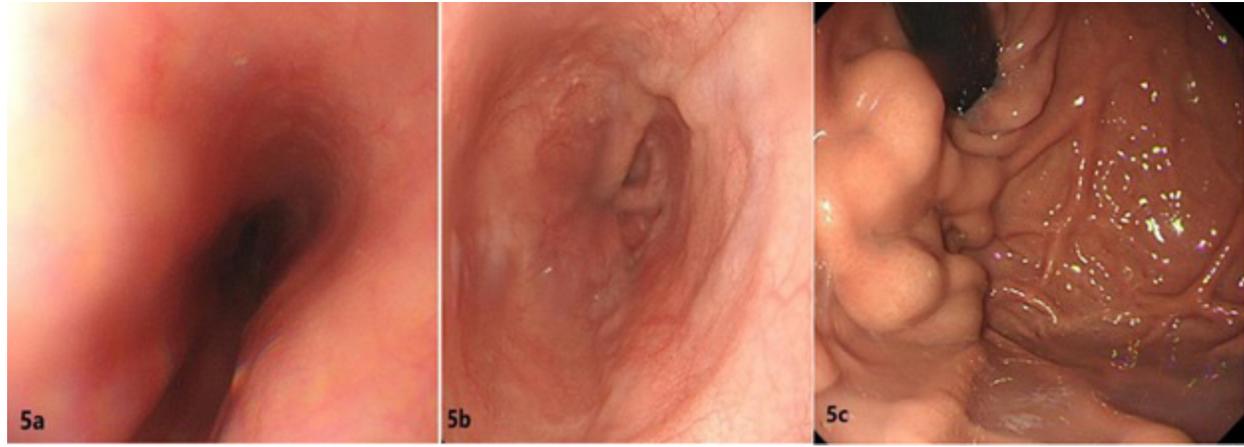


Figure 5: Gastroscopic findings 3 months after TIPS. a and b: Gastroscopy showed disappearance of the esophageal varices and disappearance of the red sign. c: Gastroscopy showed gradual narrowing of the fundus varices and no congestion or erosion of the gastric mucosa.

4. Discussion

Portal hypertension of liver cirrhosis is caused by a variety of etiologies and has complex pathophysiological changes and clinical manifestations. There are many treatment methods, including drug therapy, endoscopic band ligation sclerotherapy, splenectomy and devascularization, and portal–vena cava shunt, but so far there is no ideal solution.

TIPS is an interventional technique that uses stents to establish a shunt channel in the hepatic parenchyma between the hepatic vein and portal vein, so as to shunt the portal vein blood flow and reduce the portal vein pressure. This technique was first reported by Rösch in 1969 [6]. Richter was first used in clinical practice in 1989. Since then, TIPS has been greatly improved in terms of surgical effectiveness and safety due to the improvement of puncture and intubation techniques and the emergence of covered stents. The Baveno VII consensus meeting clearly pointed out that in patients with upper gastrointestinal hemorrhage (endoscopic classification IGV1, IGV2I, and esophageal varices), if there are high risk factors for treatment failure such as Child–Pugh class B or C with active bleeding after the initial endoscopic combination therapy, TIPS should be considered within 3 days and ideally within 24 h [7].

TIPS is widely accepted as a safe and effective method for the treatment of upper gastrointestinal hemorrhage, refractory ascites, hepatorenal syndrome, hepatopulmonary syndrome, Budd–Chiari syndrome and other related complications caused by portal hypertension. It controls the symptoms caused by portal hypertension, and has significantly improved the quality of life and survival

rate [8]. All current guidelines for the diagnosis and treatment of liver cirrhosis, including the European Association for the Study of the Liver (guidelines for decompensated cirrhosis), the American Association for the Study of Liver Diseases, Baveno VII consensus, and the Interventional Physicians Society, Chinese Medical Doctor Association Portal Hypertension Clinical Practice Guidelines for the treatment of portal hypertension in adults with cirrhosis [9, 10], do not specifically describe TIPS for treatment of portal hypertension in children with cirrhosis.

Our patient had a diagnosis of cirrhosis of unknown origin, multiple gastrointestinal bleeding, and was indicated for TIPS according to guidelines and expert consensus. In addition, the patient was 167 cm tall, weighed 60 kg, and had an adult body shape, so the use of smaller interventional devices was not considered during TIPS. RUPS-100 puncture kits, balloons, and covered stents used in TIPS are based on adult equipment standards. Local anesthesia is generally used for TIPS in adults. Considering that the child may not cooperate or be nervous during the operation, the operation is carried out by general anesthesia in children, and there are no complications related to surgery and general anesthesia after the operation. The wedge pressure of the portal vein decreased by about one third during the operation, which was not too great a reduction, considering the possibility of hepatic encephalopathy after TIPS.

After the operation, the vital signs of the child were stable. The next day's re-examination of the portal vein color Doppler showed that the blood flow in the shunt channel was smooth. Hemoglobin level was not decreased and liver function was normal. He was

then discharged from hospital. Postoperative re-examination at 1, 3, 6 and 12 months by color Doppler ultrasound showed that the intrahepatic shunt channel was unobstructed, liver function tests were normal, the blood routine showed that the cells of the three systems were low, and blood ammonia level was about 70 $\mu\text{mol/L}$ (0–60 $\mu\text{mol/L}$). Endoscopy showed that the esophageal fundus varices gradually disappeared. Enhanced CT in the middle and upper abdomen showed that the shunt channel was unobstructed, and the esophageal fundus varices gradually atrophied. The patient was reviewed and no other related treatment was performed, indicating that TIPS is safe and effective compared with TIPS after gastrointestinal hemorrhage in adults with liver cirrhosis, and its postoperative evolution is similar. It also indicates that TIPS treatment in children with liver cirrhosis is worthy of promotion.

5. Conclusion

TIPS therapy in children with cirrhotic portal hypertension is technically feasible and has a good effect for treatment and prevention of esophageal and gastric variceal bleeding. Given its high clinical success rate, good shunt patency and survival, TIPS should be considered as a reliable bridge before liver transplantation in children with cirrhotic portal hypertension.

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