CardioVascular and Interventional Radiology

Published Online: 6 November 2002

Cardiovasc Intervent Radiol (2002) 25:484–493
DOI: 10.1007/s00270-002-1913-6

Transjugular Intrahepatic Portosystemic Shunts in Children with Biliary Atresia

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Abstract

Purpose: We retrospectively evaluated the technical and long-term clinical results of transjugular intrahepatic portosystemic shunts (TIPS) in children with portal hypertension and biliary atresia (BA).
Methods: Nine children with BA and recurrent bleeding from esophagogastric and/or intestinal varices were treated by TIPS at the age of 34–156 months and followed-up in two centers. Different types of stents were used.
Results: Shunt insertion succeeded in all patients, but in two a second procedure was necessary. Seven procedures lasted more than 3 hr, mainly due to difficult portal vein puncture. Variceal bleeding ceased in all patients; however, 16 reinterventions were performed in eight patients for clinical reasons ($n = 11$) and sonographically suspected restenosis ($n = 5$). Four patients underwent successful liver transplantation 4–51 months after TIPS and five are in good clinical conditions 64–75 months after TIPS.
Conclusions: TIPS in children with BA is technically difficult, mainly due to periportal fibrosis and small portal veins. Frequency of reinterventions seems to be higher compared with adults.

Key words: Shunt, transjugular intrahepatic portosystemic (TIPS)/Biliary atresia/Children

Biliary atresia (BA) is characterized by fibrous obliteration of extrahepatic bile ducts, cholestatic syndrome and transition to biliary cirrhosis in end-stage disease. The incidence is one in 12 000–15 000 newborns; the etiology is unknown [1]. Hepatoporto-enterostomy (HPE) is the standard surgical procedure to overcome biliary obstruction [2, 3]. However, despite sustained bile flow, progression of liver fibrosis and cirrhosis causes clinical manifestations of portal hypertension in 23–29% of long-term survivors [2–5]. The patient's age at the time of HPE predicts progression of liver disease and survival [3, 6].

Hepatic fibrosis in BA predominantly surrounds portal veins, diminishing luminal diameter and causing presinusoidal portal obstruction, mostly accompanied by nearly normal liver function. Frequent complications of portal hypertension are variceal bleeding and hypersplenism. In the majority of patients bleeding originates from esophageal and gastric varices; however, HPE also predisposes to portosystemic varices at jejuno-jejunostomies and external jejunostomies [7, 8]. Hepatocellular insufficiency is present in patients with advanced cirrhosis.

The experience with transjugular intrahepatic portosystemic shunt (TIPS) in children is small, and in particular long-term results are lacking. In children with BA periportal fibrosis and the small size of the portal veins are particular problems. This article presents a retrospective analysis of the technical and clinical results in children with BA treated by TIPS and followed up in two centers.

Materials and Methods

Between August 1992 and October 1996 nine children with BA and complications of portal hypertension were treated by TIPS. Patient characteristics are listed in Table 1. Indication for treatment was recurrent massive variceal bleeding of different and sometimes

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Table 1. Patient characteristics

Patient no.	Age at TIPS	Age at HPE (weeks)	Histology at HPE	Child class	Indication for TIPS	Follow-up period
1	2 y 10 mo	7	PPF II°	A	EVB	75 mo
2	4 y 6 mo	4	Biliary cirrhosis	B	IVB	8 mo–LTx
3	6 y 0 mo	19	PPF IV°	A	EVB, IVB	69 mo
4	6 y 7 mo	13	PPF III°	A	EVB	51 mo–LTx
5	6 y 10 mo	9	Biliary cirrhosis	B	IVB, EVB	4 mo–LTx
6	10 y 3 mo	21	PPF III°	A	IVB, EVB	64 mo
7	11 y 10 mo	7	PPF IV°	A	GVB,	68 mo
8	11 y 10 mo	8	Biliary cirrhosis	B	EGVB, ascites	27 mo–LTx
9	12 y 8 mo	20	PPF III°	A	EGVB	72 mo

HPE, hepatoporto-enterostomy; y, years; mo, months; PPF, periportal fibrosis, grading I°–IV° see [6]; EVB, esophageal variceal bleeding; GVB, gastric variceal bleeding; EGVB, esophageal and gastric variceal bleeding; IVB, intestinal variceal bleeding; LTx, liver transplantation.

multiple origin. Patients with esophageal and/or gastric varices had been treated previously by means of endoscopic sclerotherapy. Bleeding from intestinal varices was suspected if endoscopy failed to prove esophageal and gastric variceal bleeding and intestinal varices were seen during angiography (Fig. 1A). In seven children hypersplenism was present with platelet counts below 100 000/μl; one child suffered from ascites and no child had signs of hepatic encephalopathy before TIPS. Five children (patients 3, 4, 5, 6 and 8) were treated and followed up at the University Hospital of Brussels (center A) and four children (patients 1, 2, 7 and 9) were treated and followed up at the University Hospital of Tübingen (center B).

Due to the small number of patients over time and because of the lack of data in the literature we did not follow a strict prospective protocol. Technical details of procedures were addressed individually (Table 2). Treatments were performed under general anesthesia via 9 Fr sheaths inserted into the right internal jugular vein. In the four children treated in center B, initially 5 Fr Cobra catheters were advanced peripherally into branches of the middle or right hepatic vein to obtain wedge veno-portography. Portography was successful in two of three children using manual iopromide injection and failed in one. In another child manual carbon dioxide injection was used successfully. In three children the hepatic veins showed a peripherally curved course impeding a stable position of guiding catheters and needles before portal vein puncture.

Portal vein puncture was initially tried in four children with miniaturized needles designed for pediatric use (TIPSI-101-PED, Cook, Denmark). In two of these cases (patients 3 and 4) 19 gauge needles were used, and in another two cases (patients 1 and 2) 17 gauge needles. Puncture failed in all four cases because of inadequate needle stiffness and insufficient needle guidance. Subsequently 16 gauge standard Colapinto needles were successfully used in these patients. In the remaining five children 16 gauge standard needles covered by Teflon sheath catheters (RTPS-100, Cook) were also used. Continuous ultrasound monitoring using 3.5 MHz and 5.0 MHz probes, respectively, was performed (US, EMS) during portal vein puncture in all patients. The probe was held by a second operator in a semisagittal plane via a lateral intercostal approach, trying to visualize both the needle tip and main right portal vein branch simultaneously. After entering a portal vein branch a steerable guidewire (0.035 inch diameter; Terumo, Tokyo, Japan) was gently advanced followed by a 5 Fr catheter for exchange of a 180 cm stiff wire (0.035 inch Amplatz wire, Boston Scientific). The parenchymal tract was predilated with 6 mm/4 cm balloons (Ultrathin, Boston Scientific) and a portal venogram with 30° right anterior oblique projection was obtained to localize the entrance into the portal vein system in relation to the portal vein bifurcation and to exclude portal vein leakage.

For shunt creation different types of stents were preferred in the two institutions. In three of the four children treated at center B, Wallstents were the first choice. The intention in using Wallstents was to obtain gently curved shunts without tight angulations and to leave 1–2 cm of the terminal portion of the stent within the main portal and hepatic veins, respectively (Fig. 2). The exception was a 6-year-old child (patient 2). In this patient a Palmaz stent was implanted initially because significant recoil was recognized during predilatation. The Palmaz stent was coaxially covered by one Wallstent, also leaving 1–2 cm of the stent within the portal and hepatic vein. In center A Palmaz stents were used for all but two patients. In one child (patient 3) significant portal vein leakage was discovered angiographically after predilatation and treated by implantation of a covered Cragg stent (MinTec, Freeport, Bahamas) followed by the insertion of two uncovered Cragg stents to complete the shunt. In another child (patient 4) two Wallstents were implanted because of a tightly angulated access to the portal vein.

Stent diameters were selected in relation to the child's age and the size of the parent portal vein. Initial stent dilatation was performed with a 6 mm diameter balloon followed by stepwise widening in order to lower the portocaval pressure gradient significantly in relation to pretreatment values. Length and number of implanted stents had to be adjusted to the tract, including the additional 1–2 cm left within the main portal vein and within the hepatic veins. We used a maximum total contrast material load (300–350 mg iodine/ml) of 4 ml per kilogram body weight. The interventional procedures lasted 3–6 hr including anesthesia. Procedure times of 5 and 6 hr in four patients were mainly due to several attempts at portal vein puncture using miniaturized needles and due to embolization of varices.

Embolization of gastric and esophageal varices was performed in five children. These patients had bled from the varices and opacification of varices persisted angiographically after final stent dilatation. For embolization Gianturco coils (Cook) were used (diameter 5–8 mm, length 50–80 mm) in four children (patients 3, 4, 5 and 6) and butylcyanoacrylate (Histoacryl, Braun-Melsungen, Germany) lipiodol (1.5 cm^3) in one child (patient 7). Materials were selected according to the personal preference of the operator. After shunt insertion 1000 IU heparin/10 kg body weight was given in four patients and heparinization was continued for 48 hr (PTT 40–60 sec) with the intention of preventing early thrombosis of small-sized shunts. In three children with major bleeding episodes

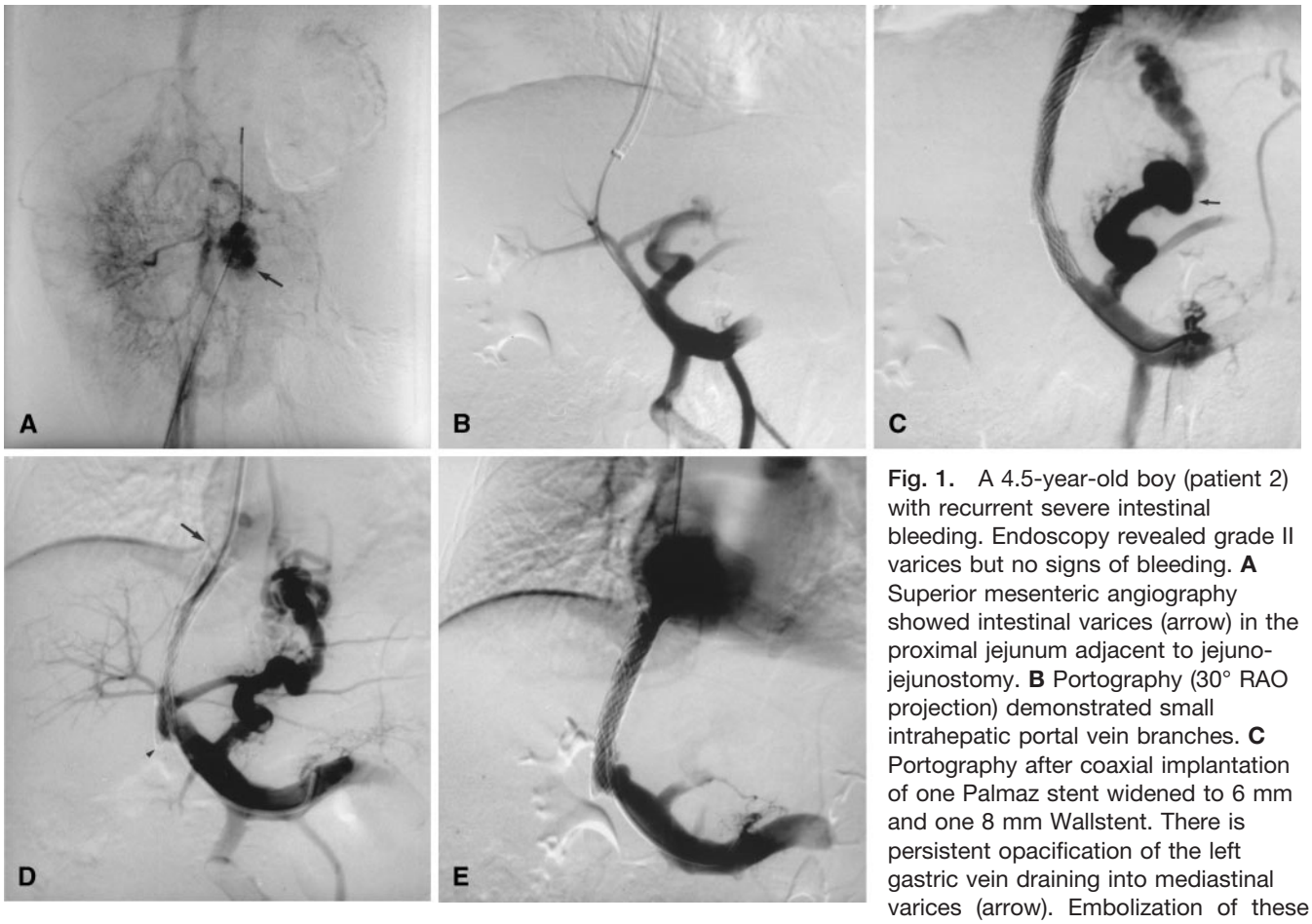

Fig. 1. A 4.5-year-old boy (patient 2) with recurrent severe intestinal bleeding. Endoscopy revealed grade II varices but no signs of bleeding. **A** Superior mesenteric angiography showed intestinal varices (arrow) in the proximal jejunum adjacent to jejuno-jejunostomy. **B** Portography (30° RAO projection) demonstrated small intrahepatic portal vein branches. **C** Portography after coaxial implantation of one Palmaz stent widened to 6 mm and one 8 mm Wallstent. There is persistent opacification of the left gastric vein draining into mediastinal varices (arrow). Embolization of these varices was not performed, because bleeding originated from intestinal varices. **D** Portography 5 months later revealed diffuse restenosis in the stent tract. There was nearly 5 mm displacement of the proximal end of the Wallstent into the vena cava (arrow) and impression of the portal vein wall by the distal stent portion (arrowhead). **E** No residual stenosis is seen after coaxial implantation of one Palmaz stent.

before treatment and in two children with procedure-related bleeding complications no heparin was given. Antibiotics were given in four children for 6–9 days to prevent infections after implantation of a covered stent (1 patient), and accidental puncture of the liver capsule (1 patient) and biliary ducts (2 patients).

During follow-up clinical examinations, laboratory investigations (serum ammonia, serum bilirubin, cholinesterase, alanine aminotransferase, aspartate aminotransferase and platelet count), sonography (morphology of liver and spleen) and color-coded Doppler sonography (flow and velocity within the shunt and in the portal vein) were performed 2 days, 1 week, 1 month, 3 months, 6 months, 9 months and 12 months after TIPS insertion and thereafter every 6 months. Shunt stenosis was suspected sonographically if one of the following criteria was present: lowering of peak velocity within the portal vein trunk by 50% or more compared with the initial post-TIPS value, peak velocity within the stent shunt below 40 cm/sec and re-establishment of a reversed flow within the intrahepatic portal veins. The mean follow-up was 69.6 (64–75) months in five children in whom liver transplantation was not performed. Follow-up data attained through September 2000 are included. In four children the mean time interval between TIPS insertion and liver transplantation was 22.5 (4–51) months.

A two-tailed paired Student's *t*-test was performed to evaluate significant changes in the laboratory data before TIPS and during follow-up.

Results

Technical Results

Shunt insertion succeeded in seven children during the first procedure and in two during a second procedure. In the latter, portal vein access could not be obtained during the first procedure despite 20–25 puncture attempts. Impeded sonographic visualization of target vessels due to injected fluids and limitation of contrast material load terminated the first sessions. During the second session, targeting was improved in one child (patient 6) by using a 0.018 inch guidewire inserted percutaneously into the portal vein via a 21 gauge needle. The puncture of portal vein branches was difficult in all patients with advanced periportal fibrosis due to high tissue resistance and small vessel diameters (Fig. 1B). Especially in small children mismatch of puncture

Table 2. Methods of TIPS insertion

Patient no.	Stents			No. of PV puncture attempts	Pc gradient (mmHg)		Procedure time
	Type	No.	Final diameter (mm)		Pre-TIPS	Post-TIPS	
1	WS	1	6	18	24	8	4 hr
2	PS	1	6		13	8	6 hr
	WS	1	8	15			
3	CS	3	7	17	16	9	5 hr 47 min
4	WS	2	7	12	18	7	5 hr 5 min
5	PS	1	7	2	12	8	1 hr 45 min
6	PS	3	7	25			
				16	16	9	3 hr 53 min
							2 hr 7 min[a]
7	WS	3	8	8	14	10	5 hr
8	PS	2	8	3	25	13	3 hr 5 min
9	WS	2	9	20			
				10	19	13	3 hr
							3 hr[a]

WS, Wallstent; PS, Palmaz stent; CS, Cragg stent; PV, portal vein; PC gradient, portocaval pressure gradient.
[a]Second procedure.

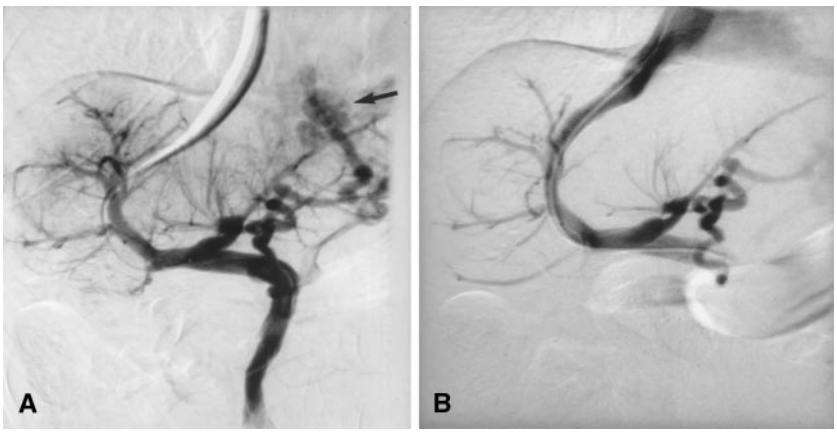

Fig. 2. A, B. A girl aged 12 years 8 months with esophageal and gastric variceal bleeding (patient 9). **A** Thirty degrees RAO portography shows the curved vessel anatomy. Implantation of a Wallstent was preferred. There is opacification of gastric varices (arrow). **B** Angiography in same projection after insertion of two Wallstents shows the gently curved shunt. There is no further opacification of varices.

needle size and vessel diameter required subtle guidewire maneuvers to enter the portal vein lumen.

Analysis of hemodynamic parameters showed that in four children (age range 34–79 months; patients 1, 2, 3 and 4) minimal shunt diameters of 6–7 mm resulted in a reduction in the portocaval pressure gradient by a mean value of 52% (38–67%). In three older children (age range 142–152 months; patients 7, 8 and 9) shunt diameters of 8–9 mm were related to reductions in pressure gradients by a mean value of 36% (29–48%) (Table 2).

Clinical Results

Hemorrhage ceased after TIPS insertion in all patients, including bleeding from gastroesophageal as well as from intestinal varices. Seven episodes of rebleeding occurred in six of nine (67%) patients within intervals of between 7 days and 48 months (Table 3). Rebleeding always originated from the same sites before TIPS insertion and was caused by shunt restenoses in all patients. Hypersplenism did not change significantly after TIPS insertion. Splenomegaly remained unchanged and platelet counts increased only in four of nine patients. Mean values of platelet counts were 81.7 (35–110) $\times$ $10^3/\mu$l before TIPS and 84.4 (52–124) $\times$ $10^3/\mu$l 1–12 months after TIPS. There was no significant difference in mean serum levels of total bilirubin, ammonia and cholinesterase before and after TIPS (Table 4). Laboratory tests revealed stable liver function in five patients (patients 1, 3, 6, 7 and 9) after TIPS. These patients are now in good clinical condition after a mean follow-up of 69.6 (64–75) months and are not active candidates for liver transplantation. Three of them had been active transplantation candidates prior to TIPS insertion. In four patients deterioration of liver function occurred during follow-up after TIPS. Two of them (patients 4 and 8) developed chronic liver dysfunction and elective liver transplantation was performed 27 and 51 months after TIPS, respectively. In two children (patients 2 and 5) staged as Child B class cirrhosis prior to TIPS, urgent liver trans-

Table 3. Reinterventions

Patient no.	Interval to TIPS reintervention	Indication	Procedure	Pc gradient (mmHg)	
				Pre-reintervention	Post-reintervention
1	19 mo	Rebleeding of EV	6 mm PTA	16	12
2	5 mo	Rebleeding of IV	PS + 7 mm PTA	14	4
3	5 mo	Stenosis at sonography	8 mm PTA	17	8
	13 mo	Stenosis at sonography	8 mm PTA	17	10
	29 mo	Stenosis at sonography	9 mm PTA	16	10
4	8 mo	Progression of EV	8 mm PTA	11	7
	25 mo	Progression of EV	PS + 8 mm PTA	15	8
	30 mo	Progression of EV	8 mm PTA	15	9
	40 mo	Rebleeding of EV	Second TIPSS (3 PS + 9 mm PTA)	15	8
	48 mo	Rebleeding of EV	9 mm PTA	16	8
5	7 d	Rebleeding of IV	8 mm PTA	11	6
6	3 mo	Rebleeding of EV	7 mm PTA	22	10
	28 mo	Stenosis at sonography	8 mm PTA	15	8
7	10 mo	Rebleeding of GV	8 mm PTA	16	10
8	10 mo	Recurrence of ascites	WS + 9 mm PTA	18	9
	17 mo	Stenosis at sonography	10 mm PTA	11	6
9	–	–	No reintervention	–	

mo, months; EV, esophageal varices; GV, gastric varices; IV, intestinal varices; PS, Palmaz stent; WS, Wallstent; Pc gradient, portocaval pressure gradient.

Table 4. Laboratory findings before TIPS insertion and during follow-up

Patient no.	Time[a]	Serum ammonia (μmol/l)	Serum bilirubin (mg/dl)	Cholinesterase (kU/l)	Platelet count ($10^3/\mu$l)	Spleen length (cm)	ALAT (U/l)	ASAT (U/l)
1	0	103	2.3	2.1	35	11	23	19
	1	n.a.	1.2	2.6	44	11	66	71
	2	44	0.6	3.1	111	13	44	48
2	0	83	8.8	1.2	83	16	81	79
	1	144	10.2	1.2	90	17	121	134
	2	125	13.6	1.3	124	14	157	125
3	0	118	1.3	n.a.	95	13	69	93
	1	145	2.9	n.a.	48	13	230	299
	2	114	1.7	n.a.	52	16	77	66
4	0	77	1.4	n.a.	82	12	52	61
	1	n.a.	1.6	n.a.	81	12	n.a.	n.a.
	2	113	3.1	3.5	68	13	64	95
5	0	125	1.1	n.a.	110	10	42	49
	1	130	1.1	n.a.	108	10	54	70
	2	190	1.9	n.a.	80	12	70	92
6	0	49	0.9	n.a.	105	14	43	70
	1	n.a.	n.a.	n.a.	104	14	49	78
	2	62	1.6	n.a.	85	13	42	60
7	0	68	1.2	4.6	42	20	17	12
	1	47	1.7	4.5	34	20	25	25
	2	42	1.2	3.1	53	20	16	13
8	0	81	1.1	2.0	90	10	72	70
	1	n.a.	1.3	n.a.	92	10	95	98
	2	107	2.7	1.7	66	14	120	132
9	0	116	1.2	2.9	93	19	102	121
	1	90	1.2	2.6	121	19	96	130
	2	75	1.2	3.6	121	18	39	53

ALAT, alanine aminotransferase; ASAT, aspartate aminotransferase; n.a., values not available.
[a]Time: 0, before. TIPS; 1, mean value 1–10 days after TIPS; 2, mean value 1–12 months after TIPS.

plantation was necessary 8 months and 4 months after shunting, respectively. In both patients bleeding was stopped initially after TIPS insertion (with reintervention in patient 5) and their general condition improved; however, progressive liver insufficiency required transplantation.

Complications

In two patients bleeding complications were related to the procedure. In a 6-year-old (patient 3) portal vein leakage was discovered angiographically at the portal vein entry site

following predilatation of the parenchymal tract. The leak was closed successfully by implantation of a covered stent, but intraperitoneal hematoma had to be removed surgically to overcome abdominal distension and to prevent infection. In a 13-year-old girl (patient 9) transcapsular liver puncture caused sonographically proven moderate intraperitoneal bleeding after successful TIPS insertion, which was managed by the application of two red blood cell packs. No bleeding complications were seen in the four children with 48 hr heparinization.

Hepatic encephalopathy developed in a 6-year-old girl (patient 5) who was classified as Child B cirrhosis prior to TIPS. Encephalopathy arose simultaneously with progressive hepatic insufficiency following shunt redilatation 1 week after shunt insertion and 4 months before liver transplantation.

Marked impression of the portal vein wall by a Wallstent with simultaneous stent protrusion into the inferior vena cave was seen 5 months after stent implantation in a 4.5-year-old boy (patient 2). The child underwent reangiography because of recurrence of jejunal variceal bleeding and the study revealed additional intrahepatic stent shunt stenosis (Fig. 1D).

Restenoses and Reinterventions

In eight of nine (89%) children a total of 16 reinterventions were performed. Indications for reintervention were rebleeding ($n = 7$), recurrent ascites ($n = 1$), progression of esophageal varices ($n = 3$) and shunt stenosis suspected sonographically during routine follow-up ($n = 5$). Four patients had one reintervention, two patients had two reinterventions, one patient had three and another one had five reinterventions. The mean interval between TIPS insertion and the first reintervention was 7.5 months (range 7 days to 19 months). In one child (patient 5) rebleeding occured 7 days after TIPS implantation due to partial occlusion of a 7 mm stent tract by clotting. In this child no heparin was given after the initial TIPS procedure because of a severe variceal bleeding episode prior to shunting. No rebleeding occured after redilatation; however, progressive liver insufficiency and hepatic encephalopathy developed during the following 4 months until liver transplantation.

In 12 of 16 procedures, balloon dilatation was sufficient to overcome shunt stenosis and in a further three stent implantation was necessary. In one case (patient 4) a second parallel shunt had to be inserted after redilatation and coaxial implantation of additional stents had failed to lower the portocaval pressure gradient sufficiently (Fig. 3). The mean portocaval pressure gradient was 15.4 (11–22) mmHg before reinterventions and 8.3 (4–12) mmHg after reinterventions. Technical details of the reinterventions are summarized in Table 3.

Discussion

BA is the most common underlying disease in children with liver cirrhosis, representing nearly 30% of cases [1, 4]. Periportal fibrosis is an important feature of BA and it seems likely that this condition makes TIPS more difficult, especially in small children. Periportal fibrosis is associated with a reduced size of the portal veins [6, 9] and is probably the cause of high resistance during portal vein puncture and the failed use of 19 gauge and 17 gauge needles in four of our patients. Similar experiences have been reported by two other groups using 18 gauge needles, with failed punctures in a total of five of five patients [10, 11]. Difficult portal vein puncture due to extensive periportal fibrosis was also described in a nearly 2-year-old girl suffering from BA using Roesch-Uchida and Colapinto standard needles [12]. In our study, portal vein punctures succeeded finally in all patients using 16 gauge needles; however, there was nevertheless a mismatch between needle size and vessel diameter necessitating qualified ultrasound monitoring and subtle guidewire maneuvers to enter the portal vein. Suitable miniaturized puncture instruments for TIPS in pediatric patients are not currently available.

Another condition impeding portal vein puncture was the peripheral course of the major hepatic veins in the right liver lobe in three children. Sonography demonstrated multiple hyperplastic parenchymal nodules located in the central parts of the liver displacing the right and the middle hepatic vein peripherally. Tortuous hepatic veins hindering portal vein puncture and precluding a TIPS procedure have also been described in a child with BA in another series [11]. Catheter-directed manual injection of carbon dioxide into peripheral hepatic veins is useful for obtaining portal vein opacification prior to portal vein puncture and to reduce the contrast material load. Successful application of this technique has been described in two children [12, 13].

Appropriate stent positioning in relation to the portal and hepatic veins is important in this group of patients, because potential liver transplantation has to be considered. After variable time intervals following HPE about 30% of patients need elective liver transplantation to overcome chronic liver failure [13, 14]. On the other hand body growth implies increasing liver size and increasing portal vein flow, especially in non-cirrhotic livers [9]. The use of Wallstents is advantageous in terms of the creation of gently curved stent tracts including intravenous stent portions. On the other hand Palmaz stents can be placed more precisely and allow the adjustment of shunt diameter if necessary during long-term follow-up. Despite favorable initial stent positioning, shrinkage of the liver parenchyma carries the risk of relative displacement of any type of stent. In one of our patients (patient 2) this was probably causative for stent protrusion into the inferior caval vein and impression of the portal vein wall (Fig. 1C, D). Shortening and straightening of the stent are additional suspected reasons. A case report in the literature describes the perforation of a 10 mm Wallstent from

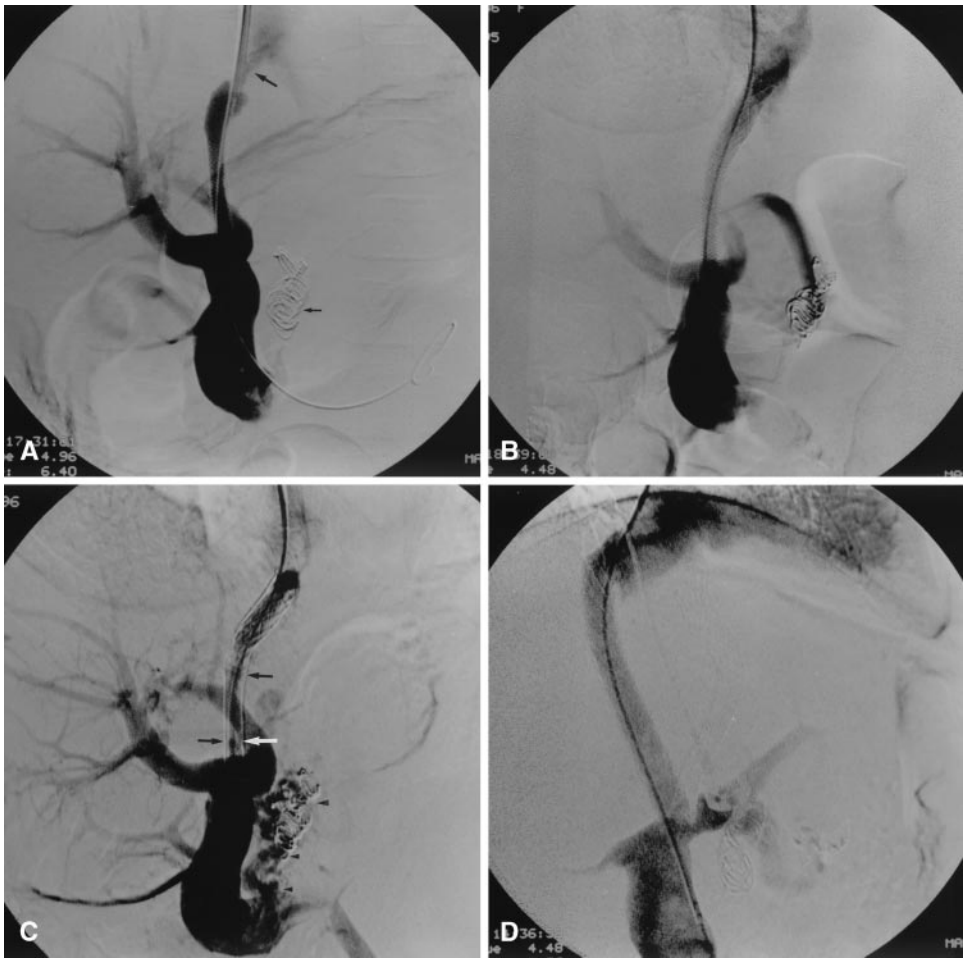

Fig. 3. A–D. Shunt reinterventions in a 6.5-year-old girl (patient 4) 25 months (**A**, **B**) and 40 months (**C**, **D**) after initial TIPS placement. **A** Angiography revealed tight hepatic vein stenosis (large arrow). The left gastric vein had been embolized by coils initially (small arrow). **B** After implantation of one 8 mm diameter Palmaz stent a sufficient lowering of the portocaval pressure gradient was achieved from 15 to 8 mmHg. **C** Fifteen months later there is diffuse restenosis (arrows) of the intrahepatic stent tract and opacification of the left gastric vein (arrowheads). Despite high-pressure balloon dilatation, the portocaval pressure gradient remained at 15 mmHg. **D** Angiographic result after creation of a parallel shunt with three 9 mm Palmaz stents, lowering the gradient to 8 mmHg.

the main portal vein into the adjacent jejunal loop of HPE followed by stent thrombosis and infection in a nearly 2-year-old child with BA [12]. Implantation of covered stents is the method of choice to treat procedure-related major lacerations of the extrahepatic portal vein associated with bleeding. Covered stents are also an option to reduce the risk of restenoses in patients with iatrogenic fistulas between the portal venous system and the biliary tree demonstrated following portal vein puncture.

Restenosis occurred in eight of nine patients, was associated with rebleeding in six and resulted in a mean number of 1.8 (16/9) reinterventions per patient during a mean follow-up of 48.7 (4–75) months. The rate of restenosis in this group of patients seems to be higher than in adults. This is probably related to the lower shunt diameters in children. The diameter of the shunt was 6–7 mm in four of five patients aged 2–7 years and 8–9 mm in two of three children aged 10–12 years, respectively. Our intention was to create initially small shunt diameters to obviate progressive hepatic encephalopathy. Intimal hyperplasia will probably cause more severe restenosis in these small shunts compared with the shunts of 9–12 mm diameter usually implanted in adults [15]. Increasing portal venous flow due to the child's growth is another potential reason necessitating shunt dilatation. In five of eight patients with restenosis the final luminal diameter after reintervention was 1–2 mm greater than the corresponding final diameter of the initial shunt (see Tables 2 and 3; excluding shunt revision on day 7 in one patient) and in one child a second parallel shunt was necessary to lower the pressure gradient sufficiently.

In children, the treatment of complications of portal hypertension by TIPS has been reported to our knowledge in 36 patients [7, 8, 10–12, 16–23], 12 of whom (33%) suffered from BA. The reported technical success rate was 94% (34 of 36). The clinical success rate in terms of controlling variceal bleeding was 93% (28/30). Successful treatment of hypersplenism by TIPS was shown in one of three patients. Indications, methods and outcome are listed by case in Table 5. Obviously technical and clinical complications are a major issue in these procedures. The most frequent complication was early restenosis and reocclusion, which occurred in eight of 34 patients (24%) during the first 30 days after TIPS. Hepatic encephalopathy was seen in five of 34 patients (15%): mild and transient in four and severe in one case. In our series only one patient (11%) with Child B liver disease prior to TIPS developed encephalopathy after treatment. Following surgical shunting in children the incidence of hepatic encephalopathy is below 5% in non-cirrhotic patients

Table 5. TIPS in children: data reported in the literature

Main author [reference]	No. of patients	Age (years)	Disease	Indication	Pc gradient (mmHg)		D (mm)	Complications and outcome	Clinical success	Follow-up	Liver Tx
					Pre-TIPS	Post-TIPS					
Kerns [19]	1	13	Cystic fibrosis	EVB	50	36	6	Transient HE	Yes	6 mo	No
Berger [18]	1	14	Cystic fibrosis	EVB	29	21	10	Rebleeding and redilatation +8 d	Yes	11 mo	No
Berger [17]	2	10	Histiocytosis	EVB	28	10	8	Reocclusion + 14 d, mild HE	Yes	9 mo	No
		11	BA	EGVB	25	13	8	No	Yes	4 mo	No
Lagier [7]	1	7	Secondary cirrhosis	SVB	10	6	8	No	Yes	4 mo	No
Weinberg [8]	2	4	Secondary cirrhosis	SVB	17	4	8	Residual stenosis and redilatation + 2 d	Yes	1 mo	Yes
		2.5	Secondary cirrhosis	SVB	12	5	8	No	Yes	10 mo	No
Ong [22]	1	10	Cystic fibrosis	GVB	n.r.	n.r.	10	Severe HE, liver failure, death +35 d	No GVB	n.r.	No
Johnson [20]	3	6	Biliary cirrhosis	EVB	18	4	8	Redilatation +2 d	Yes	14 d	Yes
		7	Congenital fibrosis	EVB	26	9	8	Redilatation +5 mo	Yes	8 mo	Yes
		11	BA	EGVB	26	12	8	Rebleeding and variceal embolization +4 d	Yes	6 d	Yes
Sergent [23]	1	1	Cystic fibrosis	IVB	26	0	8	Transient HE, liver insuff., death +22 d	No IVB	22 d	No
Heyman [11]	9	5	BA	VB	–	–	–	Technical failure, surgical shunt	–	–	No
		7	BA	VB	–	–	–	Technical failure	–	–	Yes
		5	BA	VB	n.r.	n.r.	8	No	Yes	6 d	Yes
		7	Congenital fibrosis	VB	n.r.	n.r.	6	Occlusion and recanalization +415 d	Yes	800 d	No
		9	Cryptogenic cirrhosis	VB	n.r.	n.r.	8	Occlusion and recanalization +24 d	Yes	66 d	Yes
		10	COACH syndrome	HSP	n.r.	n.r.	6	Occlusion and 2nd TIPS + 1 d	n.r.	40 d	No
		12	BA	VB	n.r.	n.r.	8	Occlusion +1 d, surgical shunt	No	1 d	No
		15	Cryptogenic cirrhosis	VB	n.r.	n.r.	8	Hepatic bleeding, death +1 d	No	1 d	No
		15	Sclerosing cholangitis	HSP	n.r.	n.r.	10	No	n.r.	35 d	Yes
Wang [12]	1	2	BA	EVB	32	4	8	Stent perforation into jejunum, occlusion 8 d	No VB	8 d	Yes
Steventon [21]	1	9	Autoimmune hepatitis	EGVB	28	11	8	No	Yes	2 d	Yes
Cao [13]	1	1	BA	EVB	17	5	5	Persisting sepsis, death +1 d	No EVB	1 d	No
Hackworth [10]	12	2.5	BA	VB	24	11	6	Mild HE +30 d(1 patient), transient pulmonary	VB: 10/10 yes	82 d	Yes
		4	BA	VB	30	13	8	edema day 0 (1 patient), shunt stenosis and		72 d	Yes
		5	Portal angiodysplasia	Ascites	26	13	7	redilatation + 186 d (1 patient)		301 d	No
		8	BA	VB	12	5	7		Ascites: 1/1 yes	357 d	No
		8	Histiocytosis	VB	n.r.	4	7			94 d	Yes
		9	Congenital fibrosis	VB	24	6	10			25 d	Yes
		9	BA	VB	15	12	7		HSP: 1/1 yes	47 d	Yes
		10	AAT deficiency	VB	21	10	8			29 d	Yes
		10	AAT deficiency	VB	18	9	10			9 d	Yes
		16	Allograft rejection	HSP	n.r.	8	8			127 d	Yes
		17	Transplant hepatitis C	VB	18	7	10			45 d	Yes
		17	Autoimmune hepatitis	VB	16	6	12			59 d	Yes

Pc gradient, portocaval pressure gradient; d, final shunt diameter; m, months; d, days; liver Tx, liver transplantation following TIPS; BA, biliary atresia; bleeding from EVB, esophageal varices; EGVB, esophageal and gastric varices; GVB, gastric varices; SVB, stomal varices; VB, varices, not specified; IVB, intestinal varices; HSP, hypersplenism; n.r., not reported; HE, hepatic encephalopathy.

[24–26], but up to 27% in cirrhotic patients [24] with time intervals to clinical manifestation reaching 20 years [27]. Therefore long-term follow-up after TIPS in children is necessary to estimate the real risk of encephalopathy.

Recurrent variceal bleeding was the main indication for TIPS insertion in our patients. Including reinterventions, this treatment was sufficient to control bleeding and to obviate further endoscopic therapy in all patients. Ascites was an additional indication in one patient. It was markedly reduced after TIPS insertion, relapsed because of restenosis and resolved again after shunt revision. Hypersplenism was not a primary indication for TIPS in our patients, but nevertheless it was our intention to improve associated findings. Peripheral platelet counts increased in four patients but decreased in five during long-term follow-up (mean factor of increase 1.03). The difference was not significant. In another series of 12 children the increase in peripheral platelet counts after TIPS was 1.2-fold [10]. Compared with these results, splenic artery embolization is more effective, causing a 2- to 3-fold increase in platelets [28, 29], which is nearly the level after splenectomy (3- to 4-fold increase). Splenomegaly did not regress in our patients after TIPS, similar to the results after surgical shunting. The results of this limited experience and reports in the literature suggest that TIPS is not a treatment modality to substantially improve hypersplenism in children.

Persistent and recurrent variceal bleeding refractory to conservative and endoscopic treatments was also the most important indication for TIPS in patients reported in the literature (Table 5). Besides bleeding from esophageal and gastric varices, hemorrhage from stomal varices was reported in three of 36 patients (8%) and from intestinal varices in one of 36 patients (3%). Bleeding was controlled by TIPS in these four patients. In our series four children (44%) had bleeding from intestinal varices, with a prominent indication for TIPS in three of them. All patients in our series had previously undergone HPE, which is probably a predisposing factor for the development of varices at jejuno-jejunostomy.

In a considerable number of children treated by TIPS, portal decompression served as a successful bridging therapy to liver transplantation. Twenty of 34 patients (59%) underwent liver transplantation following TIPS. No case has yet been reported of failed transplantation after TIPS in children. In our study eight of nine patients were active candidates for liver transplantation prior to TIPS. Four patients underwent transplantation, but in five their general condition improved considerably after TIPS, such that they were no longer active candidates for transplantation.

Because this study did not follow a defined prospective protocol, conclusions have to be drawn with caution. Considering the excellent long-term results of surgical portosystemic shunts, good palliation effects of endoscopic treatment of varices and the limited experience in terms of TIPS in pediatric patients, including those with BA, the indication for this treatment needs to be defined carefully. The results of this series support the conclusion that in children suffering from massive variceal bleeding resistant to medical and endoscopic treatment TIPS should be considered as a valuable option for bridging to liver transplantation. However, it has to be borne in mind that the procedures are more difficult, especially in children with BA and advanced periportal fibrosis, and that the frequency of reinterventions seems to be higher compared with adults.

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