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REVIEW

Transjugular intrahepatic portosystemic shunt in the treatment of Budd–Chiari syndrome: a critical review of literatures

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Abstract

An increasing number of patients with Budd–Chiari syndrome (BCS) have undergone transjugular intrahepatic portosystemic shunt (TIPS). However, the critical role of TIPS in the treatment of BCS has not been systematically reviewed. The authors identified all relevant literatures via the PubMed, EMBASE and Cochrane library databases. Overall, 160 papers from 29 countries reported the application of TIPS for BCS. The number of publications was increased over time, but the level of evidence in this field was low. Common indications for TIPS in BCS patients included refractory ascites, recurrent variceal bleeding, diffuse hepatic vein thrombosis and progressive liver failure. Successful TIPS insertion could improve the hemodynamic and clinical parameters. TIPS procedure-related complications were not infrequent (range: 0–56%), but procedure-related death was rare. Shunt dysfunction rate appeared to be higher (range: 18–100%). Compared with bare stents, covered stents could significantly decrease the rate of shunt dysfunction. Hepatic encephalopathy rate after TIPS was relatively low (range: 0–25%). Short- and long-term prognosis of BCS-TIPS patients was excellent with 1-year cumulative survival rate of 80–100% and 5-year cumulative survival rate of 74–78%. In conclusions, existing literatures supported the feasibility, safety and efficacy of TIPS in the treatment of BCS. Prospective cohort studies or randomized controlled trials were difficult due to the rarity of BCS, but might be very necessary to precisely identify the timing of transition from medical therapy and/or percutaneous recanalization to TIPS insertion and the real candidates in whom early TIPS should be promptly employed with no need of any prior therapy.

Key Words: *Budd–Chiari syndrome, review, transjugular intrahepatic portosystemic shunt, treatment*

Introduction

For more than two decades, transjugular intrahepatic portosystemic shunt (TIPS) has been successfully used for the prophylaxis and treatment of portal hypertension complications [1,2]. The classical indications for TIPS have been justified by several randomized controlled trials and meta-analyses [3,4], including the secondary prophylaxis of variceal bleeding unresponsive to medical and endoscopic therapy and the treatment of refractory ascites requiring repeated large-volume paracentesis [1,2]. A few randomized controlled trials have also confirmed the superiority of TIPS in the secondary prophylaxis of gastric variceal bleeding and the first-line treatment of acute variceal bleeding in high-risk cirrhotic patients [5–7]. Other emerging indications are supported by scattered case series, including the treatment of refractory hepatic hydrothorax [8], hepatorenal syndrome [9], portal vein thrombosis [10], Budd–Chiari syndrome (BCS) [11], etc.

BCS is a rare vascular disease of the liver characterized by the hepatic venous outflow obstruction from the small hepatic veins (HVs) to the suprahepatic inferior vena cava (IVC) [12]. The incidence of BCS is estimated to be 0.13–0.36 per million per

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year [13,14]. In the current American Association for the Study of Liver Diseases guideline and Baveno V consensus [15,16], a stepwise therapeutic strategy has been recommended as follows: 1) anticoagulation therapy should be promptly initiated to all BCS patients in the absence of contraindications; 2) local thrombolysis therapy is appropriate for acute/recent HV thrombosis; 3) percutaneous transluminal angioplasty with or without stenting can restore hepatic blood flow in patients with segmental HV or IVC obstruction; 4) TIPS should be indicated if angioplasty/stenting is not technically feasible, or severe portal hypertension complications or persistently deteriorated liver function develops and 5) liver transplantation is the ultimate option of severe BCS unresponsive to TIPS. Several large case series have further shown a high proportion of BCS patients requiring TIPS insertion [17,18]. Given that TIPS is the mainstay treatment modality of BCS, the authors systematically review the quantity and quality of scientific publications, technique, indications, efficacy, complications and prognosis of TIPS in the setting of BCS.

Methods

Search strategy and selection criteria

QX searched PubMed, EMBASE and Cochrane library databases from the inception to 20 October 2012 for this systematic review. Search items were as follows: (Budd–Chiari syndrome (all fields)) or (hepatic vein thrombosis (all fields)) or (hepatic venous thrombosis (all fields)) or (hepatic vein outflow obstruction (all fields)) or (hepatic venous outflow obstruction (all fields)) and (transjugular intrahepatic portosystemic shunt (all fields)) or (TIPS (all fields)) or (TIPSS (all fields)). Also, QX has done a manual search through reference list of published literatures, major Chinese-language interventional radiology journals and abstracts of major Gastroenterology, Hepatology, and Interventional Radiology meetings for supplementation. On the basis of the title and abstract, QX and YM excluded reviews, comments, editorials, interviews, basic studies and studies with non-primary BCS patients. In cases of uncertainty, QX and YM further read the full text. QX and YM included all publications regarding the BCS patients treated with TIPS. Sample size, publication language, publication status and publication date were not restricted.

Data extraction

QX obtained the full texts of all included papers for data extraction. If the full text of one paper was not available, QX would contact with the corresponding author. Non-English and non-Chinese full texts would be translated by Google. QX and YM extracted the detailed information for all included papers (i.e. authors, publication journal, publication date, countries, provinces and/or cities, institutions, retrospective or prospective studies, number of patients, gender proportion, age range, enrollment period, etiology of BCS, indications for TIPS, type of obstruction, bare or covered stents, technical approaches, TIPS procedure-related complications, TIPS success rate, preoperative and postoperative hemodynamic change, follow-up periods, TIPS dysfunction rate, postoperative hepatic encephalopathy rate, death rate, causes of death and prognostic factors) into an Excel table.

Data analysis

The number of publications was counted according to the published years, countries, type of papers and nature of studies. A line chart was drawn to show the trend in number of publications over time, and a bar chart was drawn to show the distribution of number of publications in different countries. These analyses were performed using the SPSS 12.0 software (SPSS Inc., Chicago, IL, USA). Meta-analyses were also performed to quantify the effect of TIPS on ascites, portosystemic pressure gradient, portal vein flow velocity, bilirubin, and creatinine in BCS patients, and the effect of covered stents on improvement of shunt dysfunction (see Appendix).

Quantity and quality of publications

A total of 507 papers were identified (311 in PubMed database, 166 in EMBASE database, 21 in Cochrane library database and 9 in manual search). Among them, 160 papers met the eligibility criteria (Figure 1). Generally, these papers were done from 29 countries, suggesting the worldwide popularity of TIPS technique in BCS (Figure 2). But only one paper was from African country. In addition, an increasing trend in the number of publications over time indicated a growing awareness of TIPS in treatment of BCS (Figure 3). In details, 19 case series described the application of TIPS in the treatment of portal hypertension due to various etiologies including BCS and others, 34 case series described the outcome of BCS patients treated with various treatment modalities including TIPS and others and 107 papers focused on the outcome of BCS treated with TIPS alone. Of the 107 papers, 90 were case reports (number of BCS-TIPS patients <10) and 17 were retrospective case series (number of BCS-TIPS patients ≥10) (Table I) [11,19–34]. No randomized controlled trial was

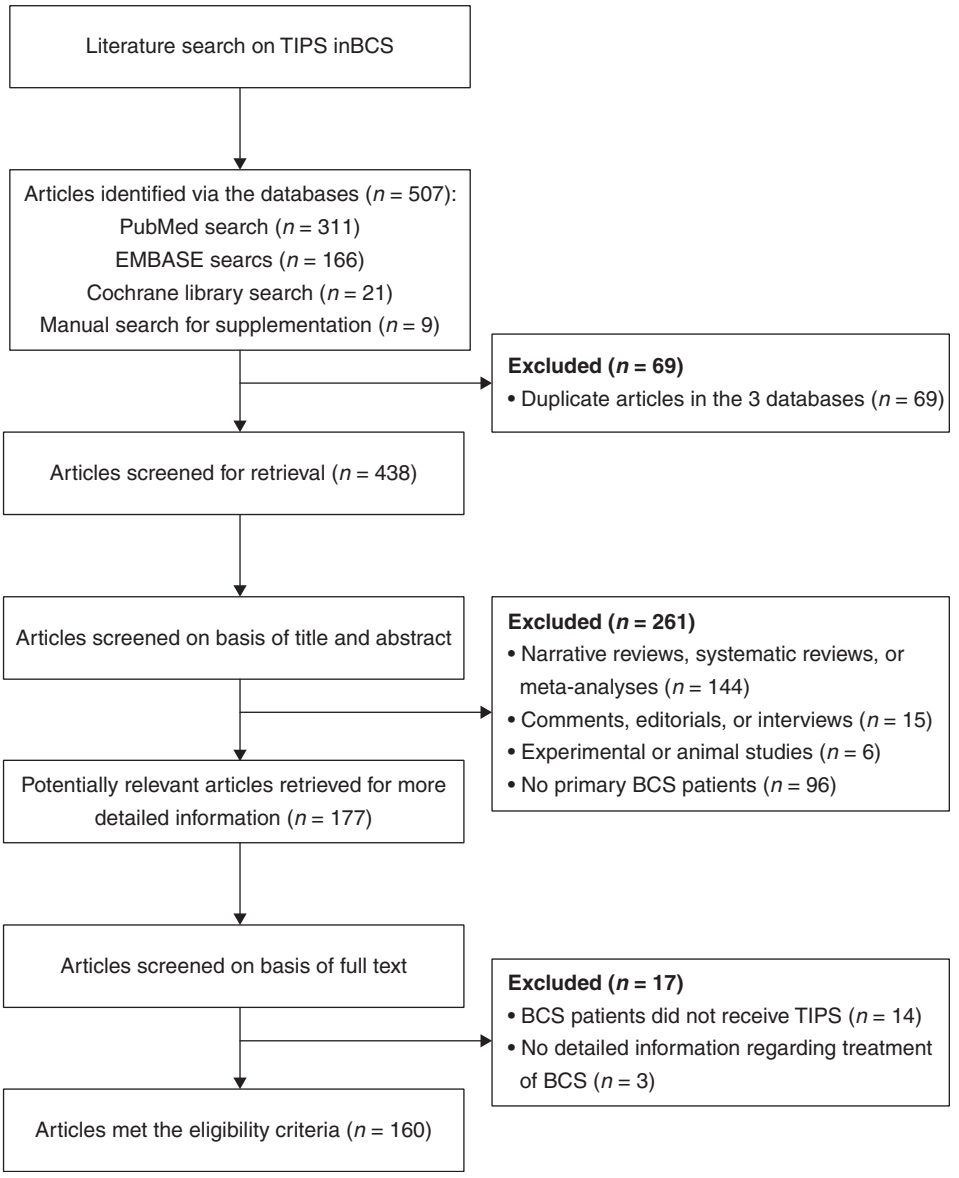

Figure 1. Flow chart of study selection.

performed, suggesting that the level of evidence was low in this field.

Techniques

The technical approach of a traditional TIPS procedure is from a patent right HV to puncture an intrahepatic right or left portal vein branch [2]. But this technical approach has to be restrained by occlusion of HVs in BCS patients [35–37]. Thus, an intrahepatic portosystemic shunt is often created via the HV remnant/stump or directly via the IVC in BCS patients. TIPS procedure by the latter technical approach is also called as "DIPS (direct intrahepatic portocaval shunt)" [38]" or "modified TIPS" [24]". To further overcome the technical difficulty of gaining the access to the portal vein, some interventional radiologists also use a reverse approach, in which percutaneous puncture of the portal vein under real-time sonographic guidance is immediately followed by puncture of the IVC [19,38,39]. Generally, TIPS success rate in these patients is high, ranging from 91% to 100% in 17 papers. Major causes of technical failure are the impossibility to puncture the liver parenchyma [26,30] and the inability to directly cannulate the occluded HV or IVC [20].

In the situations of HV occlusion combined with IVC occlusion or compression, TIPS procedure is incidentally planned after percutaneous IVC recanalization. In cases where percutaneous transluminal angioplasty can successfully dilate IVC occlusion or compression, TIPS procedure would not be

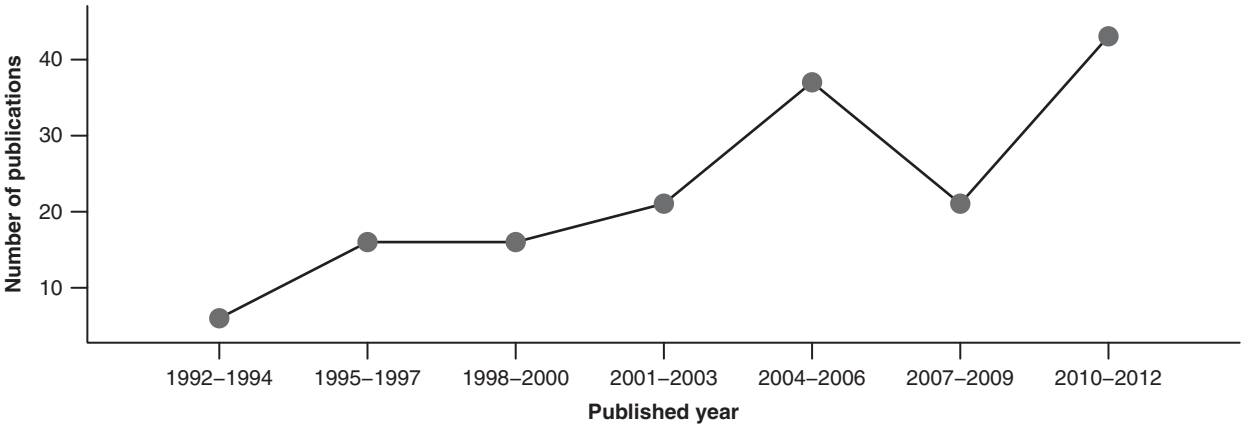

Figure 2. Number of publication according to the country.

complicated [40]. If a metallic stent is placed into the supra-hepatic portion of IVC and does not cross the HV ostium, a TIPS stent could be placed in parallel with the IVC stent [41–43]. However, if an IVC stent is necessarily placed across the HV ostium, the technical difficulty of a next TIPS insertion might be greatly increased. One case report in detail described how to place a TIPS through the strut of a previous IVC stent [44]. Because the combined HV and IVC obstruction is the most common type of BCS in China [45,46], this technique has been adopted by the authors' team (Figure 4).

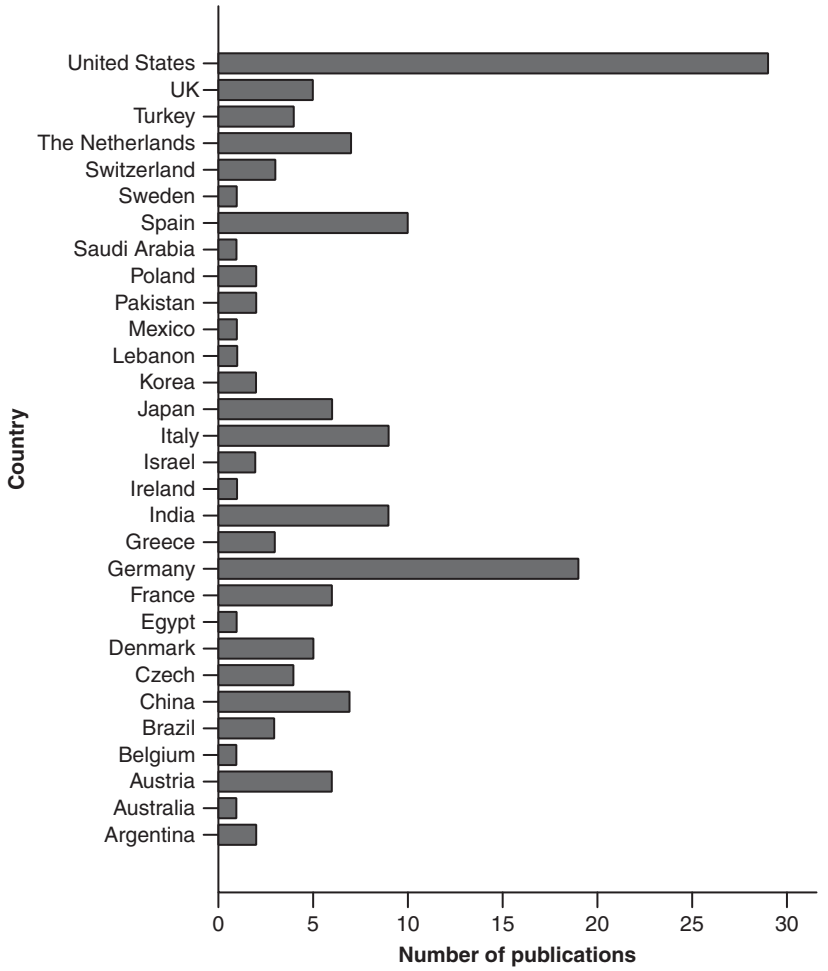

Figure 3. Number of publication according to the publication year.

Table I. An overview of 17 papers focusing on the outcome of BCS-TIPS patients.

Authors, journal (Year)	Source	Period of enrollment	No. BCS	Sex (M/F)	Age	Etiology of BCS	TIPS success rate
Blum et al. *Radiology* (1995) [28], retrospective	University Hospital Freiburg, Freiburg, Germany	1991.12–1995.1	12	6/6	Mean (range): 52 (31–71) years	PV ($n = 4$); PC deficiency ($n = 3$); PNH ($n = 1$); breast carcinoma ($n = 1$); malignant melanoma ($n = 1$); unknown ($n = 1$)	100% (12/12)
Boyvat et al. *AJR* (2008) [19], retrospective	Baskent University, Ankara, Turkey	2003.9–2006.10	11	5/6	Mean (range): 26 (6–43) years	Postpartum thrombophilia ($n = 2$); PS and PC deficiency ($n = 2$); FVL mutation ($n = 2$); PS deficiency alone ($n = 1$); unknown ($n = 4$)	100% (11/11)
Corso et al. *Radiol Med* (2008) [27], retrospective	Niguarda Hospital, Milan, Italy	1999.1–2006.12	15	7/8	Median (range): 31 (7–52) years	Myelofibrosis ($n = 4$); PV ($n = 1$); PNH ($n = 1$); FVL mutation ($n = 1$); anticardiolipin antibodies ($n = 1$); unknown ($n = 7$)	100% (15/15)
Darwish Murad et al. *Liver Int* (2008) [26], retrospective	University Medical Center, Rotterdam, the Netherlands	1994.1–2006.3	16*	6/10	Median (range): 31 (19–50) years	MPN ($n = 4$); APS ($n = 5$); thrombogenic disorders ($n = 3$); oral contraceptive ($n = 6$); BD ($n = 1$); unknown ($n = 1$)	94% (16/17)
Gandini et al. *Radiology* (2006) [29], retrospective	University of Tor Vergata, Rome, Italy	1994.1–2003.11	13	4/9	Mean (range): 35.7 (17–65) years	PV ($n = 5$); PNH ($n = 2$); osteomyelosclerosis ($n = 2$); AT deficiency ($n = 1$); PS deficiency ($n = 1$); unknown ($n = 2$)	100% (13/13)
Garcia-Pagan et al. *Gastroenterology* (2008) [11], retrospective	Six European centers	1993.7–2006.3	124#	46/78	Mean (95% CI): 38 (35–40) years	MPN ($n = 64$); PNH ($n = 13$); APS ($n = 15$); prothrombotic disorders ($n = 2$); BD ($n = 1$); MDS ($n = 1$)	93% (124/133)
Han et al. *J Intervent Radiol* (2008) [32], article in Chinese; retrospective	Xijing Hospital of Digestive Diseases, Fourth Military Medical University, Xi'an, China	2001.11–2007.3	14	10/4	Mean (range): 32 (18–49) years	NA	100% (14/14)
Hernandez-Guerra et al. *Hepatology* (2004) [23], retrospective	Hospital Clinic, Barcelona, Spain; Queen Elizabeth Hospital, Birmingham, UK	1993.1–2003.4 (Spain); 2001.1–2003.4 (UK)	21 (15, Spain; 6, UK)	6/15	Mean (range): 40 (17–54) years	MPN ($n = 10$); PNH ($n = 2$); coagulopathy disorders ($n = 6$); unknown ($n = 3$)	100% (21/21)
Mancuso et al. *J Hepatol* (2003) [25], retrospective	Royal Free Hospital, London, UK	1995.4–2002.2	15	8/7	Mean (range): 39.5 (20–73) years	MPN ($n = 5$); PNH ($n = 3$); APS ($n = 1$); PC deficiency ($n = 1$); hepatic metastases from rectal adenocarcinoma ($n = 1$); oral contraceptive ($n = 1$); unknown ($n = 3$)	93% (14/15)

Table I. (*Continued*).

Authors, journal (Year)	Source	Period of enrollment	No. BCS	Sex (M/F)	Age	Etiology of BCS	TIPS success rate
Molmenti et al. *Ann Surg* (2005) [20], retrospective	Johns Hopkins University School of Medicine, Baltimore, MD, USA	1993.1–2003.1	11	5/6	Median (range): 64 (22–78) years	PV (n = 4); PNH (n = 3); unknown (n = 5)	91% (10/11)
Rangarajan et al. *Cardiovasc Intervent Radiol* (2011) & *J Vasc Interv Radiol* (2012) [33,34], abstract; retrospective	University Hospital Birmingham, Birmingham, UK	1996.9–2009.10	46	NA	NA	NA	NA
Rossle et al. *Surgery* (2004) [30], retrospective	University of Freiburg, Freiburg, Germany	1991–2001	35	8/27	Mean ± SD (range): 43 ± 16 (12–74) years	MDS (n = 21); PNH (n = 2); plasmatic thrombophilia (n = 4); tuberculous peritonitis (n = 1); unknown (n = 7)	94% (33/35)
Safka et al. *Cas Lek Cesk* (2005) [22], retrospective	Ustav fyziologie LF UK, Hradec Kralove, Czech Republic	12 years	23	4/19	Median (range): 33.3 (13–75) years	MPN (n = 14); a defect of coagulation (n = 1); unknown (n = 7)	100% (23/23)
Wu et al. *Int J Clin Pract* (2010); Wu et al. *Zhonghua Wai Ke Za Zhi* (2006) [24,31], retrospective	Jinling Hospital, Jiangsu, China	2003–2007	11	4/7	Mean ± SD: 40.82 ± 11.45 years	NA	100% (11/11)
Zahn et al. *BMC Gastroenterol* (2010) [21], retrospective	University Hospital Heidelberg, Heidelberg, Germany	1998–2008	13	3/10	Mean ± SD (range): 36 ± 13 (20–60) years	PV (n = 3); FVL mutation and PC deficiency (n = 1); prothrombin mutation (n = 1); APS and APC resistance (n = 1); PC and AT deficiency (n = 1); APC resistance and AT deficiency (n = 1); estrogen (n = 1); unknown (n = 4)	100% (13/13)

Abbreviations: APS = antiphospholipid syndrome; AT = antithrombin; BCS = Budd–Chiari syndrome; BD = Behcet's disease; CI: confidence interval; FVL = factor V Leiden; ITP = idiopathic thrombocytopenic purpura; MDS = myelodisplastic syndrome; MPN = myeloproliferative neoplasms; NA = not available; PC = protein C; PNH = paroxysmal nocturnal hemoglobinuria; PS = protein S; PTFE = plytetrafluoroethylene; PV = polycythemia vera; SD = standard deviation; TIPS = transjugular intrahepatic portosystemic shunt.
*16 patients received successful TIPS insertions, and 17 attempted TIPS.
#124 patients received successful TIPS insertions, and 133 attempted TIPS.

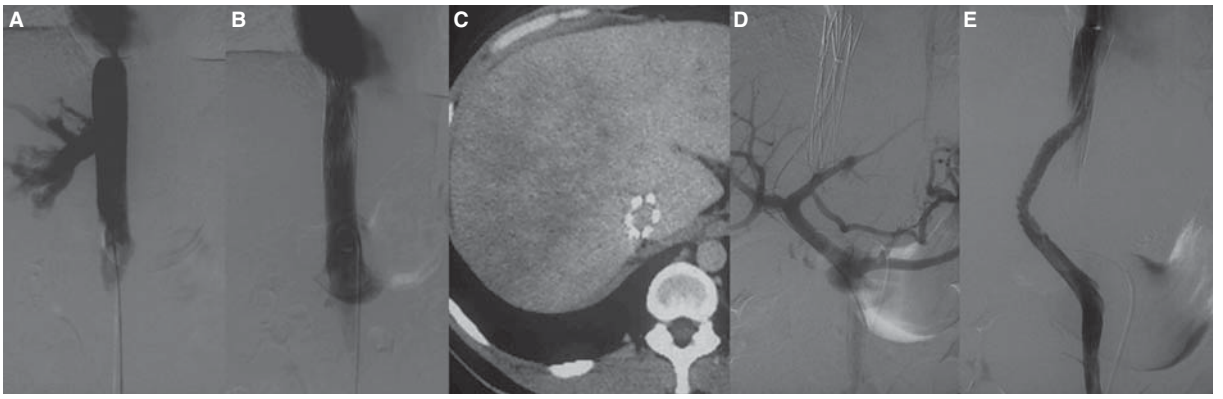

Figure 4. Creation of a transjugular intrahepatic portosystemic shunt (TIPS) after inferior vena cava (IVC) stent placement in a patient with Budd–Chiari syndrome. An IVC segmental obstruction was revealed by IVC angiography (A). Although percutaneous transluminal angioplasty and stent placement were performed (B), the patient still presented with progressive liver dysfunction and persistent liver congestion, as revealed by IVC angiography (C). Thus, a TIPS was inserted via the strut of the IVC stent (D and E).

Application of TIPS in cases of BCS and concomitant portal venous system thrombosis represents another technical challenge [47–55]. Percutaneous transhepatic, transsplenic and transmesenteric approaches can facilitate the access to the portal vein [10]. Methods used to recanalize the thrombosed portal vein include: 1) local or systemic thrombolysis during the perioperative period [47–49,52,55]; 2) mechanical disruption of portal vein thrombus by an angiographic pigtail catheter during the intraoperative period [51,53] and 3) deployment of a stent over the main portal vein and superior mesenteric vein thrombus [50,54].

Indications

The rarity of BCS leads to the difficulty in precisely identifying the candidates for TIPS by randomized controlled trials. The two common indications established for cirrhotic patients with portal hypertension (i.e. refractory ascites and recurrent variceal bleeding) appear to be extrapolated to BCS patients. Diffuse thrombosis of HVs is also a reasonable indication for TIPS, because percutaneous transluminal angioplasty with or without stent placement is often difficult to maintain the long-term HV patency. But TIPS might be unnecessary, if the clinical symptoms are lacking or mild and the compensation of intrahepatic collateral vessels is adequate [56]. In addition, TIPS should be promptly employed in cases of progressive liver failure, if medical therapy and/or percutaneous recanalization cannot fully control the disease progression [57].

Efficacy

Hemodynamic improvement

Eleven case series reported the detailed information regarding the hemodynamic parameters before and after TIPS insertions in BCS patients (Supplementary Table 1). As expected, successful TIPS insertions could decrease the portosystemic pressure gradient and increase the portal vein (PV) flow velocity and blood flow in BCS patients.

Clinical improvement

Eight case series reported the detailed information regarding the clinical parameters (i.e. frequency of ascites, albumin, bilirubin, alanin aminotransferase, asparate aminotransferase, gamma-glutamyl transferase, sodium, prothrombin time, creatinine and Child–Pugh class or score) before and after TIPS insertions in BCS patients (Supplementary Table 2). Generally, a trend in the improvement of all these clinical parameters could be observed.

Complications

Procedure-related complications

The rate of procedure-related complications is various in 16 case series, ranging from 0% to 56%. Procedure-related complications mainly include liver capsule perforation, IVC and portal vein injury, contrast materials induced acute renal failure, stent migration, etc. (Table II). Due to extensive congestion and enlargement of the liver in BCS patients, a longer hepatic parenchymal tract needs to be established during a TIPS procedure. Accordingly, reports of intrahepatic hematoma, capsular puncture and portal vein or IVC injury complicating TIPS for BCS are not infrequent [41,58–61]. Fortunately, most of patients experiencing these procedure-related complications can be cured by conservative treatment, such as close observation, dose reduction

Table II. TIPS procedure-related complications in BCS-TIPS patients in case series*.

Author (Year)	No. BCS patients attempting TIPS	TIPS procedure-related complications	Procedural complications % Procedural death %
Amarapurkar et al. (2008) [67]	15	Peritoneal hemorrhage and death (n = 1)	7% (1/15) 7% (1/15)
Attwell et al. (2004) [65]	17	Renal arterial hemorrhage, renal failure, sepsis and death (n = 1); splenic vein rupture and death (n = 1); transient hypoxia and hypotension believed to be sedation-related (n = 1); transient intraperitoneal bleeding (n = 1)	12% (2/17) 24% (4/17)
Blum et al. (1995) [28]	12	None	0% (0/12) 0% (0/12)
Boyvat et al. (2008) [19]	11	Immediate increase in preload and transient right heart failure (n = 1)	0% (0/11) 0% (0/11)
Corso et al. (2008) [27]	15	None	0% (0/15) 0% (0/15)
Darwish Murad et al. (2008) [26]	17	Liver capsule perforation (n = 3); biliary duct puncture (n = 1); bleeding complication at the site of TIPS (n = 1); stent migration (n = 1)	35% (6/17) 0% (0/17)
Eapen et al. (2006) [68]	29	Contrast materials induced acute renal failure and death (n = 1)	3% (1/29) 3% (1/29)
Eldorry et al. (2011) [64]	16#	Intraperitoneal bleeding or hemobilia (n = 4); stent migration (n = 1)	31% (5/16) 0% (0/16)
Garcia-Pagan et al. (2008) [11]	133	IVC injury, intraperitoneal bleeding, hemothorax and death (n = 1); partial occlusion of the IVC by the stent, persisting of ascites, infection and death (n = 1); subcapsular hematoma (n = 5); hemoperitoneum without hemodynamic instability (n = 3); hemobilia (n = 3); reversible heart failure (n = 3); procedure-associated infection (n = 2); supraventricular tachycardia (n = 1); jugular vein thrombosis (n = 1); IVC compression (n = 1); mild hemolysis (n = 1)	17% (22/133) 2% (2/133)
Hernandez-Guerra et al. (2004) [23]	21	None	0% (0/21) 0% (0/21)
Mancuso et al. (2003) [25]	15	Portal vein rupture and death (n = 1); intrahepatic hemorrhage complicated by liver rupture and death (n = 1); intrahepatic hematoma (n = 1)	20% (3/15) 13% (2/15)
Molmenti et al. (2005) [20]	11	None	0% (0/11) 0% (0/11)
Perello et al. (2002) [69]	13	None	0% (0/13) 0% (0/13)
Plessier et al. (2006) [18]	25	IVC injury and death (n = 1); hemobilia (n = 3); liver hematoma (n = 3); reversible pulmonary edema (n = 3); transient hepatic encephalopathy (n = 3)	56% (13/25) 4% (1/25)
Rossle et al. (2004) [30]	35	Arterio-stent fistulae (n = 3); intrahepatic hematoma (n = 2); acute renal failure (n = 1); contrast dye induced thyrotoxicosis (n = 1)	20% (7/35) 0% (0/35)
Wu et al. (2010) [24]	11	Bleeding in the portal bifurcation (n = 1)	9% (1/11) 0% (0/11)

Abbreviations: BCS = Budd–Chiari syndrome; IVC = inferior vena cava; TIPS = transjugular intrahepatic portosystemic shunt.
*Case series represent the studies in which more than 10 patients were included.
#A total of 16 TIPS procedures were performed in 15 patients, including TIPS as primary treatment in 13 patients, TIPS after angioplasty with or without stenting in 2 patients and re-TIPS after the first TIPS in 1 patient.

or discontinuation of anticoagulants and blood transfusion [41,58–61]. Only a minority of patients die of them. In addition, it should be noted that intrahepatic hematoma is not shortly observed, but 10–15 days after TIPS [41,58,59,61]. This phenomenon may be attributed to excessive use of anticoagulation immediately after multiple needle passes of liver parenchyma [41,61].

Shunt dysfunction

Shunt dysfunction appears to be more frequent in BCS-TIPS patients due to their prothrombotic states. The rate of shunt dysfunction varies from 18% to 100% in 14 case series, depending on the type of stents, number of patients and length of follow-up period. Nine case series reported the shunt dysfunction or primary

Table III. Prognosis of BCS-TIPS patients in case series*.

Authors (Year)	No. of patients	Follow-up period	Mortality rate (%)	Cumulative survival rate	Causes of death
Amarapurkar et al. (2008) [67]	15	NA	13	NA	TIPS-related complication (n = 1); persistent HE (n = 1)
Attwell et al. (2004) [65]	17	Mean (range): 3 (0.1–7) years	24	NA	TIPS-related complications (n = 2); portal vein thrombosis with hepatorenal syndrome (n = 1); decompensated liver disease and renal failure associated with TIPS occlusion (n = 1)
Blum et al. (1995) [28]	12	Median (range): 13 (0.1–36) months	25	NA	Progressive liver failure (n = 2); cardiac failure due to secondary hematochromatosis caused by multiple blood transfusions (n = 1)
Boyvat et al. (2008) [19]	11	Median (range): 17 (8–41) months	0	1-year survival rate: 100%	None
Corso et al. (2008) [27]	15	Median (range): 29.4 (3.2–68) months	13	NA	Fulminant acute leukemia (n = 1); liver failure due to portal vein and TIPS reocclusion (n = 1)
Darwish Murad et al. (2008) [26]	16	Median (range): 29 (1–110) months	25	1- and 3- year cumulative survival rates: 80% and 72%	Variceal bleeding (n = 1); MOF (n = 1); liver failure secondary to sepsis (n = 1); epithelioid ovarian sarcoma (n = 1)
Eapen CE, et al. (2006)	29	Median (range): 35 (0–93) months	17	NA	Massive cerebral infarct, severe hemolysis, renal failure (n = 1); culture negative neutrocytic ascites, septicemia, acute renal failure, worse after contrast (n = 1); subdural hematoma (n = 2); worsening HE (n = 1)
Eldorry et al. (2011) [64]	13	NA	0	1-year survival rate: 100%	None
Gandini et al. (2006) [29]	13	Mean ± SD: 22.7 ± 10.53 months in bare stent; 23.1 ± 7.81 months in covered stent	7.7	NA	Liver insufficiency (n = 1)
Garcia-Pagan et al. (2008) [11]	133	Mean (range): 36.7 (0.7–156) months	12	1-, 5- and 10-year LT-free survival rates: 88%, 78% and 69%, respectively	Liver failure (n = 4); hematological disorders (n = 4); sepsis (n = 3); TIPS-related complications (n = 2); stroke (n = 2); upper gastrointestinal bleeding of unknown origin (n = 1)
Gronbaek et al. (2011) [70]	14	Median (range): 50 (15–117) months	7	NA	NA
Hernandez-Guerra et al. (2004) [23]	21	Median (range): 21.2 (3.9–124.8) months in bare stent; 19.1 (7.7–31.3) months in covered stent	0	NA	None
Mancuso et al. (2003) [25]	15	Mean (range): 24 (8–44) months	36	NA	TIPS-related complications (n = 2); liver failure (n = 1); pulmonary edema (n = 1); metastatic disease (n = 1)
Molmenti et al. (2005) [20]	11	NA (2 lost to follow-up, 1 died, 7 available during follow-up)	18	NA	Sepsis unrelated to TIPS procedure (n = 1); surgical complications related to LT (n = 1)

Table III. (*Continued*).

Authors (Year)	No. of patients	Follow-up period	Mortality rate (%)	Cumulative survival rate	Causes of death
Perello et al. (2002) [69]	13	Mean ± SD (range): 4 ± 3 (0.2–9.1) years	8	NA	MOF (n = 1)
Plessier et al. (2006) [18]	25	Median (IQR): 32 (1–49) months in bare stents; 10 (1–21) months in covered stents	8	NA	TIPS-related complications after technical failure (n = 2)
Rangarajan et al. [33,34], abstract	46	NA	NA	Overall survival rate at 1, 5 and 10 years was 89%, 78% and 71%, respectively	NA
Rossle et al. (2004) [30]	35	Mean ± SD: 37 ± 29 months	9	1- and 5-year cumulative survival without LT: 93% and 74%	Septicemia (n = 1); acute heart failure (n = 1); unknown (n = 1)
Safka et al. (2005) [22]	23	NA	26	NA	Fulminate liver failure (n = 1); liver failure caused by acute shunt occlusion (n = 2); progression of the underlying hematooncological disease (n = 1); unknown (n = 2)
Wu et al. (2010) [24]	11	Mean ± SD: 62.55 ± 42.76 months	9	NA	Deteriorated liver function in acute HV occlusion (n = 1)
Zahn et al. (2010) [21]	13	Median (range): 4 (0.5–12) years	8	NA	Underlying hematologic disease (n = 1)

Abbreviations: BCS = Budd–Chiari syndrome; HE = hepatic encephalopathy; HV: hepatic vein; IQR = interquartile range; LT = liver transplantation; MOF = multiorgan failure; NA = not available; SD = standard deviation; TIPS = transjugular intrahepatic portosystemic shunt.
*Case series refer to the studies with more than 10 BCS-TIPS patients.

patency rate in BCS patients receiving bare and covered stents (Supplementary Table 3). Generally, shunt dysfunction is less frequent in patients receiving covered stents than in those receiving bare stents.

Hepatic encephalopathy

Hepatic encephalopathy after TIPS is uncommon in BCS patients. The rate of hepatic encephalopathy varies from 0% to 25%, primarily depending on the presence of preoperative hepatic encephalopathy and liver dysfunction. One case series reported that hepatic encephalopathy developed in 3 of 10 patients receiving covered stents and none of 5 patients receiving bare stents. But another case series reported that no hepatic encephalopathy developed in any patients receiving bare and covered stents.

Prognosis

Short- and long-term prognosis of BCS-TIPS patients is excellent. Overall mortality varies from 0% to 36% in 20 case series (Table III). The seeming heterogeneity among studies is primarily attributed to the severity of HV occlusion, degree of liver dysfunction, inclusion criteria (i.e. malignancy, other severe concomitant diseases or not), enrollment period and sample sizes. Cumulative survival rate is relatively homogeneous among studies (1-year cumulative survival rate: 80–100% in 6 case series; 5-year cumulative survival rate: 74–78% in 3 case series; 10-year cumulative survival rate: 69–71% in 2 case series).

Common causes of death include liver failure and/or multiorgan failure, progression of underlying hematological diseases and TIPS procedure-related complications. Liver failure is mainly due to the persistent deterioration of liver function itself or secondary to acute or irreversible TIPS occlusion. But it should be noted that only a minority of patients with failed recanalization of shunt occlusion or acute shunt occlusion died [22,27]. The formation of adequate intrahepatic collateral vessels [23,62–64] and timely implementation of alternative therapies (i.e. surgical shunt and/or liver transplantation) [11,20,29,30,65,66] greatly improve these patients' survival.

Given small sample size in most of studies, the prognostic analysis of BCS-TIPS patients was just performed in one large multicenter study [11] and one single-center study [33,34]. In the former study, a multivariate Cox regression analysis identified total bilirubin, age and international normalized ratio (INR) as the independent predictors of 1-year transplant-free survival. Further, a BCS-TIPS prognostic index model was proposed: age (years) $\times$ 0.08 + bilirubin (mg/dL) $\times$ 0.16 + INR $\times$ 0.63. The most specific discriminative value of the model was 7 points with a sensitivity of 58%, a specificity of 99%, a positive predictive value of 88% and a negative predictive value of 96%. In the latter study, age was an independent predictor of death ($p = 0.016$). However, age was not significant ($p = 0.45$), when a combined Cox hazard analysis was performed for all variables.

Conclusions

This systematic review of current literatures emphasizes the following: 1) although the level of evidence is low at present, the critical role of TIPS in the treatment of BCS has been increasingly recognized worldwide; 2) TIPS is technically feasible and safe in a majority of BCS patients; 3) successful TIPS insertion effectively lowers the portosystemic pressure gradient, resolves ascites and improves liver and renal function; 4) shunt dysfunction is relatively frequent in BCS patients, and covered stents should be recommended to decrease the rate of shunt dysfunction and 5) short- and long-term survival of BCS-TIPS patients is excellent, and total bilirubin, age and INR can predict 1-year transplant-free survival. Further prospective studies should be warranted in the following directions: 1) to evaluate how to use TIPS combined with percutaneous recanalization in the treatment of combined HV and IVC occlusion; 2) to explore the exact timing of transition from medical therapy and/or percutaneous recanalization to TIPS insertion and 3) to identify the real candidates in whom early TIPS should be directly considered the first-line treatment and in whom TIPS should be unnecessary.

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Supplementary material available online

Supplementary Tables 1–3.
Supplementary Figures 1–6.
Appendix.