Long Term Survival of Patients Undergoing TIPS in Budd-Chiari Syndrome

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Background: There has been significant improvement in understanding the etiology and management of Budd-Chiari Syndrome (BCS). Patients with chronic or acute-on-chronic BCS need radiological interventions in the form of angioplasty, hepatic vein/inferior vena cava stenting or Transjugular Intrahepatic Portosystemic Shunt (TIPS). Data regarding the long term follow up of patients undergoing TIPS is limited. We thus prospectively followed-up BCS patients who underwent TIPS at our center. **Methods:** This study included 42 patients with BCS who underwent TIPS with a covered stent between 2004 and 2014. We analyzed the etiology, symptoms, severity, laboratory parameters and imaging pre and post TIPS. All patients underwent surveillance for hepatocellular carcinoma. **Results:** Patients demographics included 26 males and 16 females with a mean age of 40.5 years (19–68 years). The mean Model for End-Stage Liver Disease score of the entire cohort was 15.38 (range: 9–25). Thirty-four patients were grouped into Rotterdam Class 2 and remaining into Class 3. There was significant improvement in ascites, gastrointestinal bleed, renal function and transaminase levels post TIPS. There were 11 deaths over the follow-up period – 4 within one month, 2 within six months and the rest after 3 years following TIPS. Median duration from clinical presentation to TIPS was 2.1 weeks and median survival till follow-up was 45.5 months (0–130 months). 33/42 patients underwent TIPS prior to 2013, and their median survival till follow-up was 55 months. Six out of eleven deaths that occurred within six months post-TIPS were before 2006; when the technique of TIPS creation was evolving. The cumulative 1 year, 5 years and 10 years OLT-free survival was 86%, 81% and 76%, respectively. Two patients underwent a liver transplant at 4 and 7 years after TIPS. **Conclusion:** Our results validate the role of TIPS in the management of patients with BCS. With the accessibility of TIPS, the requirement for liver transplantation has become rare. (J CLIN EXP HEPATOL 2019;9:56–61)

Budd Chiari Syndrome (BCS) occurs as a result of thrombotic occlusion of hepatic venous outflow. Hypercoagulable states account for majority of cases. Myeloproliferative disorders (polycythemia rubra

vera, occult disorders), hypercoagulable states (Factor V Leiden mutation, prothrombin G20210A gene mutation, antiphospholipid antibody syndrome, paroxysmal nocturnal hemoglobinuria, protein C and S deficiency, antithrombin III deficiency, methylene tetrahydrofolate reductase mutation), local compression from neoplasms and abscesses, systemic collagen vascular diseases, inflammatory bowel disease, sarcoidosis, oral contraceptive ingestion and pregnancy can cause BCS.

The fundamental pathophysiologic abnormality in BCS is an increase in sinusoidal pressure secondary to obstruction of hepatic venous outflow. The resultant hypoxic damage to the hepatocytes causes increase in the portal venous pressure. Sustained hepatic venous outflow tract obstruction causes further hepatic necrosis, eventually resulting in cirrhosis. Acute or subacute BCS may present with abdominal pain, hepatomegaly and ascites, whereas symptoms and signs of portal hypertension, bleeding varices, encephalopathy, coagulopathy, renal insufficiency and fatigue are prominent with chronic disease. Doppler ultrasonography is a practical noninvasive screening test for BCS. Hepatic Vein (HV) occlusion can be demonstrated with contrast-enhanced computed tomography and magnetic resonance imaging. Treatment is

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Abbreviations: AT III: Antithrombin III; BCS: Budd-Chiari Syndrome; CT: Computerized Tomography; CTP: Child-Turcotte-Pugh; HV: Hepatic Vein; INR: International Normalized Ratio; IVC: Inferior Vena Cava; JAK-2: Janus Kinase 2; MELD: Model for End-Stage Liver Disease; MRI: Magnetic Resonance Imaging; MTHFR: Methylene Tetrahydrofolate Reductase; OLT: Orthotopic Liver Transplantation; SD: Standard Deviation; TIPS-BCS PI score: BCS-TIPS Prognostic Index score; TIPS: Transjugular Intrahepatic Portosystemic Shunt

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based upon the underlying etiology, the extent of thrombosis and severity of liver disease. The aims of treatment are to relieve the hepatic outflow tract obstruction, to ascertain and treat the underlying cause, and to abate symptoms. Anticoagulation is the foundation of medical management. If the liver disease is progressive despite medical management, decompressive surgery and Transjugular Intrahepatic Portosystemic Shunting (TIPS) are often resorted.

Anticoagulation as a standalone therapy is less likely to achieve sufficient recanalization of occluded vessels. Porto-systemic shunt has not shown survival benefit in recent studies.^{1,2} TIPS, on the other hand has shown heartening results and survival benefit.³⁻⁸ Treatment recommendations for BCS are based on few retrospective studies^{9,10} due to dearth of prospective studies.² In this study, our objective was to evaluate the long term outcome of patients with BCS undergoing TIPS at our tertiary care center.

MATERIALS AND METHODS

The analysis was performed on prospectively maintained data by the author (DA) for all patients. All patients were evaluated and followed-up by the same author (DA). Data of all patients admitted with a primary diagnosis of BCS between 2004 and 2014 was scrutinized. All available medical records were carefully evaluated. Demographic data like age, sex and medical co-morbidities were recorded. Signs and symptoms at presentation and major events in the history including complications like gastrointestinal bleeding, ascites, jaundice, hepatic encephalopathy, hepato-renal syndrome were documented. Laboratory data, radiological imaging and procedures, surgical interventions and discharge cards were reviewed.

The date of the first investigation when the criteria for diagnosis were satisfied was considered as the date of diagnosis. BCS was defined as per the European network for vascular disorders of the liver (En-Vie) criteria and the last Baveno consensus based on imaging showing an obstructed venous outflow tract.¹¹ Doppler ultrasonography, Magnetic Resonance Imaging (MRI) or Computerized Tomography (CT) was used to diagnose BCS. All successive adult (≥ 19 years) patients who satisfied the diagnostic criteria for BCS and who successfully underwent TIPS were included in the analysis. All patients with age ≤ 19 years, those who did not consent for TIPS and those in whom the procedure of TIPS failed (nontarget TIPS insertion after multiple attempts, transgression of liver capsule with catheter) were excluded.

Hematological evaluation of hypercoagulable state was performed in all patients. This included blood work-up for Factor V Leiden mutation, Janus Kinase 2 (JAK-2) mutation, prothrombin G20210A gene mutation, antiphospholipid antibody syndrome, paroxysmal nocturnal

hemoglobinuria, protein C and S deficiency, Antithrombin III (AT III) deficiency and Methylene Tetrahydrofolate Reductase (MTHFR) mutation causing hyperhomocysteinemia. Bone marrow biopsy was performed in indicated patients. Laboratory data included complete blood count, liver profile (serum proteins, serum albumin, liver transaminases, serum bilirubin, serum alkaline phosphatase and serum gamma-glutamyl transpeptidase along with prothrombin time and INR) and renal profile (serum creatinine and blood urea nitrogen). Laboratory data was evaluated pre-TIPS, one and six months post-TIPS.

Standard set institutional protocols were followed for all the patients regarding approach, investigations and treatment. TIPS was created using standard techniques and a 10-mm-diameter covered metal stent.¹²⁻¹⁴ The portal vein was punctured directly from the Inferior Vena Cava (IVC) in absence of sufficient stump/remnant of HVs. Post-TIPS, anticoagulation was started as per guidelines.¹¹ A check TIPS-gram or TIPS Doppler were performed in all patients prior to discharge. Occurrence of gastrointestinal bleeding, ascites, hepatic encephalopathy and renal insufficiency post TIPS till last follow-up was recorded. Deaths occurring till follow-up were recorded. Such patients were censored at the time of death. If patients required a TIPS-plasty or TIPS reduction for TIPS dysfunction, a note was made in the analysis. TIPS dysfunction was defined as any of the following: significant luminal narrowing on venography, increase in portal pressure gradient above 12 mm Hg, or appearance of ascites or other complication of portal hypertension during follow-up.

Statistical Analysis

Descriptive statistics were analyzed as mean $\pm$ Standard Deviation (SD) and as range. Statistical significance was defined as a *P* value less than 0.05. Paired *t*-test was used to analyze laboratory data. The Kaplan-Meier survival analysis was used to estimate the survival function from the data. Microsoft Excel and SPSS (SPSS Inc., Chicago, IL) were used to carry out all analysis. The study protocol conformed to the guidelines of the hospital ethics committee.

RESULTS

Patient Characteristics

Total 42 patients (26 males and 16 females) were included in the analysis. The average age of the cohort was 40.52 ± 14.26 years (range 19–68 years; median 42.5 years). The baseline characteristics of the cohort are summarized in Table 1. Thirty-four patients had no comorbidities. All patients had ascites. Thirty-six patients who presented to us with BCS were not included in the analysis (13 patients were lost to follow-up; 8 patients

Table 1 Baseline Characteristics of the Cohort.

	n = 42
Males	26 (61.9%)
Females	16 (38.1%)
Age	40.52 ± 14.26 years
Co-morbidities	
Diabetes	2 (4.7%)
Pulmonary tuberculosis	2 (4.7%)
Hypertension	1 (2.3%)
Bronchial asthma	1 (2.3%)
Hypothyroidism	1 (2.3%)
Abdominal distension (ascites related)	42 (100%)
Abdominal pain	8 (19%)
Nausea and vomiting	3 (7.1%)
Icterus	22 (52.4%)
Gastrointestinal bleeding	13 (30.9%)
Renal insufficiency	5 (11.9%)
Hepatic encephalopathy	2 (4.7%)

underwent HV angioplasty, 6 patients underwent IVC stenting; 5 patients had acute BCS; 2 patients had procedural failure and 2 patients were unwilling for TIPS.

Laboratory work-up to determine the etiology of BCS was performed in all patients. A definite etiology could be determined in 24 patients. Myeloproliferative disease (especially polycythemia vera) was the commonest etiology in 17 (40.47%) patients. 3 patients tested positive for JAK-2 mutation. 5 (11.9%) patients had MTHFR mutation with resultant hyperhomocysteinemia. AT III deficiency and presence of antiphospholipid antibody syndrome was seen in one patient each. None of the patients had a combination of procoagulant disorders. All patients had

obstruction of all three HV on radiological imaging studies. 7 patients (17%) had IVC obstruction in addition to HV obstruction.

Upper gastrointestinal endoscopy revealed esophageal and gastric varices in 33 (78.57%) patients and portal hypertensive gastropathy in 7 (16.67%) patients. Laboratory values pre TIPS and one and six months post TIPS are depicted in Table 2. All patients had tested negative for hepatitis B surface antigen and anti-HCV antibody prior to TIPS. The mean MELD score of the entire cohort was 15.38 (range: 9–25). 7 patients were classified into Child-Turcotte-Pugh (CTP) Class A, 29 patients in Class B and rest of the 6 patients into Class C. Mean CTP score was 8.04 (range: 6–11). 34 (81%) patients were grouped into Rotterdam Class 2 and the rest into Rotterdam Class 3. There were 2 deaths in patients belonging to Class 3. The BCS-TIPS Prognostic Index score (TIPS-BCS PI score) has shown to predict transplant free survival in patients who receive TIPS. It is defined as: age (years) × 0.08 + bilirubin (mg/dl) × 0.16 + International Normalized Ratio (INR) × 0.63. The cutoff of 7 points had a sensitivity of 58%, a specificity of 99%, a positive predictive value of 88%, and a negative predictive value of 96% for death or OLT 1 year after TIPS.⁹ Only one patient had a score above seven and 14 (33.33%) patients had a score above five.

Treatment and Outcomes

Data was systematically evaluated at 1 month and 6 months post TIPS, and up to the last follow-up. Statistically significant improvement was observed in the total bilirubin, gamma-glutamyl transpeptidase, creatinine and total leukocyte counts pre-TIPS and post-TIPS (Table 2). Improvement was also observed in the serum transaminase values, total protein and serum albumin level. Till the

Table 2 Laboratory Values Pre TIPS and One and Three Months Post TIPS.

Laboratory value (normal range)	Pre TIPS	Post TIPS (1 month)	Post TIPS (6 months)
Total Leukocyte Count (TLC) (4000–10,000/ μ L)	16,671 ± 7024.3	7435.7 ± 2959 ($P = 0.111$)	6365.9 ± 2029 ($P = 0.001$)
Bilirubin (total) (0.2–1.3 mg/dl)	2.73 ± 2.87	2.40 ± 2.23 ($P = 0.266$)	1.75 ± 1.76 ($P = 0.007$)
Aspartate aminotransferase/SGOT (5–40 units/L)	62.88 ± 32.75	44.7 ± 23.91 ($P = 0.281$)	31.8 ± 25.05 ($P = 0.169$)
Alanine aminotransferase/SGPT (7–56 units/L)	78.6 ± 38.47	38.21 ± 17.17 ($P = 0.115$)	39.9 ± 20.11 ($P = 0.129$)
Gamma-glutamyl Transpeptidase (15–73 units/L)	72.93 ± 40.23	50.50 ± 17.52 ($P = 0.002$)	53.42 ± 30.87 ($P = 0.018$)
Serum albumin (3.5–5.0 g/dl)	3.33 ± 0.65	3.17 ± 0.66 ($P = 0.145$)	3.52 ± 0.49 ($P = 0.151$)
International Normalized Ratio (INR)	1.62 ± 0.47	2.26 (on anticoagulation) ± 0.65 ($P = 0.001$)	2.37 (on anticoagulation) ± 0.57 ($P = 0.001$)
Serum creatinine (0.8–1.5 mg/dl) Males: (0.8–1.5 mg/dl) Females: (0.7–1.2 mg/dl)	1.06 ± 0.42	0.91 ± 0.24 ($P = 0.009$)	0.91 ± 0.47 ($P = 0.098$)

last follow-up, none of the patients had recurrent gastrointestinal bleed or renal insufficiency, 8 patients had recurrent ascites post TIPS, and 3 patients had hepatic encephalopathy (Table 3). TIPS plasty was performed in 3 patients during follow-up at 6 months, 91 months and 102 months respectively for refractory ascites. TIPS dysfunction was secondary to thrombosis or pseudointimal hyperplasia, which could be managed by dilatation. TIPS diameter reduction was performed in one patient for hepatic encephalopathy within two weeks post TIPS. The patient however could not survive. Two patients developed anticoagulation related bleeding (Gangliocapsular bleed and subdural hematoma, respectively). One patient developed hepatocellular carcinoma.

There were total 11 deaths [hematologic disorder ($n = 4$); hepatic encephalopathy ($n = 3$); procedure-related death ($n = 2$; intra-abdominal bleeding in both); gastrointestinal bleeding ($n = 1$); myocardial infarction ($n = 1$)]. Out of the 11 deaths in the follow-up period, 4 patients could not survive beyond one month post TIPS. There were 2 deaths within six months and the rest (5 deaths) three years post-TIPS. A noteworthy point is that all the six out of eleven deaths that occurred within six months post-TIPS were before 2006; when the technique of TIPS creation was evolving. None of the prognostic indices

(TIPS-BCS PI score; Rotterdam class; CTP score or MELD score) could predict survival in our cohort.

Median duration from clinical presentation to TIPS procedure was 2.1 weeks (mean 6 weeks; range: 0–44 weeks) and median survival till follow-up was 45.5 months (mean 55.3 months; range: 0–130 months). 34/42 (80.95%) patients underwent TIPS prior to 2013, and their median survival till follow-up was 55 months (mean 66.13 years, range: 0–130 months). The cumulative 1 year; 5 years and 10 years OLT-free survival was 86%, 81% and 76%, respectively. Two patients underwent a liver transplant at 4 and 7 years after TIPS. Indication for transplantation was hepatocellular carcinoma in one and progressive liver failure in other. The Kaplan–Meier survival curve is depicted in Figure 1.

DISCUSSION

In this study, an assessment of the clinical course and long term outcome of patients with BCS referred to our tertiary hospital and then undergoing TIPS was done by analyzing the data of all patients admitted with a primary diagnosis of BCS between 2004 and 2014. TIPS was first used by Rösch et al. in 1969¹⁵ in dogs following which the technique was attempted in a cirrhotic patient by Colapinto in

Table 3 Complications Pre and Post-TIPS.

<i>n</i> = 42	Pre-TIPS		Post-TIPS	
	Yes	No	Yes	No
GI bleed	13 (30.95%)	29 (69.04%)	0 (0%)	42 (100%)
HE	2 (4.7%)	40 (95.3%)	3 (7.14%)	40 (92.85%)
Renal insufficiency	5 (11.9%)	37 (88.09%)	0 (0%)	42 (100%)
Ascites	42 (100%)	0 (0%)	8 (19.05%)	34 (80.95%)

Figure 1 Kaplan–Meier survival curve.

1982.¹⁶ The aim of TIPS was nonsurgical reduction in portal hypertension.

TIPS, has since then been extensively tested and has demonstrated encouraging results (control of ascites, reversal of liver failure) as demonstrated by a large series of patients published in 2008.⁹ The technique of TIPS placement may be challenging in situations of thrombosed hepatic veins and caudate lobe hypertrophy. Such patients may require lifelong anticoagulation. Covered stents have improved the long-term patency of TIPS as compared to bare metal stents used previously. TIPS has thus become the desired method of performing a portosystemic shunt. Encephalopathy rates are significantly lower when TIPS is performed for BCS as compared against other etiologies for refractory ascites.⁹

In our cohort, 15 patients (35.71%) underwent TIPS within one week of presentation. 14 patients (33.33%) underwent TIPS within one week to four weeks of presentation. 6 patients (14.28%) underwent TIPS within four week to eight weeks of presentation. Rest of 7 patients underwent TIPS beyond 8 weeks (range 12–44 weeks).

Seijo et al. looked at long term outcome of BCS with step-wise management.¹⁷ Sixty-two patients underwent TIPS (39.5%). Main indications were refractory ascites (69%), liver failure (13%), and variceal bleeding (7%). Four of these (6.45%) had rescue Orthotopic Liver Transplant (OLT) at a median of 1.8 months after TIPS. Three of these four patients died at a median of 35 months after OLT. Of the remaining 58 patients, 10 (17%) died within 6 months and 48 (83%) were alive after a median follow-up of 51 months (range, 0.3–69.0). One, 3-, and 5-year actuarial survival and OLT-free survival of patients treated with TIPS was 88%, 83%, and 72% and 85%, 78%, and 72%, respectively. No differences in overall survival or OLT-free

survival were observed in patients with TIPS performed before or after the first month after diagnosis. Similar results were observed when comparing patients receiving TIPS before or later than 3 or 6 months from diagnosis. On univariable analysis, only age and BCS-TIPS PI score were significantly associated with survival. Our results were similar to the study by Seijo et al.¹⁷ Overall survival was similar to that observed in a previous retrospective multicenter European study including 124 BCS patients treated with TIPS.⁹ Similar 5 year transplant free survival rates between 77 and 100% after TIPS were observed in other studies.^{3–9} Few noteworthy studies are depicted in Table 4.

A recent study by Tripathi et al. concluded good clinical outcomes following TIPS in BCS. The cohort consisted of 67 patients with a mean follow-up of 82 months. Primary patency rates significantly favored covered stents. Shunt re-interventions were significantly lesser when covered stents were deployed.¹⁸ A systematic review of 79 studies revealed that, the median 1-, 5- and 10-year survival rate was 93, 83 and 73% after interventional radiological treatment; 81, 75 and 72.5% after surgery other than liver transplantation; 82.5, 70.2 and 66.5% after liver transplantation and 68.1, 44.4% and unavailable after medical therapy alone. Bilirubin, creatinine and ascites had prognostic significance.¹⁹

Our observed survival post TIPS matches well with the encouraging results seen in other studies.^{8,9,17,18} These results undeniably indicate that TIPS should be the treatment of choice in all patients with chronic BCS in whom medical therapy fails and radiological interventions (angioplasty, HV/IVC stenting) are unsuccessful. The use of TIPS instead of OLT-first approach will reduce the pressure over the ever increasing transplant-awaiting

Table 4 Comparative Data Regarding OLT-Free Cumulative Survival in BCS.

Author	Number of patients	Age (years)	MELD/CTP score	Indication (%age)	Mortality	Follow-up (months)	Cumulative Survival (1;5;10 years)
Martin Rösslé et al., 2004 ⁸	35	–	–/9.2 ± 1.9	–	9%	37 ± 29	93%;74%;–
Garcia-Pagan et al., 2008 ⁹	124	38 (35–40)	17 (15.7–18.7)/9 (8.6–9.5)	Refractory ascites (59), liver failure (27), variceal bleed (9.5), portal hypertension complications (9.5)	13%	36.7	88%;78%;69%
Seijo et al., 2013 ¹⁷	62	37 (16–83)	–/8 (5–13)	Refractory ascites (69), liver failure (13), and variceal bleeding (7)	20.9%	51	88%;83%;72%
Tripathi et al., 2014 ¹⁸	67	39 ± 14.3	16.1 ± 7/8.8 ± 2.0	Refractory ascites (91) and variceal bleed (9)	11.9%	82	92%;80%;72%
Amarapurkar and Sonavane et al. (present study); 2016	42	40.52 (13–68)	15.38 (9–25)/8.04 (6–11)	Refractory ascites (69), variceal bleed (31)	26.1%	45.5	86%;81%;76%

list of patients. This is clinically relevant in a country like India where organ donation has yet not picked up momentum nationally.

A major limitation of our study was the long enrollment period, which could have resulted in the heterogeneity of the technique, making the learning curve of the procedure a confounding factor. 2 patients had procedural failure and were not included in the analysis.

There is lack of a controlled trial comparing TIPS with liver transplantation. However, considering the good long term survival post-TIPS validate its use as a first line treatment.⁹ The critical role of TIPS in the treatment of BCS has been increasingly recognized. Liver transplantation can thus be reserved for TIPS failure. Our experience with TIPS in BCS conforms to this conclusion. Indications for liver transplant in BCS thus include (1) end-stage chronic liver disease, (2) fulminant liver failure, (3) worsening of liver function in spite of portosystemic shunting, and (4) hepatocellular carcinoma.

COMPLIANCE WITH ETHICAL STANDARDS

All procedures performed were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments. Informed consent was obtained from all individual participants included in the study.

CONFLICTS OF INTEREST

The authors have none to declare.

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