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OPEN Evaluation of outcome from endovascular therapy for Budd-Chiari syndrome: a systematic review and meta-analysis

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This study was performed to evaluate the outcome of endovascular intervention therapy for Budd-Chiari syndrome (BCS) and compare recanalization, transjugular intrahepatic portosystemic shunt (TIPS)/direct intrahepatic portosystemic shunt (DIPS), and combined procedure treatment. For the meta-analysis, 71 studies were identified by searching four databases. The individual studies' samples were used to calculate a confidence interval (CI 95%), and data were pooled using a fixedeffect model and random effect model. The pooled measure and an equal-weighted average rate were calculated in all participant studies. Heterogeneity between the studies was assessed with I², and T² tests, and publication bias was estimated using Egger's regression test. A total of 4,407 BCS patients had undergone an endovascular intervention procedure. The pooled results were 98.9% (95% CI 97.8–98.9%) for a technical success operation, and 96.9% (95% CI 94.9–98.9%) for a clinical success operation. The re-intervention rate after the initial intervention procedure was 18.9% (95% CI 14.7–22.9%), and the survival rates at 1 and 5 years after the initial intervention procedure were 98.9% (95% CI 96.8–98.9%) and 94.9% (95% CI 92.9–96.9%), respectively. Patients receiving recanalization treatment (98%) had a better prognosis than those with a combined procedure (95.6%) and TIPS/DIPS treatment (94.5%). The systematic review and meta-analysis further solidify the role of endovascular intervention treatment in BCS as safe and effective. It maintains high technical and clinical success and long-term survival rates. The recanalization treatment had a better prognosis and outcome than the combined procedures and TIPS/DIPS treatment.

Budd-Chiari syndrome (BCS) is a rare hepatic venous disease. It presents with thrombosis, located anywhere from the hepatic veins (HV) to the suprahepatic of the inferior vena cava (IVC). The result is an outflow obstruction of hepatic veins^{1,2}. The obstruction of BCS is classified as primary or secondary depending on the site of hepatic vein obstruction. The obstruction site can be a thrombus inside the vein or outside the vein due to compression with tumors³. The pathogenesis of BCSs remains unclear, but some known risk factors include myeloproliferative neoplasm, use of oral contraceptive drugs, and coagulation factors^{4,5}. An HV outflow obstruction might cause centrilobular congestion and hepatocyte necrosis. If not treated in time, this can lead to liver cirrhosis, portal hypertension, and ascites. The clinical manifestations of BCS are abdomen pain, hepatomegaly, and ascites^{6,7}. The cause and type of BCS vary by geographical regions; in Western countries, the common cause is HV obstruction, but IVC obstruction is predominate in Eastern countries^{8,9}. Most frequent cause of BCS is thrombophilia, which is detected in more than 84% of patients with BCS^{10,11}. The European Association for the Study of the Liver has recommended a step-wise therapeutic algorithm for BCS. The algorithm depends on treatment response, medical therapy with anticoagulant drugs, angioplasty, stent implantation, thrombolysis, transjugular intrahepatic portosystemic shunt (TIPS), and liver transplantation¹². The progressive improvement in radiological intervention therapy in the past two decades has provided a better survival rate for BCS treatment

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with an intervention procedure than other treatment modalities. Recently, there has been an increase in the number of BCS patients managed with endovascular intervention therapy.

This systematic review and meta-analysis aimed to evaluate the technical and clinical success rates of endovascular intervention operation and re-intervention (including re-occlusion, re-stenosis stent, and shunt dysfunctions). We evaluated the success rates after the initial intervention procedure and the survival rate at 1 and 5 years after the initial intervention procedure. Moreover, this review compares the difference in outcome between recanalization, TIPS/DIPS, and a combined procedure (recanalization and TIPS/DIPS).

Methods

Search strategy. The PubMed, EMBASE, Cochrane Library and Science-Direct databases were searched for relevant published papers. The last search was performed on May 28, 2021. The following search terms were used: Budd-Chiari syndrome, hepatic venous outflow obstruction, hepatic vein stenosis, hepatic vein occlusion, hepatic vein obstruction, supra-hepatic IVC obstruction, membranous obstruction of IVC, endovascular treatment, interventional procedure, transjugular intrahepatic portosystemic shunt (TIPS), direct intrahepatic portosystemic shunt (DIPS), percutaneous transluminal balloon angioplasty(PTBA), percutaneous transluminal angioplasty (PTA) of the hepatic vein, vascular recanalization of the hepatic vein, vascular stent implantation in the hepatic vein, and vascular stent implantation in IVC.

Selection criteria. The following criteria were used to determine those studies to include: (1) study had more than ten case participants; (2) retrospective studies, prospective studies, including case series, and case-control studies were eligible; (3) all participants of any age, race, origin with a diagnosis of BCS; (4) full article papers with detailed information and statistical results of intervention treatment; and (5) there were no publication data, publication language or publication status restrictions. Exclusion criteria were: (1) duplicates studies; (2) studies that were not original papers; (3) case reports; (4) comments, (5) essays; (6) abstracts; (7) small case series; (8) not reporting relevant clinical outcomes; (9) lack of detailed results; (10) review articles; (11) less than ten patients; (12) studies unmatched inclusion criteria; (13) studies with missing survival rate, re-intervention rate or clinical success. The study selection process followed the Preferred Reporting Items for Systematic Reviews and Meta-analysis (PRISMA) guideline flowchart (Fig. 1)¹³. The PRISMA checklist is provided in (Supplementary Table 1).

Data extraction. The following data were extracted for further analysis: (1) First author, publication year, enrollment period, country, number of BCS patients with endovascular intervention treated, age, gender, site of the obstruction (HV, IVC and combination), type of intervention procedures, technical and clinical success rate, rate of the re-intervention (re-occlusion, re-stenosis stent and dysfunction of shunt), and survival rate at 1 and 5 years.

Ouality assessment. Studies were considered higher quality if they fulfilled all the following predetermined criteria: (1) patients were admitted to the hospital; (2) the interval of enrollment and eligibility criteria was recorded; (3) the site of obstruction of BCS patients was reported; and (4) Patients were diagnosed with BCS and treated with endovascular intervention procedures.

HV-angioplasty. When the stiff guide wire was established, a balloon dilator catheter of 12–15 mm diameter was inserted from the right jugular vein puncture site to the obstructed part of HV/IVC via the guide wire. Next, the balloon catheter was dilated twice, and each dilatation occurred for approximately 40 s. If there was more than 30% residual stenosis on HV venography after balloon dilation then a stent was inserted in the stenosis part of the HV.

IVC-angioplasty. Venography was performed (right femoral vein or right jugular vein) to evaluate the IVC anatomy and obstruction characteristics. Next, a guidewire with a balloon catheter (25–30 mm) was used to dilate IVC obstructive lesions. A self-expandable metallic stent was used if the IVC narrowed immediately after balloon dilatation or more than 30% residual stenosis on IVC venography after balloon dilation.

Combined HV and IVC angioplasty. Combined HV and IVC stenting were performed in patients having short-segment HV and IVC obstructions.

Recanalization. Recanalization (PTA with or without stent placement) has been used in 31 (43.66%) studies with or without stent placement. In the subgroup, we analyze the technical and clinical success rate of recanalization, re-intervention treatment, and survival rate at 1 and 5 years of recanalization procedure. It was performed with balloon dilation or endovascular stent placement in the stenosis part of HV and IVC.

TIPS/DIPS. TIPS/DIPS were used in 17 (23.94%) studies. In the subgroup, we analyze the technical and clinical success rate of TIPS/DIPS, re-intervention treatment, and survival rate at 1 and 5 years of TIPS/DIPS procedure. This was performed in symptomatic patients with non-recanalization HV obstruction with small collaterals draining into IVC, portal hypertension, refractory ascites, variceal bleeding, and long segment obstruction HV. DIPS usually used in failed TIPS, occluded three major HVs and anomalies of HVs.



Figure 1. PRISMA flowchart of studies selection.

Recanalization and TIPS/DIPS (combined procedures). Recanalization (PTA with or without stent placement) and TIPS/DIPS were used in 23 (32.39%) studies. We analyze the technical and clinical success rate, re-intervention treatment, and survival rate at 1 and 5 years of combined procedures in the subgroup.

Definition. *Technical success.* Technical success of recanalization was defined as the complete elimination of HV or IVC obstruction and confirmed by venography. Technical success of TIPS was defined as successful placement of artificial stent between the hepatic vein and the portal vein. The stent position was confirmed by angiography, and the contrast medium flowed back into the right atrium smoothly through the intrahepatic shunt.

Clinical success. Clinical success of recanalization, combined procedures, and TIPS/DIPS was defined as an improvement of BCS related-symptoms and liver function after a technical success within day one to 90 days.

Statistical analysis. The individual studies' sample sizes were used to calculate a confidence interval (CI: 95%). The pooled measure and an equal weighted average rate were calculated in all participant studies. The data were pooled using a fixed effect model and random effect model. Heterogeneity between studies was assessed with the I² and T² tests (I²>50% or $P \le 0.10$ was considered statistically significant heterogeneity). Publication bias was estimated using Egger's regression asymmetry test ($P \le 0.05$ represented statistically significant publica-

tion bias). Subgroup analyses were performed according to the continent of objectives. Statistical analyses were carried out using the R-version 3.5.3 software.

Results

Study characteristics. Overall, a total of 536 papers were identified in four databases. Among them, 71 original articles^{9,14-83} were eligible for systematic review and meta-analysis (Fig. 1). The general characteristics of the included studies are listed in Table 1. All included studies were published between 1995 and 2019. Among them, 33 (46.4%) were published between 2015 and 2019, and four (5.6%) before 2000. Most of the papers were published after 2010. Thirty-five (50%) studies were conducted in China, ten (14.2%) studies in India, four studies in the UK, three studies in Germany and Egypt, and two studies each in the USA, Italy, Netherland, and Turkey (Table 1).

A total of 4407 patients underwent endovascular intervention procedures. Among them, 98.9% of patients were considered technical successes and 96.9% achieved clinical improvement. The site of obstruction was documented in 53 (75.7%) studies, including 42.25% in HV, 30.98% in the IVC, and 26.76% in combined (HV and IVC) (Table 1). In subgroup analysis, recanalization was used in 31(43.66%) studies, combined procedures (recanalization and TIPS) in 23 (32.39%) studies, and TIPS in 17 (23.94%) studies (Table 1).

Study quality assessment. Patients were consecutively admitted in 57 (80.28%) studies $^{9,12,14-25,28,30-39,41-45,47-57,59,64,66,68,70-75,78-83}$. Fifty one (71.83%) studies were considered to be of high-quality $^{9,12,14,16-19,21-23,25,26,31-34,39,41-43,45-47,49-56,58,59,61-64,66-68,70-75,78-83}$ and six (8.45%) studies were of poor-quality 13,15,29,65,76,77 . The site of obstruction was clearly reported in 53 (75.7%) studies (Table 1). The interval of enrollment and eligibility criteria were recorded in all included studies. All patients were diagnosed with BCS and treated with endovascular intervention procedures.

The technical success rate of endovascular intervention procedures. The technical success rate of all individual studies is shown in Fig. 2. The pooled result of total technical success procedures was 98.9% (95% CI 97.8–98.9%), with statistically significant heterogeneity among studies ($I^2 = 54\%$, P < 0.01). The pooled results of the recanalization, combined procedures, and TIPS subgroups were 97.9% (95% CI 96.8–98.9%), 98.9% (95% CI 97.9–99.9%), respectively.

The clinical success rate of endovascular intervention treatment. The clinical success rates of all cases of BCS are shown in Fig. 3. The pooled result of the total patients with a clinical success rate was 96.9% (95% CI 94.9–98.9%), with statistically significant heterogeneity among studies (I^2 =83%, P<0.01). The pooled results of the recanalization, combined procedures, and TIPS subgroups were 97.9% (95% CI 95.9–99.9%), 95.6% (95% CI 92.7–98.9%), and 94.0% (95% CI 88.5–98.8%), respectively.

The rate of re-intervention at 5 years after initial intervention treatment. The vascular re-occlusion, stent stenosis, and shunt dysfunction at 5 years after initial endovascular intervention procedures of BCS are shown in Fig. 4. The pooled result of total re-intervention was 18.9% (95% CI: 14.7–16.9%), with statistically significant heterogeneity among studies (I^2 =90%, P<0.01). The pooled results of the recanalization, combined procedures, and TIPS subgroups were 10.8% (95% 7.5–13.8%), 17.9% (95% CI 10.9–24.9%), and 42.9% (95% CI 29.9–56.8%), respectively.

The survival rate at 1 and 5 years after endovascular intervention procedures. The survival rate of endovascular intervention therapy of BCS patients at 1 and 5 years after initial intervention procedures are shown in Figs. 5 and 6. The pooled result of the total survival rate at 1 year was 98.9% (95% CI 96.8–98.9%), with statistically significant heterogeneity among studies ($I^2 = 60\%$, P < 0.01). The pooled results of the recanalization, combined procedures, and TIPS subgroups were 99.9% (95% CI 98.9–99.9%), 96.9% (95% CI 94.8–97.9%), and 94.9% (95% CI 91.9–96.7%), respectively. Similarly, the pooled result of the total survival rate at 5 years was 94.9% (95% CI: 92.5–96.9%), with statistically significant heterogeneity among studies ($I^2 = 77\%$, P < 0.01). The pooled results of the recanalization, TIPS, and combined procedures subgroups were 97.9% (95% CI 94.8–97.9%), espectively. 84.9–91.9%), and 93.9% (95% CI 90.9–95.9%), respectively.

Publication bias. The results of publication bias in the studies evaluated with Egger's test. The publication bias for the technical success rate of endovascular intervention procedures (P=0.0335), clinical success (P=0.5567), re-intervention (P=0.08108), the survival rate in one year (P=0.01549) and the survival rate at five years (P=0.8909). Although the P value of technical success and survival rate at 1 year was statistically significant.

Discussion

This extensive study evaluates and updates the clinical efficacy and long-term outcome of endovascular therapy in BCS patients and compares recanalization, TIPS/DIPS, and combined procedures. The technical and clinical success rates were 98.9% and 96.9%. After the initial endovascular treatment, the re-intervention rate was 18.9%, and the survival rates at 1 and 5 years after the initial endovascular treatment were 97.9% and 94.9%, respectively. The findings indicate that endovascular intervention treatment is safe, effective, and provides long term survival rates in patients with BCS.

1st author/					Ste of ste	nosis		Type of treatment		Success rate		Re-Intervention		Survival rate				
published/ reference	Country	N.P.	M/F	Mean Age	HV	IVC	Both	Recanalize	TIPS/DIPS	Stent	Angio	Thrombo	Technical (%)	Clinical (%)	Re-stenosis (%)	Dysfunction (%)	1 year (%)	5 years (%)
Fu YF 2015 ²²	China	20	11/9	22-56	20	-	-	20	-	2	18	-	100	100	15	-	100	NA
Ding PX 201814	China	108	69/39	25-74	-	1	107	107	-	13	94	12	99.1	99.5	16.5	-	95	86
Nagral A 2010 ¹⁵	India	11	5/6	4 m-11 y	11	-	-	5	6	2	3	-	100	100	0	-	90.9	NA
Rossle M 2004 ¹⁶	Germany	35	8/27	12-74	NA	NA	NA	-	33	-	-	-	94.2	100	57.5	57.5	91.4	91.4
Blum U 1995 ¹⁷	Germany	12	6/6	31-71	NA	NA	NA	-	12	-	-	-	100	83.3	41.6	41.6	75	NA
Pavri TM 2014 ¹⁸	USA	21/47	16/31	31-69	NA	NA	NA	-	21	-	-	-	100	85.7	57	52.3	100	81.5
Xu ke 1996 ¹⁹	China	32	6/26	20-56	12	20	-	31	-	17	20	-	100	96.8	37.5	-	96.8	96.8
Kathuri R 2014 ²⁰	India	25	16/9	2-16	20	01	04	25	-	20	5	-	100	100	25	-	96	96
KhurooMS 2005 ²¹	Soudi arabia	16/40	17/23	15-64	16	19	-	6	8	-	6	-	87.5	92.8	14.2	62.5	92.8	92.8
Jagtap N 2017 ²³	India	88	52/36	20-56	33	42	13	75	0/13	64	73	-	98.8	86.3	17.2	-	95.4	93.1
Zahan A 2010-*	Germany	13	3/10	14-60	11	-	2	-	13	-	-	-	100	100	84.6	84.6	92.3	92.3
2017 ²⁵	China	47	33/14	21-71	33	-	14	61	-	-	61	-	100	100	10.8	-	100	100
Yang F 2019 ²⁶	china	33	16/17	44-74	-	33	-	33	-	15	18	-	100	100	9	-	100	100
Amara DN 2008 ²⁷	India	38/49	24/25	1-57	29	10	10	22	15	22	2	-	97.5	100	16.2	-	94.5	94.5
Cheng D 201328	china	141/145	90/55	10-82	45	8	92	133	1	16	133	48	95	100	4.4	-	99	NA
Fu Y-F 2015 29	China	17	13/4	43-72	17	-	-	17	-	4	13	-	100	100	11.7	-	100	NA
Huang Q 2016	China	265	131/134	18-79	-	-	265	263	-	56	263	-	99.5	100	14.6	-	99.6	98
2003 ³¹	India	17	NA	30-50	NA	NA	NA	15	-	-	15	-	88.5	100	20	-	NA	NA
Mo A 2017 ³²	Australia	27	11/14	21-76	NA	NA	NA	11	18	11	11	-	92.6	96	56	77.7	96	81
Zhang B 2013 33 Meng X 2016 ³⁴	China	18	39/14	19-50 NA	-	4	-	53	3	- 47	53	- 13	96.5	100	16.6	-	100 90	86
Chen ZK 2017 35	China	68	39/29	22-52	68	-	-	68	-	8	60	-	100	95.6	27.9	-	96.9	93.4
Rathod K 2016 ³⁶	India	190	102/88	15-55	147	40	3	84	106	84	78	-	100	80.5	10	10	100	100
Sang H-F 2014 ³⁵	China	48	31/17	25-65	NA	NA	NA	43	-	31	43	5	89.6	100	9.3	-	100	100
Rosenqvit K 2016 ³⁸	Sweden	13	6/7	16-63	NA	NA	NA	-	13	-	-	-	100	100	15.3	30.7	100	93
Bi Y 2018 ³⁹	China	60	48/12	12-76	35	-	25	31	27	-	31	-	96.6	78	23.3	62.9	98.3	98.3
Darwish M 2009 ⁹	Netherland	64/163	70/93	16-83	NA	Na	N A	22	56	-	8	10	100	100	14	16	83	NA
Al-Warraky 2015 ⁴⁰	Egypt	103	30/73	14-44	88	9	6	26	55/22	-	26	-	98	99	30.6	22.6	98	92
Eapen CE 2005 ⁴¹	UK	61	22/39	16-67	58	3	-	32	29	8	24	6	100	100	65.5	65.5	94	87
Li T 2009 ⁴²	China	101	52/49	15-57	101	-	-	92	-	2	92	-	91	100	13	-	100	NA
Tripathi D 2014 ⁴³	UK	67	21/46	15-70	NA	NA	NA	-	67	-	-	-	100	97	44.7	44.7	92	80
Fan X 2016 ⁴⁴	China	60	27/33	18-60	51	-	9	27	33	-	27	-	100	96.6	13.3	13.3	96.6	96.6
Seijo S 201345	Europe	70	NA	16-83	NA	NA	NA	8	62	-	8	9	100	94.2	0	-	84.2	84.2
Srinivas 201246	India	12	7/5	28-55	-	12	-	12	-	5	7	-	100	100	8.3	-	100	100
Qiao T 200547	China	44	25/19	19-77	8	32	4	45	-	45	-	-	93.1	100	8.5	-	100	100
Cheng D 2019 ⁴⁸ Tripathi D	China	162	94/68	18-78	-	-	162	157	-	35	208	47	96.9	92.9	8.2	-	100	NA
2016 ⁴⁹ Sonavane	UK	63	27/36	15-55	55	3	5	63	-	31	32	8	100	73	17.4	-	97	89
201850	India	42	26/16	19-68	42	-	-	-	42	-	-	-	100	100	7.1	7.1	86	81
2003 ⁵¹	China	115	65/50	17-67	13	85	17	122	-	122	-	-	92.4	99.1	4.7	-	100	100
Bi V 201853	China	54 40	20/34	15-6/	54	-	- 27	- 40	3	-	- 40	- 24	98	67.9	51	41.5	96	83
Bi Y 2018 ⁵⁴	China	72	43/29	22-76	-	3	69	91	-	-	91	12	97.5	79.2	0	-	100	91.5
Ding PX 201955	China	456	264/192	22-74	-	456	-	455	5	25	455	85	99.8	99.3	19.4	-	98.5	91.2
Shalimar 2017 ⁵⁶	India	80	40/40	12-50	61	-	19	-	80	-	-	-	100	88.8	13.7	13.7	93.7	90
Ding PX 2015 ⁵⁷	China	93	59/34	15-72	65	-	28	93		2	93	-	100	100	11.8	-	98.9	97.8
Darwish M 2007 ⁵⁸	Netherland	17	10/6	19-50	16	-	11	-	16	-	-	-	94.1	94	0	62.5	80	72
Fu Y 2011 ⁵⁹	China	18/29	13/16	23-67	4	18	-	22	-	-	22	-	100	100	5.5	-	100	100
Eldorry A 2011 ⁴⁰	Egypt	25	9/16	14-57	NA	NA	NA	12	13	10	12	-	100	96	12	38.34	100	NA
Cheng DL 2018 ⁶¹	China	69	43/26	15-72	66	-	-	66	-	11	66	19	95.7	92.4	0	-	98.5	94
Yu C 2019 ⁶²	China	56	30/26	29-65	-	56	-	55	-	-	55	-	98.2	100	12.7	-	100	100
Wu T 2002 63	China	42	28/14	12-62	-	42	4	41	-		41	-	97.6	100	12.1	-	100	100
Han G 201364	China	177	93/75	12-62	50	33	94	168	-	117	168	-	95	90	14.8	-	96	83
Cui Y-F 201566	China	143	58/78	14-74	- 143	-	-	140	3	16	124	-	90.0	97.1	20.5	-	97.7	93.5
Boyvat F 2008 ⁶⁷	Turkey	11	5/6	6-43	NA	NA	NA	-	- 11	-	-	-	100	81.8	45.4	81.8	100	NA
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1st author/					Ste of stenosis Type of tr				pe of treatment				Success rate		Re-Intervention		Survival rate	
years of published/ reference	Country	N.P.	M/F	Mean Age	нv	IVC	Both	Recanalize	TIPS/DIPS	Stent	Angio	Thrombo	Technical (%)	Clinical (%)	Re-stenosis (%)	Dysfunction (%)	1 year (%)	5 years (%)
Kucukay F 2016 ⁶⁸	Turkey	32	18/14	20-42	NA	32	NA	30	-	-	30	-	94	100	10	-	100	100
Lee BB 2006 ⁶⁹	South Korea	17/28	13/15	28-68	2	26	-	15	2	6	15	-	100	82.3	23.5	-	100	NA
Griffith JF 1996 ⁷⁰	UK	18	8/10	16-65	12	-	6	18	-	6	18	5	100	56	27.7	-	89	78
Cui YF 2015 ⁷¹	China	17	8/6	25-66	14	-	-	14		2	12	-	82.3	100	21.4	-	100	NA
Yang XL 1996 ⁷²	China	42	28/14	16-56	-	38	-	38	-	-	38	-	91	100	2.6	-	100	100
Xue H 2009 ⁷³	China	53	39/14	11-70	11	38	4	47	2	34	13	-	92.5	100	0	-	93.8	93.8
Molmenti 2005 ⁷⁴	USA	11	5/6	22-78	NA	NA	NA	-	10	-	-	-	91	100	0	-	100	100
Garcia-pag 2008 ¹¹⁴	Italy	133	78/46	35-40	NA	NA	NA	-	124	-	-	-	93.2	82.2	49.1	49.1	95	87
Katerina 2013 ¹¹⁵	Greece	14	3/11	3-66	NA	NA	NA	-	14	-	-	-	100	100	28.5	28.5	100	100
Neumann 2013 ⁷⁶	Denmark	14	3/11	17-66	NA	NA	NA	-	14	-	-	-	100	100	78.5	100	100	92.8
Wang R 2013 ⁷⁷	China	29	NA	NA	-	29	-	28	-	18	-	15	96.6	100	14.2	-	100	100
Corso R 200878	Italy	15	7/8	7-52	NA	NA	NA	-	15	-	-	15	100	100	40	40	86.6	86.6
Ding PX 2010 ³⁹	China	13	9/4	39-74	-	13	-	13	-		13	13	100	100	0	-	100	NA
Fu YF 2015 ⁸⁰	China	66	34/32	21-79	66	-	-	66	-	18	50	-	100	100	16.6	-	100	100
Mukund A 2018 ⁸²	India	136	96/40	1-67	106	30	-	92	44	64	92	4	100	87.5	5.1	5.1	94.1	94.1
Mohamed 2018 ⁸³	Egypt	118	43/75	20-45	118	-	-	-	118	-	-	-	100	83.0	40.74	40.74	95.8	91.5

Table 1. Overview on baseline of the included studies.

Most of the studies were conducted in Asian countries, half of the study sample was from China (50%), and 45.7% of the study sample was published from 2015 to 2019. Most of the patients were treated with endovascular recanalization with or without stent placement. The subgroups' pooled result showed that the re-intervention treatment rate was high in TIPS/DIPS, the technical success rate higher in combined procedures, and the clinical success rate and the survival rate at 1 and 5 years were higher with recanalization. It was interesting to find that the most common obstruction site was HV in the Asian countries. Also, most Asian studies reported the most common obstruction sites IVC and combined (HV and IVC)^{48,84,85}. However, some studies have reported HV obstruction as the most common cause of BCS in the Asian population^{27,86}.

BCS can be classified according to etiology (primary and secondary), site of obstruction (HV, IVC, and combined HV + IVC), the manifestation of the disease (fulminant or non-fulminant), and duration of the disease (acute, subacute or chronic)². The clinical presentation is highly variable but may be categorized as acute/ fulminant hepatic failure, as subacute without evidence of cirrhosis and as chronic with evidence of portal hypertension and cirrhosis⁸⁷. In this meta-analysis, we found most of the studies were treated according to the site of obstruction (42.25% in HV, 30.98% in the IVC, and 26.76% in combined HV + IVC). Recanalization and TIPS treatments for BCS depend on the anatomical site and the extent of obstruction and liver function⁵⁸. HV recanalization and TIPS have become the main treatment for HV-type BCS ^{16,33,38}.

BCS is a rare disorder and therefore management guidelines are based on the retrospective case series, expert opinion and clinical presentation^{75,88–90}, due to the lack of randomized controlled trials study⁹. BCS is more prevalent in developing countries such as, China, India, Nepal and South Africa. In contrast, the most common cause is membranous obstruction, an underlying thrombotic disorder that has been in only a few patients²⁸, where the treatment choice is recanalization. However, only 29–41% of Western patients have membranous or segment obstruction^{41,91}, and pure hepatic vein thrombosis accounts for more than half of BCS cases⁹². In contrast, recanalization is not applicable in most Western patients with BCS, and TIPS is a preferable treatment⁴¹.

Membranous obstruction of IVC is a common cause of hepatic venous outflow obstruction, which has short web narrowing to a long segmental occlusion with or without narrowing of hepatic vein^{46,93}. In the West, HV thrombosis is the most common cause, while in Asian countries isolated IVC membranous webs are more common^{84,85}, and two-thirds of IVC obstructions are due to membranous or segment obstruction. The long-term treatment outcome of endovascular intervention treatment was better for membranous obstruction of IVC rather than segmental obstruction of IVC. PTA alone could be the optimal treatment for membranous obstruction and stenting should be more strongly recommended for a segment of obstruction of IVC³⁰.

The thrombophilic factors are responsible to development of BCS, which is detected in up to 84% of BCS patients^{10,11}. The most common thrombophilic factors are myeloproliferative disease and factor V linden¹¹. In over 25% of BCS cause more than one thrombophilic state may be present with BCS patients⁹⁴. Most inherited thrombophilias result increased thrombosis due to an impaired neutralization of thrombin or failure to control of generation of thrombin⁹⁵. Data show that prothrombotic disorders are not common in china as a cause of unknown factors in Chinese BCS patients⁹⁶. The thrombophilia is more commonly found in western BCS patient than Chinese BCS patients⁹⁷.

HV recanalization was performed in patients with short-segment HV obstruction (<3 cm), and stenting was performed in long segment HV occlusion (>3 cm) with large collateral vein drainage³⁶. HV recanalization is usually difficult for BCS patients with segmental obstruction, whereas TIPS placement has been widely used for BCS patients who fail to HV recanalization^{41,98}. In patients with compensatory but obstruction accessory hepatic vein (AHV), Fu et al.²² reported that recanalization of the AHV is a simple, safe, and effective treatment

Study	Events	Total		Proportion	95%-CI	Weight (fixed)	Weight (random)
treatment = Recanaliza	tion						
Yufe Fu 2015	20	20		1.00	[0.83; 1.00]	0.4%	0.9%
Ding P. Au 2016	32	32		1.00	[0.95; 1.00]	2.4%	2.1%
Pengli zhou 2017	47	47		1.00	[0.92; 1.00]	1.0%	1.5%
Fang yang 2019	33	33		1.00	[0.89; 1.00]	0.7%	1.2%
Yu fei fu 2015	17	17		1.00	[0.80; 1.00]	0.4%	0.8%
Qian Xin 2016 Mishra 2003	263	265		0.99	[0.97; 1.00]	5.8%	2.6%
Zhonake 2017	68	68	· · · · ·	1.00	[0.95: 1.00]	1.5%	1.7%
Hong fei 2014	43	48		0.90	[0.77; 0.97]	1.1%	1.5%
Tianxiao Li 2009	92	101		0.91	[0.84; 0.96]	2.2%	2.0%
Srinivas BC 2012	12	12		1.00	[0.74; 1.00]	0.3%	0.6%
Delei cheng 2019	157	162		0.93	[0.93: 0.99]	3.6%	2.3%
D.tripathi 2016	63	63		1.00	[0.94; 1.00]	1.4%	1.7%
Chun qing 2003	106	115		0.92	[0.86; 0.96]	2.5%	2.1%
Yonghua Bi 2018	70	72		0.97	[0.90; 1.00]	1.6%	1.8%
Yang fu 2011	18	18		1.00	[0.81: 1.00]	0.4%	0.8%
Delei cheng 2018	66	69		0.96	[0.88; 0.99]	1.5%	1.8%
Chaowen 2019	55	56		0.98	[0.90; 1.00]	1.2%	1.6%
Tongguo 2002	41	42		0.98	[0.87; 1.00]	0.9%	1.4%
Gaunong 2013 Yufei Eu 2015	60	62		0.95	[0.91; 0.98]	3.9%	2.4%
Fahrettin 2016	30	32		0.94	[0.79; 0.99]	0.7%	1.2%
J.F Griffith 1996	18	18		1.00	[0.81; 1.00]	0.4%	0.8%
Yan feng Yu 2015	14	17	· · · · · · · · · · · · · · · · · · ·	0.82	[0.57; 0.96]	0.4%	0.8%
Ruibua 2013	38	42 29		0.90	[0.77; 0.97]	0.9%	1.4%
P.Xu Ding 2010	13	13		1.00	[0.75: 1.00]	0.3%	0.6%
Yu fei Fu 2015	66	66		1.00	[0.95; 1.00]	1.5%	1.7%
Fixed effect model		1958	•	0.98	[0.97; 0.99]	43.3%	
Random effects model	0.005		\$	0.98	[0.97; 0.99]		45.2%
Heterogeneity: $I^- = 56\%$, τ^-	= 0.005	1, <i>p</i> < 0.01					
treatment = Recanalization	tion + Tl	PS/DIPS					
Albha N 2010	11	11		1.00	[0.72; 1.00]	0.3%	0.6%
Rachna 2014	25	25		1.00	[0.86; 1.00]	0.6%	1.0%
Monammed-2005	14	16 -	·	0.88	[0.62; 0.98]	1.9%	0.7%
Deepak N 2008	37	38		0.95	[0.86: 1.00]	0.8%	1.3%
Delei cheng 2013	134	141		0.95	[0.90; 0.98]	3.1%	2.2%
Allison 2017	25	27		0.93	[0.76; 0.99]	0.6%	1.1%
Bozhang 2013	18	18		1.00	[0.81; 1.00]	0.4%	0.8%
Kranti 2016	190	190		1.00	[0.87, 1.00]	4.2%	2.4%
Yonghua 2018	58	60		0.97	[0.88; 1.00]	1.3%	1.6%
Darwish 2009	64	64		1.00	[0.94; 1.00]	1.4%	1.7%
Warraky 2015	101	103		0.98	[0.93; 1.00]	2.3%	2.0%
Eapen 2005 Xinvin fan 2016	61	60		1.00	[0.94; 1.00]	1.3%	1.7%
Susanaseiio2013	70	70		1.00	[0.95; 1.00]	1.5%	1.8%
Yonghua Bi 2018	40	40		1.00	[0.91; 1.00]	0.9%	1.3%
P. Xu Ding 2019	455	456	-	1.00	[0.99; 1.00]	10.0%	2.8%
Ahmed 2011 Venfong Cui 2015	25	25		1.00	[0.86; 1.00]	0.6%	1.0%
B. boong 2006	140	143		1.00	[0.80: 1.00]	0.4%	0.8%
Huixue 2009	49	53		0.92	[0.82; 0.98]	1.2%	1.6%
Mukund A 2018	136	136	1	1.00	[0.97; 1.00]	3.0%	2.2%
Fixed effect model		1897	4	1.00	[0.99; 1.00]	41.8%	26.0%
Heterogeneity: $l^2 = 53\% \tau^2$	² = 0.0036	6. p < 0.01	Ŷ	0.99	[0.00, 1.00]		30.0%
treatment = TIPS/DIPS	-	05			10.04.0.000	0.00	
Illrich 1995	33	35		1.00	[0.81; 0.99]	0.8%	0.6%
Tanya M 2014	21	21		1.00	[0.84; 1.00]	0.5%	0.9%
Alexeanda2010	13	13		1.00	[0.75; 1.00]	0.3%	0.6%
Kerstin 2016	13	13		1.00	[0.75; 1.00]	0.3%	0.6%
D. Iripathi 2014 Sonayang 2019	67	67		1.00	[0.95; 1.00]	1.5%	1.7%
Georges 2016	53	54		0.98	[0.92, 1.00]	1.2%	1.6%
Shalimar 2017	80	80	-	1.00	[0.95; 1.00]	1.8%	1.9%
S .darwsh 2007	16	17		0.94	[0.71; 1.00]	0.4%	0.8%
F. boyvat 2008	11	11		1.00	[0.72; 1.00]	0.3%	0.6%
Juncarlos 2008	10 124	133		0.91	[0.59; 1.00] [0.88: 0.971	2.3%	2.2%
Katerina 2013	14	14		1.00	[0.77; 1.00]	0.3%	0.7%
Anders 2013	14	14		1.00	[0.77; 1.00]	0.3%	0.7%
R.corso 2008	15	15		1.00	[0.78; 1.00]	0.3%	0.7%
Monamed 2021	118	118 670	Ť	1.00	[0.97; 1.00]	2.6%	2.1%
Random effects model		070	e Ø	1.00	[0.98; 1.00]	14.9%	18.8%
Heterogeneity: $I^2 = 33\%$, τ^2	² = 0.0033	3, <i>p</i> = 0.09			L.100, 1100]		
Fixed effect model		4525	2	0.99	[0.99; 1.00]	100.0%	100.0%
Heterogeneity: $I^2 = 55\% \tau^2$	² = 0.004	$7. \rho < 0.01$		0.99	[0.90, 0.99]		100.0%
Residual heterogeneity: I2	= 51%, p	< 0.01 0.6	0.7 0.8 0.9 1				

Figure 2. The Forest plot of technically success rate of intervention procedures in BCS patients, horizontal lines indicate 95% confidence intervals, square size indicates study specific statistical weight, and diamond indicates the overall treatment effect with 95% confidence intervals.

option for long segmental obstruction of the HV. However, TIPS is often the treatment choice for long segmental obstruction of $\rm HV^{41,76}$.

Study	Events	Total		Proportion	95%-CI	Weight (fixed)	Weight (random)
treatment = Recanalizat	tion						
Yufe Fu 2015	20	20		1.00	[0.83; 1.00]	0.5%	1.2%
Ding P. Xu 2018	99	107		0.93	[0.86; 0.97]	2.4%	1.7%
Ke Xu 1996	31	32		0.97	[0.84; 1.00]	0.7%	1.4%
Fengli znou 2017	47	47		1.00	[0.92; 1.00]	0.8%	1.5%
Yu fei fu 2015	17	17		1.00	[0.80: 1.00]	0.4%	1.1%
Qian xin 2016	263	263		1.00	[0.99; 1.00]	5.9%	1.8%
Mishra 2003	15	15		1.00	[0.78; 1.00]	0.3%	1.1%
Zhongke 2017	65	68		0.96	[0.88; 0.99]	1.5%	1.6%
Hong fei 2014	43	43		1.00	[0.92; 1.00]	1.0%	1.5%
Srinivae BC 2012	92	12		1.00	[0.96; 1.00]	2.1%	1.7%
Tonggiao 2005	35	35		1.00	[0.90: 1.00]	0.8%	1.4%
Delei cheng 2019	146	157		0.93	[0.88; 0.96]	3.5%	1.7%
D.tripathi 2016	46	63		0.73	[0.60; 0.83]	1.4%	1.6%
Chun qing 2003	120	121		0.99	[0.95; 1.00]	2.7%	1.7%
Yonghua Bi 2018	57	70		0.81	[0.70; 0.90]	1.6%	1.6%
P. Au Ding 2015 Vana fu 2011	93	93		1.00	[0.96; 1.00]	2.1%	1.7%
Delei cheng 2018	61	66		0.92	[0.83: 0.97]	1.5%	1.6%
Chaowen 2019	55	55		1.00	[0.94; 1.00]	1.2%	1.5%
Tongguo 2002	41	41		1.00	[0.91; 1.00]	0.9%	1.5%
Gauhong 2013	158	168		0.94	[0.89; 0.97]	3.8%	1.8%
Yutei Fu 2015	60	60		1.00	[0.94; 1.00]	1.4%	1.6%
Failfellin 2016	30	30 18 —		1.00	[0.31.0.79]	0.7%	1.4%
Yan feng Yu 2015	14	14	· · · · ·	1.00	[0.77; 1.00]	0.4%	1.0%
Xueliang 1996	38	38		1.00	[0.91; 1.00]	0.9%	1.4%
Ruihua 2013	28	28		1.00	[0.88; 1.00]	0.6%	1.3%
P.Xu Ding 2010	13	13		1.00	[0.75; 1.00]	0.3%	1.0%
Yu fei Fu 2015	66	66		1.00	[0.95; 1.00]	1.5%	1.6%
Fixed effect model		1903	10	0.98	[0.98; 0.99]	43.0%	44.69/
Heterogeneity: $l^2 = 81\% \tau^2$	- 0.017	3 0 < 0 0	-	0.96	[0.96; 1.00]		44.0 %
neterogeneity. r = 0176, t	- 0.0170	5, p < 0.0					
treatment = Recanalizat	tion + Tl	PS/DIPS					
Albha N 2010	11	11		1.00	[0.72; 1.00]	0.3%	0.9%
Rachna 2014	25	25		1.00	[0.86; 1.00]	0.6%	1.3%
Nonammed-2005	13	14		0.93	[0.66; 1.00]	2.0%	1.0%
Deepak N 2008	37	37		1.00	[0.91: 1.00]	0.8%	1.4%
Delei cheng 2013	134	134	-	1.00	[0.97; 1.00]	3.0%	1.7%
Allison 2017	24	25		0.96	[0.80; 1.00]	0.6%	1.3%
Bozhang 2013	18	18		1.00	[0.81; 1.00]	0.4%	1.2%
Xiang 2016	45	53		0.85	[0.72; 0.93]	1.2%	1.5%
Yonghua 2018	47	58		0.81	[0.74, 0.80]	4.3%	1.6%
Darwish 2009	64	64		1.00	[0.94: 1.00]	1.4%	1.6%
Warraky 2015	100	101		0.99	[0.95; 1.00]	2.3%	1.7%
Eapen 2005	61	61		1.00	[0.94; 1.00]	1.4%	1.6%
Xinxin fan 2016	58	60		0.97	[0.88; 1.00]	1.4%	1.6%
Susanasejio2013	66	70		0.94	[0.86; 0.98]	1.6%	1.6%
P Xu Ding 2019	452	455		0.92	[0.79, 0.90]	10.9%	1.4%
Ahmed 2011	24	25		0.96	[0.80; 1.00]	0.6%	1.3%
Yanfeng Cui 2015	136	140	+	0.97	[0.93; 0.99]	3.1%	1.7%
B. boong 2006	14	17		0.82	[0.57; 0.96]	0.4%	1.1%
Huixue 2009	49	49		1.00	[0.93; 1.00]	1.1%	1.5%
Fixed offect model	119	136	j	0.88	[0.81; 0.93]	3.1%	1.7%
Random effects model		1070		0.96	[0.93; 0.99]		34.1%
Heterogeneity: $I^2 = 87\%$, τ^2	= 0.0213	3, <i>p</i> < 0.0					
treatment - TIDO/DIDO							
Martin 2004	30	33		1.00	FO 89- 1 001	0 8%	1 /0/-
Ulrich 1995	10	12		0.83	[0.52: 0.98]	0.3%	1.0%
Tanya M 2014	18	21		0.86	[0.64; 0.97]	0.5%	1.2%
Alexeanda2010	13	13		1.00	[0.75; 1.00]	0.3%	1.0%
Kerstin 2016	13	13		1.00	[0.75; 1.00]	0.3%	1.0%
D. Iripathi 2014	65	67		0.97	[0.90; 1.00]	1.5%	1.6%
Sonavane 2018	42	42		0.68	[0.92; 1.00]	1.0%	1.5%
Shalimar 2017	71	80		0.89	[0.80; 0.95]	1.8%	1.6%
S .darwsh 2007	15	16	+	0.94	[0.70; 1.00]	0.4%	1.1%
F. boyvat 2008	9	11		0.82	[0.48; 0.98]	0.3%	0.9%
Ernesto 2005	10	10		1.00	[0.69; 1.00]	0.2%	0.9%
Juncarios 2008 Katerina 2012	102	124		0.82	[0.74; 0.89]	2.8%	1.7%
Anders 2013	14	14		1.00	[0.77; 1.00]	0.3%	1.0%
R.corso 2008	15	15		1.00	[0.78; 1.00]	0.3%	1.1%
Mohamed 2021	98	118	— <u>—</u>	0.83	[0.75; 0.89]	2.7%	1.7%
Fixed effect model		656	\$	0.91	[0.89; 0.94]	14.9%	
Heterogeneity: 12 - 769/ -2	- 0.024	10-00		0.94	[0.89; 0.98]		21.3%
neterogeneity: $I = I $ %, τ	= 0.0210	σ, μ < 0.0					
Fixed effect model		4429	6	0.97	[0.97; 0.98]	100.0%	
Random effects model	0.00		<pre></pre>	0.97	[0.95; 0.98]		100.0%
neterogeneity: $I^- = 84\%$, τ^-	= 0.0215	$p, p < 0.0^{\circ}$					

Residual heterogeneity: $I^2 = 82\%$, p < 0.01 0.4 0.5 0.6 0.7 0.8 0.9 1

Figure 3. The Forest plot of clinically success rate after intervention treatment in BCS patients, horizontal lines indicate 95% confidence intervals, square size indicates study specific statistical weigh, and diamond indicates the overall treatment effect with 95% confidence intervals.

In Western countries, where HV extensive thrombosis is more common mostly due to myeloproliferative neoplasm^{92,99}, TIPS placement is used to treat most patients. In Asia, where HV obstruction is mostly due to membranous webs⁸⁴, recanalization (PTA and stenting) is a more common treatment. In this extensive

Study	Events	Total	Proportion	n 95%–Cl	Weight (fixed)	Weight (random)
reatment = Recanalizat	tion					
Yute Fu 2015	3	20	0.1	5 [0.03; 0.38]	0.5%	1.3%
Xe Xu 1996	12	32	0.3	[0.10, 0.25] 3 [0.21: 0.56]	0.7%	1.4%
Pengli zhou 2017	5	47		[0.04; 0.23]	1.1%	1.5%
ang yang 2019	3	33	0.09	9 [0.02; 0.24]	0.8%	1.4%
Yu fei fu 2015	2	17	0.1	2 [0.01; 0.36]	0.4%	1.2%
Jian Xin 2016 Mishra 2003	35	245	0.1	0.11; 0.20]	5.6%	1.0%
Zhongke 2017	19	68	0.2	3 [0.18; 0.40]	1.6%	1.5%
Hong fei 2014	4	43		9 [0.03; 0.22]	1.0%	1.5%
Fianxiao Li 2009	12	92	0.1	3 [0.07; 0.22]	2.1%	1.6%
Srinivas BC 2012	1	12	0.00	3 [0.00; 0.38]	0.3%	1.1%
Delei chena 2019	13	157	- 0.0	3 [0.02; 0.23]	3.6%	1.6%
D.tripathi 2016	11	63	0.1	7 [0.09; 0.29]	1.4%	1.5%
Chun qing 2003	5	106	- 0.0	5 [0.02; 0.11]	2.4%	1.6%
Yonghua Bi 2018	0	70			1.6%	1.5%
Yang fu 2011	1	18	0.0	6 [0.00; 0.20]	0.4%	1.2%
Delei cheng 2018	0	66	- 0.0	0 [0.00; 0.05]	1.5%	1.5%
Chaowen 2019	7	55	0.13	3 [0.05; 0.24]	1.3%	1.5%
Tongguo 2002	1	41		2 [0.00; 0.13]	0.9%	1.5%
aaunong 2013 Yufei Eu 2015	25	168	0.1	5 [0.10; 0.21] 3 [0.10: 0.30]	3.8% 1.4%	1.5%
Fahrettin 2016	3	30	0.10	0 [0.02: 0.27]	0.7%	1.4%
J.F Griffith 1996	5	18	0.22	3 [0.10; 0.53]	0.4%	1.2%
Yan feng Yu 2015	3	14		[0.05; 0.51]	0.3%	1.2%
Kueliang 1996	1	38		3 [0.00; 0.14]	0.9%	1.4%
Ruinua 2013 2 Xu Ding 2010	4	28		1 [0.04; 0.33]	0.6%	1.4%
Yu fei Fu 2015	11	66	0.1	7 [0.09: 0.28]	1.5%	1.5%
Fixed effect model		1866	♦ 0.1	[0.09; 0.12]	42.7%	
Random effects model			♦ 0.1	[0.08; 0.14]		44.4%
Heterogeneity: $I^{a} = 73\%$, τ^{a}	= 0.0111	l, p < 0	1			
reatment = Recanalizat	tion + Tl	PS/DI	5			
Albha N 2010	0	11	0.00	0 [0.00; 0.28]	0.3%	1.1%
Mohammed_2005	5	20		5 [0.09; 0.49] 5 [0.13: 0.65]	0.5%	1.3%
Nitin jagat 2017	15	87	0.1	7 [0.10; 0.27]	2.0%	1.6%
Deepak N 2008	6	37	0.10	6 [0.06; 0.32]	0.9%	1.4%
Delei cheng 2013	6	134	- 0.0	4 [0.02; 0.09]	3.1%	1.6%
Allison 2017 Bozbang 2013	14	25		5 [0.35; 0.76]	0.6%	1.3%
Kiang 2016	8	53		5 [0.07: 0.28]	1.2%	1.5%
Kranti 2016	19	190		0 [0.06; 0.15]	4.3%	1.6%
Yonghua 2018	31	58		3 [0.40; 0.67]	1.3%	1.5%
Darwish 2009	9	64	0.1	1 [0.07; 0.25]	1.5%	1.5%
apen 2005	40	61	0.3	[0.22; 0.41] 3 [0.52: 0.77]	2.3%	1.6%
Kinxin fan 2016	6	58		0 [0.04; 0.21]	1.3%	1.5%
Susanasejio2013	0	70	- 0.00	0 [0.00; 0.05]	1.6%	1.5%
Yonghua Bi 2018	2	39		5 [0.01; 0.17]	0.9%	1.4%
Ahmed 2011	10	442	0.13	9 [0.16; 0.23]	0.6%	1.7%
Yanfeng Cui 2015	28	136		[0.14; 0.28]	3.1%	1.6%
3. boong 2006	4	17	0.24	4 [0.07; 0.50]	0.4%	1.2%
Huixue 2009	0	49	- 0.0	0 [0.00; 0.07]	1.1%	1.5%
Nukund A 2018	/	136	· 0.0	5 [0.02; 0.10]	3.1%	1.6%
Random effects model		1040	0.1	3 [0.11; 0.25]		33.5%
Heterogeneity: $I^2 = 92\%, \tau^2$	= 0.0366	8, <i>p</i> < 0	н			
reatment = TIPS/DIPS						
Martin 2004	19	33	0.58	3 [0.39; 0.75]	0.8%	1.4%
Jlrich 1995	5	12	0.4	2 [0.15; 0.72]	0.3%	1.1%
Tanya M 2014	12	21	0.5	7 [0.34; 0.78]	0.5%	1.3%
Alexeanda2010	11	13		0.55;0.98]	0.3%	1.1%
D.Tripathi 2014	30	67		5 [0.33: 0.57]	1.5%	1.5%
Sonavane 2018	3	42	*	7 [0.01; 0.19]	1.0%	1.5%
Georges 2016	22	53	0.4	2 [0.28; 0.56]	1.2%	1.5%
Snallmar 2017 S. darweb 2007	11	80	0.1	+ [U.U/; 0.23]	1.8%	1.6%
5. bovvat 2008	5	10	0.6	5 [0.17: 0.77]	0.4%	1.1%
Ernesto 2005	0	10	0.0	0 [0.00; 0.31]	0.2%	1.0%
Juncarlos 2008	61	124	0.49	0 [0.40; 0.58]	2.8%	1.6%
Katerina 2013	4	14	0.2	0.08; 0.58]	0.3%	1.2%
Anuers 2013 Ricorso 2008	14 e	14) [0.77; 1.00]) [0.16:0.69]	0.3%	1.2%
Mohamed 2021	48	118	0.4	[0.32: 0.50]	2.7%	1.6%
Fixed effect model		656	♦ 0.3	0 [0.35; 0.43]	15.1%	
Random effects model	0.047		0.4	3 [0.31; 0.55]		22.2%
Heterogeneity: $I^* = 87\%$, τ^*	= 0.0470	, p < 0				
Fixed effect model		4367	٥.1u	6 [0.15; 0.17]	100.0%	100 00/
Heterogeneity: $I^2 = 90\%$. τ^2	= 0.0382	2, p < 0	0.1	0.15; 0.23]		100.0%
2	070/ -		0.0 0.4 0.0 0.0 1			

Residual heterogeneity: $l^2 = 87\%$, p < 0.010 0.2 0.4 0.6 0.8 1

Figure 4. The Forest plot of the re-intervention rate after initial intervention procedures in BCS patients, horizontal lines indicate 95% confidence intervals, square size indicates study specific statistical weigh, and diamond indicates the overall treatment effect with 95% confidence intervals.

meta-analysis, TIPS placement was more used in Western countries than Asian countries, and membranous webs had better outcomes than extensive thrombosis.

Study	Events	Total		Proportion	95%-CI	Weight (fixed)	Weight (random)	
treatment = Recanaliza	tion		1					
Yufe Fu 2015	20	20		1.00	[0.83; 1.00]	0.5%	0.9%	
Ding P. Xu 2018	102	107		0.95	[0.89; 0.98]	2.4%	2.0%	
Ne Au 1996 Popali zbou 2017	47	32		1.00	[0.64; 1.00]	0.7%	1.2%	
Feligii 2000 2017 Fang yang 2019	47	33		1.00	[0.92, 1.00]	0.8%	1.3%	
Yu fei fu 2015	17	17		1.00	[0.80: 1.00]	0.4%	0.8%	
Qian xin 2016	262	263		1.00	[0.98; 1.00]	5.9%	2.4%	
Zhongke 2017	66	68		0.97	[0.90; 1.00]	1.5%	1.8%	
Hong fei 2014	43	43		1.00	[0.92; 1.00]	1.0%	1.4%	
Tianxiao Li 2009	92	92		1.00	[0.96; 1.00]	2.1%	2.0%	
Srinivas BC 2012	12	12		1.00	[0.74; 1.00]	0.3%	0.7%	
Tongqiao 2005 Delei ebeng 2010	41	41		1.00	[0.91; 1.00]	0.9%	1.4%	
Delei cheng 2019 D tripathi 2016	61	63		0.97	[0.98, 1.00]	3.3%	2.2%	
Chun aina 2003	106	106	19	1.00	[0.03, 1.00]	2.4%	2.0%	
Yonghua Bi 2018	70	70		1.00	[0.95; 1.00]	1.6%	1.8%	
P. Xu Ding 2015	92	93		0.99	[0.94; 1.00]	2.1%	2.0%	
Yang fu 2011	18	18		1.00	[0.81; 1.00]	0.4%	0.9%	
Delei cheng 2018	65	66		0.98	[0.92; 1.00]	1.5%	1.7%	
Chaowen 2019	55	55		1.00	[0.94; 1.00]	1.2%	1.6%	
Coubong 2012	41	41		1.00	[0.91; 1.00]	0.9%	1.4%	
Vufei Eu 2015	59	601		0.90	[0.92, 0.96]	3.0%	2.3%	
Fahrettin 2016	30	30		1.00	[0.88; 1.00]	0.7%	1.2%	
J.F Griffith 1996	16	18		0.89	[0.65; 0.99]	0.4%	0.9%	
Yan feng Yu 2015	14	14		1.00	[0.77; 1.00]	0.3%	0.7%	
Xueliang 1996	38	38		1.00	[0.91; 1.00]	0.9%	1.4%	
Ruihua 2013	28	28		1.00	[0.88; 1.00]	0.6%	1.2%	
P.Xu Ding 2010	13	13		1.00	[0.75; 1.00]	0.3%	0.7%	
Yu fei Fu 2015 Fixed offoot model	66	1970		1.00	[0.95; 1.00]	1.5%	1.7%	
Fixed effects model		1879	30	1.00	[0.99; 1.00]	42.1%	44.6%	
Heterogeneity: $l^2 = 15\% \tau^2$	= 0.0007	$7 \cdot p = 0$	3	1.00	[0.33, 1.00]		44.0 /0	
		, ,-	- -					
treatment = Recanaliza	tion + TI	PS/DIP	3					
Albha N 2010	10	11		0.91	[0.59; 1.00]	0.3%	0.6%	
Rachna 2014	24	25		0.96	[0.80; 1.00]	0.6%	1.1%	
Nonammed-2005	13	14		0.93	[0.66; 1.00]	0.3%	1.0%	
Deenak N 2008	35	37		0.95	[0.89, 0.99]	0.8%	1.3%	
Delei chena 2013	133	134	-	0.99	[0.96; 1.00]	3.0%	2.2%	
Allison 2017	24	25		0.96	[0.80; 1.00]	0.6%	1.1%	
Bozhang 2013	18	18		1.00	[0.81; 1.00]	0.4%	0.9%	
Xiang 2016	48	53		0.91	[0.79; 0.97]	1.2%	1.6%	
Kranti 2016	190	190		1.00	[0.98; 1.00]	4.3%	2.3%	
Yonghua 2018	5/	58		0.98	[0.91; 1.00]	1.3%	1.7%	
Warraky 2015	93	101		0.83	[0.71, 0.91]	2.3%	2.0%	
Eapen 2005	57	61		0.93	[0.84: 0.98]	1.4%	1.7%	
Xinxin fan 2016	58	60		0.97	[0.88; 1.00]	1.4%	1.7%	
Susanasejio2013	59	70		0.84	[0.74; 0.92]	1.6%	1.8%	
Yonghua Bi 2018	39	40		0.98	[0.87; 1.00]	0.9%	1.4%	
P. Xu Ding 2019	448	455		0.98	[0.97; 0.99]	10.3%	2.6%	
Ahmed 2011	25	25		1.00	[0.86; 1.00]	0.6%	1.1%	
B boong 2006	137	140		1.00	[0.94; 1.00]	0.4%	0.8%	
B. boong 2000 Huixue 2009	46	49		0.94	[0.83: 0.99]	1.1%	1.5%	
Mukund A 2018	128	136		0.94	[0.89; 0.97]	3.1%	2.2%	
Fixed effect model		1870	¢.	0.98	[0.97; 0.99]	42.4%		
Random effects model			\$	0.97	[0.95; 0.98]		36.0%	
Heterogeneity: $I^{*} = 70\%$, τ	= 0.0076	6, <i>p</i> < 0.	1 8					
treatment = TIPS/DIPS								
Martin 2004	30	33		0.91	[0.76; 0.98]	0.8%	1.3%	
Ulrich 1995	9	12 -	i	0.75	[0.43; 0.95]	0.3%	0.7%	
Tanya M 2014	21	21		1.00	[0.84; 1.00]	0.5%	1.0%	
Alexeanda2010	12	13		0.92	[0.64; 1.00]	0.3%	0.7%	
Kerstin 2016	13	13		1.00	[0.75; 1.00]	0.3%	0.7%	
D. Iripatni 2014	62	67		0.93	[0.83; 0.98]	1.5%	1.8%	
Georges 2016	51	42		0.00	[0.71, 0.95]	1.0%	1.4%	
Shalimar 2017	75	80		0.94	[0.86: 0.98]	1.8%	1.9%	
S .darwsh 2007	13	16	i	0.81	[0.54; 0.96]	0.4%	0.8%	
F. boyvat 2008	11	11		1.00	[0.72; 1.00]	0.3%	0.6%	
Ernesto 2005	10	10		1.00	[0.69; 1.00]	0.2%	0.6%	
Juncarlos 2008	118	124	-=	0.95	[0.90; 0.98]	2.8%	2.1%	
Katerina 2013	14	14		1.00	[0.77; 1.00]	0.3%	0.7%	
Anuers 2013	14	14		1.00	[0.77; 1.00]	0.3%	0.7%	
Mohamed 2021	113	118		0.07	[0.90: 0.90]	2 7%	2.1%	
Fixed effect model	115	656	\$	0.96	[0.93; 0.97]	15.0%		
Random effects model			\diamond	0.95	[0.93; 0.97]		19.4%	
Heterogeneity: $I^2 = 21\%$, τ^2	= 0.0018	B, p = 0.	1					
Eived offect model		440F		0.00	0.08.0.001	100 0%		
Random effects model		-1-100		0.99	[0.97; 0.99]		100.0%	
Heterogeneity: $I^2 = 60\%$, τ^2	² = 0.0060), p < 0.						

Residual heterogeneity: $l^2 = 48\%$, p < 0.01 0.5 0.6 0.7 0.8 0.9 1

Figure 5. The Forest plot of the survival rate at 1 year after initial intervention procedures in BCS patients, horizontal lines indicate 95% confidence intervals, square size indicates study specific statistical weigh, and diamond indicates the overall treatment effect with 95% confidence intervals.

The step-wise therapeutic algorithm of BCS includes medical therapy with anticoagulant drugs and thrombolysis—recanalization with or without stent placement—TIPS/DIPS and liver transplantation^{45,100}. However, due to poor long-term medical therapy outcomes, most of the studies used recanalization with or without stent

Study	Events Total		Proportion	95%-CI	Weight (fixed)	Weight (random)
treatment = Recanaliza	tion					
Ding P. Xu 2018	92 107		0.86	[0.78; 0.92]	2.8%	2.2%
Ke Xu 1996	31 32		0.97	[0.84; 1.00]	0.8%	1.6%
Pengli zhou 2017	47 47		1.00	[0.92; 1.00]	1.2%	1.8%
Fang yang 2019	33 33		1.00	[0.89; 1.00]	0.9%	1.6%
Qian xin 2016	258 263		0.98	[0.96; 0.99]	6.9%	2.4%
Zhongke 2017	64 68		0.94	[0.86; 0.98]	1.8%	2.0%
Hong fei 2014	43 43		1.00	[0.92; 1.00]	1.1%	1.8%
Srinivas BC 2012	12 12		1.00	[0.74; 1.00]	0.3%	1.0%
Tongqiao 2005	41 41		1.00	[0.91; 1.00]	1.1%	1.7%
D.tripathi 2016	56 63		0.89	[0.78; 0.95]	1.7%	2.0%
Chun qing 2003	106 106		1.00	[0.97; 1.00]	2.8%	2.2%
Yonghua Bi 2018	64 70	<u> </u>	0.91	[0.82; 0.97]	1.8%	2.0%
P. Xu Ding 2015	91 93	1	0.98	[0.92; 1.00]	2.4%	2.1%
Doloi obong 2019	60 66		1.00	[0.01, 1.00]	1 70/	1.2%
Chaowon 2019	55 55		1.00	[0.85, 0.98]	1./ 70	2.0%
	JJ JJ 41 41		1.00	[0.94, 1.00]	1.4%	1.5%
Gaubong 2013	139 168		0.83	[0.31, 1.00]	4 4%	2.3%
Yufei Eu 2015	57 60		0.95	[0.86: 0.99]	1.6%	1.9%
Fabrettin 2016	30 30		1 00	[0.88:1.00]	0.8%	1.6%
J F Griffith 1996	14 18		0.78	[0.52: 0.94]	0.5%	1.2%
Xueliang 1996	38 38		1.00	[0.91: 1.00]	1.0%	1.7%
Ruihua 2013	28 28		1.00	[0.88: 1.00]	0.7%	1.5%
Yu fei Fu 2015	66 66		1.00	[0.95; 1.00]	1.7%	2.0%
Fixed effect model	1566	\$	0.97	[0.96: 0.98]	41.0%	
Random effects model		\diamond	0.98	[0.95; 0.99]		43.6%
Heterogeneity: $I^2 = 79\%$, τ^2	$p^2 = 0.0148, p < 0.01$					
treatment = TIPS/DIPS						
Martin 2004	30 33		0.91	[0.76; 0.98]	0.9%	1.6%
Tanya M 2014	18 21		0.86	[0.64; 0.97]	0.6%	1.3%
Alexeanda2010	12 13		0.92	[0.64; 1.00]	0.4%	1.0%
Kerstin 2016	12 13		0.92	[0.64; 1.00]	0.4%	1.0%
D.Tripathi 2014	54 67		0.81	[0.69; 0.89]	1.8%	2.0%
Sonavane 2018	34 42		0.81	[0.66; 0.91]	1.1%	1.8%
Georges 2016	44 53		0.83	[0.70; 0.92]	1.4%	1.9%
Shallmar 2017	72 80		0.90	[0.81; 0.96]	2.1%	2.1%
S Juarwsh 2007	10 10		1.09	[0.41, 0.69]	0.4%	0.0%
luncarlos 2008	108 124		0.87	[0.09, 1.00]	3 20%	0.9%
Katerina 2013	14 14		1.00	[0.00, 0.02]	0.2%	1 1%
Anders 2013	13 14		0.93	[0.66:1.00]	0.4%	1.1%
B corso 2008	13 15		0.87	[0.60; 0.98]	0.4%	1.1%
Mohamed 2021	108 118		0.92	[0.85: 0.96]	3.1%	2.2%
Fixed effect model	633	\diamond	0.89	[0.86: 0.91]	16.7%	
Random effects model		\diamond	0.89	[0.85; 0.92]		22.6%
Heterogeneity: $I^2 = 18\%$, τ^2	² = 0.0013, <i>p</i> = 0.26					
treatment = Recanaliza	tion + TIPS/DIPS					
Rachna 2014	24 25		0.96	[0.80; 1.00]	0.7%	1.5%
Mohammed-2005	13 14		0.93	[0.66; 1.00]	0.4%	1.1%
Nitin jagat 2017	81 87		0.93	[0.86; 0.97]	2.3%	2.1%
Deepak N 2008	35 37		0.95	[0.82; 0.99]	1.0%	1.7%
Allison 2017	20 25		0.80	[0.59; 0.93]	0.7%	1.5%
Boznang 2013	18 18		1.00	[0.81; 1.00]	0.5%	1.2%
Kiang 2016	46 53		0.87	[0.75; 0.95]	1.4%	1.9%
Vonghua 2019	57 59		1.00	[0.98; 1.00]	5.0%	2.3%
Worroky 2015	57 56 02 101		0.90	[0.91, 1.00]	1.5%	1.9%
Fanon 2005	53 61		0.92	[0.85, 0.97]	2.0%	2.2%
Xinxin fan 2016	58 60		0.97	[0.88: 1.00]	1.6%	1.9%
Susanaseijo2013	59 70		0.84	[0.74; 0.92]	1.8%	2.0%
Yonghua Bi 2018	36 40		0.90	[0.76: 0.97]	1.1%	1.7%
P. Xu Ding 2019	415 455		0.91	[0.88; 0.94]	11.8%	2.5%
Yanfeng Cui 2015	131 140		0.94	[0.88; 0.97]	3.7%	2.3%
Huixue 2009	46 49		0.94	[0.83; 0.99]	1.3%	1.8%
Mukund A 2018	128 136		0.94	[0.89; 0.97]	3.5%	2.3%
Fixed effect model	1619	4	0.94	[0.93; 0.95]	42.3%	
Random effects model		\$	0.94	[0.91; 0.96]		33.8%
Heterogeneity: $I^2 = 74\%$, τ^2	² = 0.0086, <i>p</i> < 0.01					
Eived offect model	2010		0.05	10 04. 0 001	100 00/	
Random effects model	3010	2	0.95	[0.93 · 0.96]	100.0%	100 0%
Heterogeneity: $l^2 = 77\%$, τ^2	$p^2 = 0.0127$, $p < 0.01$		0.00	[5.55, 5.55]		

Residual heterogeneity: $I^2 = 72\%$, p < 0.01 0.5 0.6 0.7 0.8 0.9 1

Figure 6. The Forest plot of the survival rate at 5 years after initial intervention procedures in BCS patients, horizontal lines indicate 95% confidence intervals, square size indicates study specific statistical weigh, and diamond indicates the overall treatment effect with 95% confidence intervals.

placement as the first-line treatment for BCS^{14,15,22,26,35,59,80}. Moreover, TIPS was used in circumstances of failed recanalization, refractory ascites, portal hypertension, variceal bleeding, and long segment obstruction or diffused obstruction of the HV^{21,24,41,43,52}. Recanalization is a physiological procedure that maintains the natural blood flow in HV/IVC^{33,36,41}. It can minimize the risk of hepatic encephalopathy, and remains a first-line treatment option for BCS patients^{35,61}. However, TIPS has less portal vein blood perfusion in the liver than recanalization and a high risk of hepatic encephalopathy due to the formation of a blood ammonia level and impaired liver function after shunt placement¹⁹. The secondary patency of recanalization with angioplasty + stent (79% and 92%) was higher than recanalization with only angioplasty (64% and 69%) at 1 and 5 years⁴⁹. The treatment of BCS with an expandable metallic stent was introduced to decreasing the re-stenosis rate after angioplasty¹⁰¹. This study found that most studies adopted recanalization (44.28%) as a first-line treatment because it is a relatively simple and quick procedure. Also, the risk of hepatic encephalopathy after recanalization is lower than TIPS/DIPS. TIPS/DIPS has only been applied as an alternative treatment option for selective cases of BCS, but it may have a high risk of complication after shunt implantation^{49,102}. However, several previous studies have reported the high patency rate and long-term outcome of TIPS/DIPS for BCS^{43,75,103-106}. Liver transplantation is a second surgical option for BCS when a rapidly progressive liver failure occurs before or after TIPS^{107,108}.

In this meta-analysis, we found that the survival of recanalization and TIPS were 99.9% and 94.9% at 1 year and 97.9% and 87.9% at 5 years, respectively. The survival of patients in this study seems comparable to that of a previous meta-analysis Zhang et al.¹⁰⁹, which showed the survival of recanalization and TIPS were 95.9% and 87.3% at 1 year and 88.6% and 72.1% at 5 years, respectively. Tripathi et al.'s⁴⁹ retrospective study showed the survival of recanalization and TIPS were 97% and 88% at 1 year, 89% and 79% at 5 years, and 85% and 73% at 10 years, respectively. Garcia-pagan et al.⁷⁵ reported that the survival of TIPS with liver transplantation at 1and 5 years were 88% and 78%, respectively. Mentha et al.¹¹⁰ reported that survival of liver transplantation for BCS at 1, 5, and 10 years were 76%, 71%, and 68%, respectively. Nonetheless, our meta-analysis results indicate a progressive improvement in survival rate with endovascular therapy for BCS treatment.

Our results show that recanalization therapy had a better prognosis than TIPS therapy. Similarly, the prognosis of recanalization was shown by previous meta-analyses¹⁰⁹. Mukund et al.⁸² reported that BCS patients treated with recanalization have improved biochemical profile and overall outcome relative to DIPS treatment. However, the survival and clinical improvement were similar in both groups, and Tripathi et al.⁴⁹ also reported no significant difference in the results of patients treated with recanalization and TIPS.

Recently, endovascular intervention treatment has emerged as an advanced therapeutic option for BCS patients. The TIPS/DIPS procedures have rapidly replaced the traditional surgical shunt due to minimal invasiveness, less blood loss, low infection rate, quick recovery, shorter hospital stay, and increased long-term survival rate^{9,24}. The technical success rate of TIPS in BCS has been reported to be between 75 and 100%. Shunt dysfunction at 5 years ranges between 40 and 75%, and the survival rate at 1 and 5 years after the initial intervention treatment was 85% and 75%, respectively^{16,24,74,111,112}. It was found that the TIPS/DIPS technical success rate was 98.9%, while shunt dysfunction was 42.9%, and the survival rates at 1 and 5 years were 94.9% and 87.9%, respectively.

The development of new techniques and improvements in radiological intervention has established endovascular intervention therapy as a treatment of choice for BCS patients. This method provides an effective treatment modality for BCS patients and prevents progression to life threatening conditions, such as portal hypertension and other related complications^{47,113}.

In this updated analysis, most of the included study was original articles published after 2010. The survival rates at 1 and 5 years were 97.9% and 94.9%, the success rate of operation was 98.9%, and the re-intervention episode was 18.9%. Similarly, the survival rates of recanalization, combined procedures, and TIPS/DIPS in BCS at 1 and 5 years were 99.9%, 96.9%, and 94.9% and 97.9%, 93.9%, and 87.9%, respectively. Publication bias of technical success (P=0.0335), clinical success (P=0.5567), re-intervention (P=0.08108), the survival rate at 1 year (P=0.01549) and survival rates at 5 years (P=0.8909) were observed. The patients with recanalization treatment had a better prognosis and outcome than the combined procedures and TIPS/DIPS treatment. Additionally, the clinical success rate, shunt dysfunction rate, combined procedures, and obstruction site were analyzed. Overall, comparatively the statistical results are progressively more favorable than the previous study¹⁰⁹.

Despite the latest update on the role of endovascular intervention therapy for BCS, the present study has several limitations: First, studies on endovascular intervention therapy for BCS worldwide are limited. Retrievable articles were available between 1995 and 2019. Most of the relevant studies were published between 2015 and 2019 and only four studies were published before 2000. Second, some articles were excluded during the selection because of a lack of information about re-intervention and long-term survival rates. Third, there was an unequal distribution based on studies conducted in different geographical regions. Most of the study samples were from Asian and European countries; the African and American data were scarce. Also, some studies were excluded due to low study quality.

Conclusion

The systematic review and meta-analysis findings further solidify the role of endovascular intervention treatment in BCS as safe and effective. It maintains high technical and clinical success, and long-term survival rates. The recanalization treatment had a better prognosis and outcome than the combined procedures and TIPS/DIPS treatment. The endovascular intervention procedures are the preferred first-line treatment in selected patients with BCS. However, randomized controlled multidisciplinary centers studies are needed to further evaluation.

Data availability

The data presented in this study are available on request from the corresponding author. The data are not publicly available due to legal restrictions.

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Author contributions

G.M., and X.Z., retrieved of all data and analyzed data, interpretation of data, wrote manuscript and final approval; X.H., and D.J., conception and designed of the study, supervised the work, draft manuscript, critical revision and final approval; G.P., interpretation data and revised manuscript and final approval; Y.L. and S.P., revised manuscript and final approval.

Competing interests

The authors declare no competing interests.

Additional information

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