Percutaneous hepatic vein recanalization in pediatric Budd–Chiari syndrome – 10 years' experience from a tertiary center

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ABSTRACT

Background : **Budd – Chiari syndrome (BCS) due to hepatic venous outflow obstruction is a rare cause of liver disease with dismal outcome, often amenable to catheter intervention.**

Materials and Methods : **This retrospective single-center study analyzed the clinical profile and medium-term outcome of interventional treatment with balloon angioplasty ± stenting in all pediatric BCS over a 10-year period. Clinical, laboratory, imaging, and interventional data were retrieved. Transhepatic (TH) access was utilized in the recent 3 years.**

Results : **We included a total of 27 patients. Acute and subacute BCS comprised 93% of subjects. Ascites was the most common symptom. COVID-19 infection and Takayasu arteritis were two novel etiologies in our study. There was isolated hepatic vein (HV) narrowing in 11 (41%), isolated inferior vena cava obstruction in 4, and combined occlusion in 12 (44%). Intervention was successful in 22 (82%) patients. Stenting was required in 14 (64%) patients and the rest underwent balloon angioplasty. The immediate outcome was better with stenting than balloon (91% vs. 64%). Transhepatic access in 6 patients allowed HV cannulation in all and achieved patency in five patients. Two patients from the balloon group (25%) and 9 from the stent group (64%) are alive with patent veins at a median follow-up of 60 months, indicating a high attrition rate.**

Conclusion : **Catheter interventions restored physiological blood flow in pediatric BCS. TH route improved cannulation of occluded HV compared to other accesses. Immediate and medium-term outcomes were better after stenting with lower rates of reinterventions than balloon angioplasty. Life-long surveillance is required as mortality is high on follow-up.**

Keywords : Budd–Chiari syndrome, portal hypertension, transhepatic access

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INTRODUCTION

Budd–Chiari syndrome (BCS) is characterized by hepatic venous outflow obstruction leading to hepatic venous congestion and back pressure changes ultimately causing cirrhosis and liver cell failure. Obstruction can be at the hepatic veins (HV) and/or at inferior vena cava (IVC) due to luminal thrombosis or rarely extrinsic compression.[1,2] BCS can be acute (<1 month), subacute, or a chronic process (>6 months).[3] The physiological hallmark is portal hypertension causing ascites, hepato-splenomegaly, and varices. Variceal hemorrhage can occur due to rupture of portosystemic collaterals. Encephalopathy and jaundice occur in acute BCS or advanced liver failure.[1]

The etiological factors can be genetic mutation (JAK 2 mutation), antithrombin III deficiency, Protein C and Protein S deficiency, Factor V Leiden mutation, and other thrombophilic states.[2,3] BCS must be considered a possible diagnosis in children with firm hepatomegaly, ascites and altered liver function.[4] Diagnosis is through imaging by ultrasound, computed tomography (CT), or magnetic resonance imaging.[5] The crux of management is to restore HV flows to decompress the portal veins. Medical management alone with anticoagulation has dismal outcomes with <15% survival.[1] Thrombolysis, either local or systemic, may benefit during the acute phase of illness. Creation of surgical portosystemic shunt carries significant morbidity and mortality. Transjugular intrahepatic portosystemic shunt (TIPS) has improved 5-year survival to 71% or more. Percutaneous hepatic or IVC recanalization (HIVR) provides physiological restoration of sinusoidal flow. This remains the first choice of therapy whenever anatomy is feasible. Patients receiving HIVR show improved liver synthetic functions compared to those with TIPS.[6] Liver transplantation remains the final option when all other modalities have failed.

Abundant literature is available on adult BCS describing newer interventional techniques and long-term outcomes, including international guidelines.[6-9] The studies in children are limited, largest ones being four from India and one from China.[3,10-13] The success of interventional therapy seen in the adult population has not been replicated to the same extent in children. Technical skill, small size of patients, and lack of suitable stents with further dilatation potential are the major limiting factors. This article describes the evolution of technique and outcomes of HIVR over a period of 10 years in pediatric BCS from a tertiary care cardiac center. Various scoring systems have been used for prognostication of a child with liver disease.[6,14-16] We have also made an attempt at prognostication of BCS using these scoring systems (see footnote).

MATERIALS AND METHODS

Data of all children with BCS admitted at our center from January 2012 to December 2021 were retrieved. Demographic data, clinical profile, and investigations were tabulated. Ultrasound with Doppler and/or CT was performed. All children with diagnosis of BCS were taken up for percutaneous recanalization of IVC ± HV. Patients were initially optimized by the correction of anemia, coagulopathy, and tapping of ascitic fluid. All procedures were performed with strict aseptic precautions under conscious sedation. Heparin 100 units/kg was administered at the beginning of the procedure. Postprocedure, all children were started on oral anticoagulation overlapping with heparin until therapeutic International Normalized Ratio (INR) of 2–3 was targeted. Oral anticoagulation has been continued indefinitely for all children. Children were followed up monthly for 3 months and then every 6 months. Clinical data, laboratory tests for INR, liver functions, and ultrasound were documented at follow-up.

Institutional Ethical Committee clearance was obtained to conduct the study. Descriptive statistics were applied for the demographic data. All parameters including scoring system were tested for statistical significance by the Mann–Whitney *U* test using SPSS (IBM trial version 28.0.1.0 [142]) (IBM corporation, New York, United States). Receiver Operating Characteristic (ROC) was plotted for statistically significant parameters to elicit desirable cut-offs.

Interventional procedure

Initial angiograms were performed from femoral vein or jugular vein to profile IVC and locate the stump of blocked HVs [Figure 1]. In the initial years of the study, HIVR was performed through femoral and jugular access. From the beginning of 2019, we adopted transhepatic access (THA) for cannulating the HVs. It was performed under ultrasound guidance by an interventional radiologist. The HV with shortest length of narrowing and well visualized lumen was chosen. A 22G long puncture needle was passed through lower intercostal space in anterior or midaxillary line to enter the HV, which was confirmed by free back flow of blood and contrast injection [Figure 1a and Supplementary Video 1]. Only one HV was punctured. Once the vein was entered, a 0.018″ hydrophilic guide wire (Terumo Corporation, Japan) or a 0.014″ coronary wire was inserted to allow the insertion of a 5Fr short sheath. Angiograms were performed to locate the site, length, and severity of stenosis. Using a multipurpose catheter and coronary wire the obstruction was negotiated. Graded dilatation was done with coronary balloons. The right atrium was entered with the catheter and wire combination. A stiffer 0.035″ Teflon wire was introduced. This was

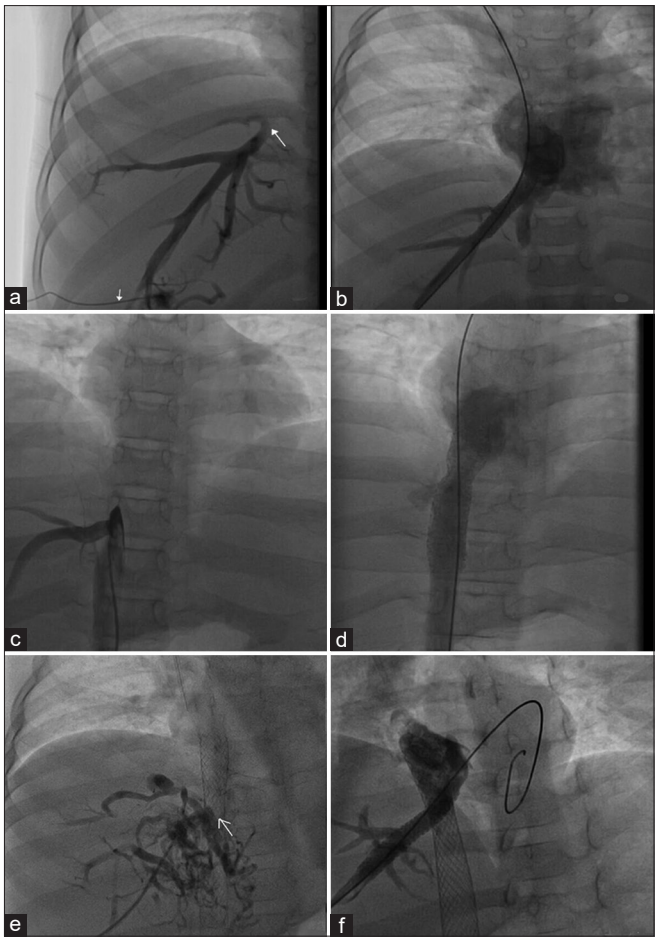

Figure 1: (a) THA with long 22G needle. Position in HV being confirmed with contrast injection. Long arrow showing blocked HV ostia. Short arrow showing cannula in liver (b) Post HV stenting angiogram across showing good flow across the ostia (c) IVC angiogram in AP view showing complete obstruction of hepatic IVC. Arrow pointing to blind end of IVC (d) Post stent deployment across IVC narrowing showing unobstructed flows. Arrow pointing to free reflux into unobstructed HV (e) Trans hepatic access of middle HV post IVC stent with arrow showing blocked ostia of HVs (f and e) Same baby as in e with posthepatic stent angiogram through struts of IVC stent showing good flows. IVC: Inferior vena cava, HV: Hepatic veins, THA: Transhepatic access

snared out from the jugular vein to advance a larger sheath from the neck [Supplementary Video 2]. Further procedures were performed from the jugular sheath. Appropriate sized balloons were used for dilatation. Stenting was performed when residual gradient exceeded 4 mmHg or persistent luminal narrowing was present [Figure 1b and Supplementary Video 3]. Stenting was not preferred in children under 2 years of age. Isolated IVC obstruction with patent HV was treated with stenting in older children [Figure 1c and d]. ATLAS (Bard Peripheral Vascular, USA) or ACCURA (Vascular Concepts Limited, UK) balloons were used for IVC dilatation and self-expanding stents like WALL stent (Boston Scientific, USA) was used for IVC stenting except one where a FORMULA (Cook Medical, USA) stent was used [Figure 1c, d and Supplementary Videos 4, 5]. The most common stent used for HV was OMNILINK (Abbott, Illinois, USA) followed by PROTÉGÉ (Medtronic, Ireland) in one, GENESIS (Cook Corporation, USA) in one and FORMULA stent in 1 patient. Some patients required combined IVC and HV intervention. In younger children balloon dilatation of HV and IVC was done. When stenting was required, the IVC stent was placed by the side of HV stent or HV stent was placed through IVC stent after balloon dilatation of struts [Figure 1e, f and Supplementary Videos 6-8]. While removing the sheath from hepatic puncture site, embolization coils were deployed to close the track and prevent peritoneal bleed [Supplementary Video 9]. Pressure gradient <4 mmHg with the absence of luminal narrowing was considered as a successful outcome. Patients were monitored for 24 h for abdominal girth, hemodynamics, hemoglobin, sensorium, and glucose levels.

RESULTS

Clinical details

A total of 27 patients were admitted with BCS over the last 10 years [Table 1 and Figure 2]. Girls were predominant. The median age was 8 years and duration of symptoms was 30 days. Ten children had acute symptoms of <1 month. Fifteen had subacute BCS of duration <6 months. Abdominal distension due to ascites, pedal edema, and engorged abdominal veins were universally present. Esophageal varices, encephalopathy, and jaundice were present in one third or less patients. Few children had severe anemia requiring transfusion. Liver enzymes were normal in 62%. Hypoalbuminemia was the most common laboratory abnormality in 70%. Prothrombin time was prolonged in most of the patients. The diagnosis was confirmed by ultrasound with Doppler in 38%. The rest underwent CT scan for the diagnosis of BCS. Nonvisualization of HV, narrowing of IVC due to caudate lobe enlargement or luminal thrombus and presence of intrahepatic collaterals were the common findings. Due to logistic and financial constraints, etiology could be established in only 5 out of 27 patients. Two had Protein C deficiency, two had post COVID-19 thrombotic state, and one had Takayasu arteritis.

Isolated HV obstruction (11) and combined HV and IVC (12) obstruction were the most common pattern in nearly 90% of patients. Isolated IVC obstruction was present in four patients. Access was transhepatic (TH) in combination with jugular vein in six patients. THA achieved HV cannulation in all attempted patients, but with jugular and femoral vein access, cannulation was successful in 57% (13/23) of patients. In one patient, patency could not be established due to long segment narrowing despite THA. The average fluoroscopy time for procedure with THA was 20 min, whereas it was 45 min for jugular or femoral access.

Table 1: Profile of pediatric Budd-Chiari syndrome including intervention and outcomes – present study in comparison to literature

	Shukla *et al.*[3]	Sharma *et al.*[10]	Singh *et al.*[11]	Redkar *et al.*[12]	Wang *et al.*[13]	Present
N (Total/Intervened)	36/16	32/26	69/55	35/32	25/24	27/22
HIVR/TIPS	12/3	11/15	43/5	31/1	24/0	22/0
M:F	2.6:1	1:1.5	1.8:1	4:3	16:9	3:4
Acute/Subacute/Chronic %	14/25/61	40/16/44	Chronic	-		38/55/7
Median duration of illness (month)	12	9	8	8		1
Age (years)(range)		1.2	10	2.6	1.3	8 (0.25-18)
Jaundice%(*n*)	33	20	15	-		6 (27)
Encephalopathy (*n*)	0	-	-	-		4
Upper GI bleed(*n*)	8	9	15	3		5
Mean Albumin g/dl	3.16		3.6	-		2.8
Hypoalbuminemia%(*n*)		55%(10)				69%(19)
Mean AST (IU/ml) (SD)	39	80	-	-		74 (57.4)
Mean ALT (IU/ml)(SD)	153	45	-	-		68 (102.1)
INR (mean) ProlongedINR%(*n*)(SD)	1.4	45.5%(10)	1.2-1.8	-		1.7 (0.68)
HV/IVC/IVC + HV %(*n*) involvement	66/8/20	80%/0/5%	74/2/24	85/0/15	60/20/20	40/12/44 (11/4/12)
Balloon%(*n*)	75 (12)(B+S)	75 (9)	16 (7)	100 (27)	80 (20)	50 (11)
Stent%(*n*)		18 (2)	84 (36)	-	20 (4)	64 (14)
TIPS %(*n*)	3	57 (14)	10 (5)	3 (1)	0	0
Success %(*n*) (excluding TIPS)	75%	55%	86%	78%	96%	82%(22)
Mortality%	9	20	29	25.7	0	28
Median follow up (months)(range)	41	44	48		25	60 (3 – 108)
Recurrence (overall)	-	28.5%(TIPS)	18%	18%	29%	37%(B)
Balloon		57%(B)	20%(TIPS)			25%(S)
Stent		33%(S)	27%(S)			
Transhepatic access				1	-	6
LFU		23%	9%	34%	-	26%

HIVR – Hepatic and IVC venous recanalization, TIPS – transjugular intrahepatic portosystemic shunt, GI – hematemesis, INR – international normalised ratio, LFU – lost to follow up, B – balloon, S -stent

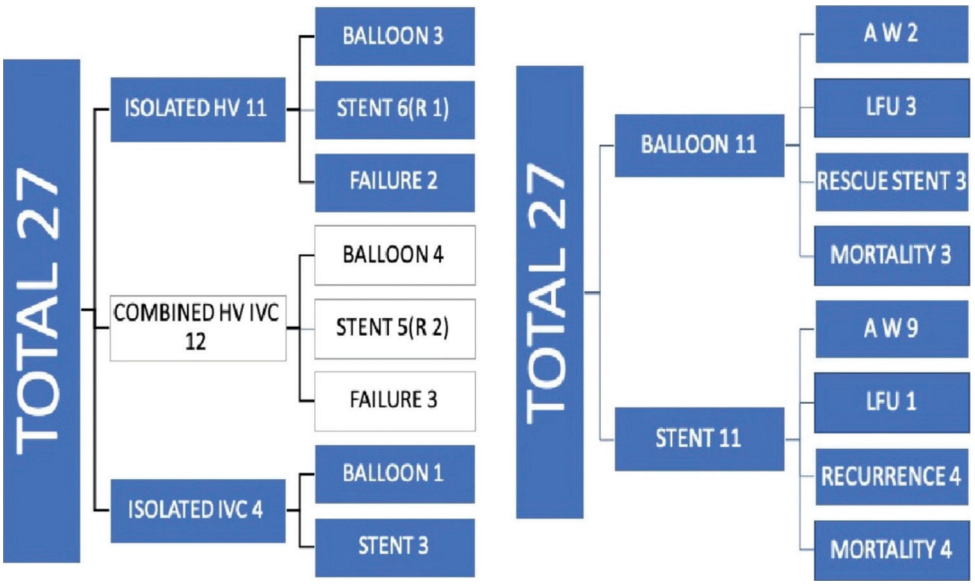

Figure 2: Flow chart showing different subtypes of occlusion, intervention performed and medium term follow up

Immediate success was achieved in 82%. Half the children underwent balloon dilatation as primary procedure. It was the choice, either because of small size of the patient (5 children <2 years) or achievement of good flow (7 children). Three children after balloon dilatation underwent secondary stenting within 72 h in view of increasing ascites and turbulent flows in veins. A total of 64% (14/22) required stenting. Among them isolated HV was in 6, isolated IVC in 3, and combined IVC and HV in 5 children.

Isolated inferior vena cava obstruction

Among total of 4 patients, stenting was done in 3 and one received only balloon dilatation. They had free reflux into the HVs after the IVC intervention demonstrating no obstruction in HV [Arrow in Figure 1d].

Isolated hepatic veins obstruction

In total, 11 patients had isolated HV obstruction. All three HVs showed occlusion of varying lengths. Six were stented, 3 were balloon dilated, and 2 were unsuccessful. In 3 patients, THA was route for intervention. One was a rescue stenting after a failed balloon angioplasty.

Combined hepatic veins + inferior vena cava obstruction

Among 12 patients, simultaneous HV and IVC side by side stenting was done in two patients. One patient had IVC balloon angioplasty followed by HV stenting. In two patients, balloon dilatation of struts of IVC stent was done followed by HV stenting. Four children had balloon dilatation alone of IVC and HV. The procedure was unsuccessful in three patients. In two patients THA was the route. Two were rescue stentings after a failed balloon angioplasty. Details of the stent type and size are provided in the Supplementary Table 1.

Middle HV was the most common vein to be recanalized in 8 children followed by right HV in 7 and left HV in 2 children. Only one child had combined intervention for middle and left HV.

Outcomes (balloon vs. stenting) [Figure 2]

Immediate successful outcome was better with stenting compared to balloon dilatation (91% [10/11]–64% [7/11]). Ten children had good flows post stenting. One child with post multisystem inflammatory syndrome associated with COVID (COVID-MISC) died within 48 h of stenting [Supplementary Videos 10 and 11]. Thrombolysis and repeat procedure were not successful. Postballoon, three children required stenting within 72 h and one baby had suboptimal flows.

Follow-up – mortality

Three children died within 2 months of balloon angioplasty. They had acute BCS and died in spite of having single patent HV. Overall outcomes are poor following balloon angioplasty as only 2 are alive on follow up.

One baby aged 1 year, with suboptimal flows, postballoon angioplasty is on follow-up for about 8 months, with intermittent symptoms of portal hypertension [Figure 1e, f and Supplementary Videos 12, 13]. At a median follow-up of 60 months (range 2–108 months), 64% (9/14) are alive in the stenting group and 25% (2/8) are alive in the balloon group. Mortality was 37% (3/8) in balloon group and 28% (4/14) in stent group [Figure 2].

Recurrence

Among the 14 children undergoing stenting, four children (28%) had restenosis at 1 month, 1 year, 2-, and 4-years postprocedure. Three children with recurrence did not reach our facility for treatment. They succumbed at home to recurrence of similar symptoms. Restenosis was picked in one child during routine follow-up at 4 years and responded to repeat balloon dilatation of stent.

Outcomes with respect to patho-anatomy

Children with IVC obstruction had no recurrence. One child was lost to follow-up and three were alive and well. Both groups of isolated HV and combined HV-IVC obstruction have four children alive and well on follow-up in each group. Four children have died in the combined obstruction group with one lost to follow-up. Three children have died and two are lost to follow-up in isolated HV obstruction group [Figure 2].

Procedure was unsuccessful in 18% (5/27). All of them were placed on medical management. Two succumbed and one is alive with 3 years follow-up with intermittent symptoms. Two are lost to follow-up.

Correlation of outcomes with the different clinical scores

We analyzed the clinical variables and the four scoring systems to compare the survivors and fatal outcome [Table 2]. Symptom duration and Rotterdam scoring system were found to be statistically significant for predicting poor outcomes. Symptom duration of <26 days was suggestive of poor prognosis with sensitivity of 90% and specificity of 75%. Rotterdam score ROC had AUC of 0.8. The score cutoff of 17.52 had a sensitivity of 81% and specificity of 75% to predict the poor outcomes.

Table 2: Comparison of children with good outcomes versus those who succumbed

	Alive and well (*n*=11)	Mortality (*n*=8)	*P*
Age (years)	10	7	0.263
Duration of symptoms (days), mean	**90**	**14.25**	**0.007**
Total protein (g/dL), mean	6.06	5.75	0.272
Albumin (g/dL), mean	3.04	2.47	0.152
Bilirubin (mg/dL), mean	1.26	1.85	0.351
Encephalopathy mean	9%	37%	0.152
AST (IU/mL), mean	63	114	0.091
ALT (IU/mL), mean	42	124	0.129
INR mean	1.56	1.68	0.717
PELD score, mean	11.08	15.27	0.492
Child score, mean	9.18	10.27	0.062
Zietoun score, mean	5.21	5.44	0.238
Rotterdam score, mean	**16.8**	**20.21**	**0.020**

Symptom duration, laboratory parameters, and different scoring systems are the variables analyzed for the possibility of a statistical association for poor outcomes. Those which were statistically significant are in bold fonts. PELD score: 0.436 (Age [<1 year]) – 0.687 × Log_e (albumin g/dL) + 0.480 × Log_e (total bilirubin mg/dL) + 1.87 × Log_e (INR) + 0.667 (growth failure [<–2 SD])[2], Rotterdam score: 1.27×encephalopathy + 1.04 × ascites + 0.72 × prothrombin time + 0.004 × bilirubin (mg/dL)[3], Zeitoun index: (Ascites score × 0.75) + (Pugh score × 0.28) + (age × 0.037) + (creatinine × 0.0036)[5], CPT score[6], (bilirubin, albumin, ascites, encephalopathy, PT), MELD score: 11.2 × ln (INR) + 9.57 × ln (creatinine [mg/dL]) + 3.78 × ln (bilirubin [mg/dL]) × 6.43)[7]. PELD: Pediatric end-stage liver disease, CPT: Child–Pugh-Turcotte, MELD: Model for end-stage liver disease, INR: International Normalized Ratio, ALT: Alanine transaminase, AST: Aspartate transaminase, SD: Standard deviation

DISCUSSION

This is a large single-center retrospective case series of pediatric BCS managed by transcatheter recanalization of the occluded veins. Shukla *et al.* and Singh *et al.* have published series of 16 children (<10 years) and 43 children, respectively, treated with percutaneous intervention.[3,11] Redkar *et al.* and Wang *et al.* published series of 26 and 25 children, respectively, treated with balloon angioplasty alone.[12,13] Our study population differed significantly from other studies [Table 1]. Most of our cases were acute or subacute BCS (84%) unlike other studies where they constituted <50%. Hence, we had low median duration of illness of 1 month as compared to 9 months in other studies. Acute onset of clinical manifestations probably suggests rapid widespread thrombosis. Upper GI bleeding was higher in the series by Dilawari *et al.* and Kathuria *et al.*[2,17] as compared to our series, probably due to high proportion acute BCS patients, with insufficient time for opening of portosystemic collaterals. Higher incidence of encephalopathy, jaundice, lower mean albumin, and higher mean INR in our study suggest a more severe liver cell injury.

We had a higher percentage of combined HV-IVC obstruction. Extensive thrombosis might be a reason for this occurrence. TIPS, although a bridge to transplant has the risk of hepatic encephalopathy, inadequately restores the synthetic functions, and risk of hepatocellular carcinoma remains in the long run. TIPS is not practiced at our institute. HV recanalization which restores the physiological flow can overcome these drawbacks and should be undertaken as the first choice of therapy.[6] It has shown good outcomes in most of the adult studies.

Ultrasound-guided THA provides direct entry to HVs and is useful in short-and medium-segment stenosis of HVs.[18-20] Acute angulation of HV entry into IVC makes femoral or jugular access less suitable. IVC obstruction or compression due to caudate lobe hypertrophy also makes entry into HV technically difficult. THA for HIVR has been extensively used in adult patients with good success rates of up to 91%.[8,9,18,21,22]

Success of HIVR in the literature varies from 40% to 86% in various pediatric series. THA has not gained popularity in the management of pediatric BCS. TH route was used in one patient in Redkar *et al.* series.[12] They had overall technical success in 75%. This access has been used as last resort in some series. We successfully employed this route in our patient population. Access to HV was possible in all patients in whom it was attempted. The reported rate of complications in literature has been <5%. Hepatic capsule rupture, hemopericardium, and hepatic artery pseudoaneurysm have been some of them. Prior ascitic tapping and adequate embolization of exit track reduce the rate of these complications. Six patients in our study were intervened using this route. We did not have any serious complications related to THA. It also reduces the fluoroscopy time of the procedure by almost half.

Immediate outcomes are better with stenting than angioplasty. Intermediate duration patency is also superior with stenting, both in our study and literature. Overall success of interventional therapy (HIVR) including angioplasty and stenting was comparable with other studies.[14] Recurrence with balloon was significant in other studies when compared to stenting, which was replicated in our study. Single patent HV may not decompress the liver adequately in acute BCS as venous collaterals have not formed. Reduced recurrence risk with stenting has also been demonstrated in adult studies.[9] Availability of appropriate stents with potential for expansion to adult size is a major limiting factor in interventional therapy in children. It could be resolved with advances in design and technology in the coming future. Stent patency has been shown to be 82% at 5 years by Singh *et al.*[11] We have a patency rate of 64% at 5 years but restenosis patients did not come for timely repeat intervention which may be the reason for low patency rates.

Two new etiologies hitherto undescribed were found in our study – post-COVID MISC in two children and procoagulant state of aorto-arteritis (Takayasu arteritis) in one child.

Acute BCS with symptoms of <1 month and Rotterdam score more than 17.5 predicted a poor prognosis in our study. Such patients probably require watchful anticoagulation and more frequent follow up. Of the various scores tested in the study, CHILD score predicts survival following portosystemic shunt surgery.[14] Model for end stage liver disease/Pediatric end stage liver disease (PELD) is used to prioritize patients for liver transplant.[15] Zeiuton score is used to predict the survival in adult BCS.[16] Rotterdam score predicted 3-month mortality in adult BCS following medical or nonsurgical interventional therapy.[6] There may be many reasons why only Rotterdam score predicted outcomes in our study. The PELD score did not include key indicators like ascites and encephalopathy that reflect the severity of HV occlusion. The drawback with Child-Pugh score was presence of discrete ranges for prothrombin time, albumin, and bilirubin rather than continuous variables. Zeiuton score was an extension of Child Pugh score. Rotterdam score included ascites and encephalopathy as a significant contributor and covered the synthetic and excretory function of liver, mirrored the severity of portal hypertension and hence probably predicted the outcomes.

CONCLUSIONS

Percutaneous recanalization of HVs provides a physiological pathway for hepatic venous flow and should

be the first choice of treatment. It has an acceptable success rate, a lesser cost with reasonable long-term patency. Immediate-and short-term patency is better with stenting than angioplasty alone. Ultrasound-guided THA provides direct entry to HVs increasing the success of recanalization. Lifelong surveillance and anticoagulation are required.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Wanless IR. Budd-Chiari syndrome. In: Xingshun QI, editor. Pathology of Budd-Chiari Syndrome and Hepatic Vein Obstruction. Budd-Chiari Syndrome. 1st ed. Singapore: Springer Publishers; 2020. p. 27-38.
2. Kathuria R, Srivastava A, Yachha SK, Poddar U, Baijal SS. Budd-Chiari syndrome in children: Clinical features, percutaneous radiological intervention, and outcome. Eur J Gastroenterol Hepatol 2014;26:1030-8.
3. Shukla A, Bhatt P, Gupta DK, Modi T, Patel J, Gupte A, *et al.* Budd-Chiari syndrome has different presentations and disease severity during adolescence. Hepatol Int 2018;12:560-6.
4. Gentil-Kocher S, Bernard O, Brunelle F, Hadchouel M, Maillard JN, Valayer J, *et al.* Budd-Chiari syndrome in children: Report of 22 cases. J Pediatr 1988;113:30-8.
5. Van Wettere M, Bruno O, Rautou PE, Vilgrain V, Ronot M. Diagnosis of Budd-Chiari syndrome. Abdom Radiol (NY) 2018;43:1896-907.
6. Mukund A, Mittal K, Mondal A, Sarin SK. Anatomic recanalization of hepatic vein and inferior vena cava versus direct intrahepatic portosystemic shunt creation in Budd-Chiari syndrome: Overall outcome and midterm transplant-free survival. J Vasc Interv Radiol 2018;29:790-9.
7. Han G, Qi X, Zhang W, He C, Yin Z, Wang J, *et al.* Percutaneous recanalization for Budd-Chiari syndrome: An 11-year retrospective study on patency and survival in 177 Chinese patients from a single Center. Radiology 2013;266:657-67.
8. Tripathi D, Sunderraj L, Vemala V, Mehrzad H, Zia Z, Mangat K, *et al.* Long-term outcomes following percutaneous hepatic vein recanalization for Budd-Chiari syndrome. Liver Int 2017;37:111-20.
9. Shukla A, Shreshtha A, Mukund A, Bihari C, Eapen CE, Han G, *et al.* Budd-Chiari syndrome: Consensus guidance of the Asian Pacific Association for the study of the liver (APASL). Hepatol Int 2021;15:531-67.
10. Sharma VK, Ranade PR, Marar S, Nabi F, Nagral A. Long-term clinical outcome of Budd-Chiari syndrome in children after radiological intervention. Eur J Gastroenterol Hepatol 2016;28:567-75.
11. Singh SK, Sen Sarma M, Yadav R, Kumar S, Prasad R, Yachha SK, *et al.* Prognostic scoring systems and outcome of endovascular radiological intervention of chronic Budd-Chiari syndrome in children. Liver Int 2018;38:1308-15.
12. Redkar R, Raj V, Tewari S, Sharma R, Joshi S. Balloon venoplasty as primary modality of treatment in children with Budd-Chiari syndrome. J Pediatr Surg 2020;55:2183-6.
13. Wang L, Zu MH, Gu YM, Xu H, Zhang QQ, Wei N, *et al.* Budd-Chiari syndrome in children and adolescents: Therapeutic radiological intervention. Zhonghua Er Ke Za Zhi 2013;51:590-4.
14. Peng Y, Qi X, Guo X. Child-Pugh versus MELD score for the assessment of prognosis in liver cirrhosis: A systematic review and meta-analysis of observational studies. Medicine (Baltimore) 2016;95:e2877.
15. Chang CH, Bryce CL, Shneider BL, Yabes JG, Ren Y, Zenarosa GL, *et al.* Accuracy of the pediatric end-stage liver disease score in estimating pretransplant mortality among pediatric liver transplant candidates. JAMA Pediatr 2018;172:1070-7.
16. Zeitoun G, Escolano S, Hadengue A, Azar N, El Younsi M, Mallet A, *et al.* Outcome of Budd-Chiari syndrome: A multivariate analysis of factors related to survival including surgical portosystemic shunting. Hepatology 1999;30:84-9.
17. Dilawari JB, Bambery P, Chawla Y, Kaur U, Bhusnurmath SR, Malhotra HS, *et al.* Hepatic outflow obstruction (Budd-Chiari syndrome). Experience with 177 patients and a review of the literature. Medicine (Baltimore) 1994;73:21-36.
18. Chunqing Z, Lina F, Guoquan Z, Lin X, Zhaohai W, Tao J, *et al.* Ultrasonically guided percutaneous transhepatic hepatic vein stent placement for Budd-Chiari syndrome. J Vasc Interv Radiol 1999;10:933-40.
19. Bansal V, Gupta P, Sinha S, Dhaka N, Kalra N, Vijayvergiya R, *et al.* Budd-Chiari syndrome: Imaging review. Br J Radiol 2018;91:20180441.
20. Madhusudhan KS, Sharma S. Ultrasonography in pediatric Budd-Chiari syndrome. Pediatr Radiol 2020;50:1768-80.
21. Wang Q, Han G. Image-guided treatment of Budd-Chiari syndrome: A giant leap from the past, a small step towards the future. Abdom Radiol (NY) 2018;43:1908-19.
22. Li T, Zhai S, Pang Z, Ma X, Cao H, Bai W, *et al.* Feasibility and midterm outcomes of percutaneous transhepatic balloon angioplasty for symptomatic Budd-Chiari syndrome secondary to hepatic venous obstruction. J Vasc Surg 2009;50:1079-84.

Supplementary Table 1: Patient age, site of obstruction, and intervention done

Patient	Age (years)	HV/IVC	Success/failure	Balloon/stent	HV stent	IVC stent
1	0.9	IVC/HV	Success	Balloon - RHV	-	
2	4	HV	Failure	-	-	
3	10	IVC/HV	Success	Stent - MHV	7×39 OMNI MHV	16×30 WALL
4	6	HV	Success	Stent - RHV	9×59 OMNI	
5	14	HV	Failure	-	-	
6	1	IVC	Success	Balloon	-	
7	8	IVC/HV	Success	Balloon - MHV	-	
8	14	HV	Success	Stent - MHV	8×39, 9×39 OMNI	
9	13	IVC	Success	Stent		10×60 FORMULA
10	3	HV	Success	Stent - LHV	9×30 FORMULA	
11	1.8	HV	Success	Balloon - RHV	-	
12	0.8	HV	Success	Balloon - RHV	-	
13	14	IVC/HV	Failure	-	-	
14	16	IVC/HV	Success	Stent - MHV	9×29 OMNI	14×50 WALL
15	3	HV	Success	Stent - RHV	10×30 EV3	
16	12	IVC/HV	Success	Stent - RHV	9×39 OMNI	
17	7	HV	Success	Balloon - LHV		
18	1	IVC/HV	Success	Balloon - MHV RHV	-	
19	9	IVC/HV	Failure	-	-	
20	8	IVC/HV	Success	Balloon/stent - MHV	9×59 OMNI	B
21	9	HV	Success	Stent - MHV	8×39 OMNI	
22	16	IVC/HV	Success	Stent - RHV	8×39 OMNI	20×50 WALL
23	18	IVC/HV	Success	Stent - MHV	8×18 GENESIS	20×50 WALL
24	1.5	IVC/HV	Failure	-	-	
25	8	IVC/HV	Success	Balloon - RHV		
26	18	IVC	Success	Stent		14×50 WALL
27	16	IVC	Success	Stent		16×60 WALL

IVC: Inferior vena cava, HV: Hepatic vein, RHV: Right hepatic vein, MHV: Middle hepatic vein, LHV: Left hepatic vein