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Changes in arterial oxygenation after portal decompression in Budd–Chiari syndrome patients with hepatopulmonary syndrome

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Abstract

Objectives To evaluate the changes in arterial oxygenation after portal decompression in Budd–Chiari syndrome (BCS) patients with hepatopulmonary syndrome (HPS).

Methods From June 2014 to June 2015, all patients with BCS who underwent balloon angioplasty or transjugular intrahepatic portosystemic shunt (TIPS) creation at our institution were eligible for inclusion in this study. Arterial blood gas analysis was performed with the patient in an upright position and breathing room air at 2–3 days and 1 and 3 months after the procedure.

Results Eleven patients with HPS and 14 patients without HPS were included in this study. The procedure was technically successful in 24 patients. One patient with HPS had technically unsuccessful TIPS creation. Reobstruction or TIPS dysfunction was not detected in any patient within 3 months after the procedure. For patients with HPS, the alveolar–arterial oxygen gradient ($A\text{-}aO_2$) remained comparable to baseline 2–3 days after the procedure (-3.2 ± 11.9 mmHg; $p = .412$), significantly improved 1 month after the procedure (-11.7 ± 6.4 mmHg; $p < .001$), and then returned to baseline 3 months after the procedure (-1.3 ± 12.5 mmHg; $p = .757$). For patients without HPS, the $A\text{-}aO_2$ remained comparable to baseline at all three time points after the procedure (+1.4 ± 8.3 mmHg, +3.5 ± 8.1 mmHg, and +1.3 ± 8.2 mmHg; $p = .543$, $p = .137$, and $p = .565$).

Conclusions Arterial oxygenation transiently improves after portal decompression in BCS patients with HPS.

Key Points

- *Intrapulmonary vascular dilation and hepatopulmonary syndrome are common in patients with Budd–Chiari syndrome.*
- *Arterial oxygenation transiently improves after portal decompression in Budd–Chiari syndrome patients with hepatopulmonary syndrome.*

Keywords Hypertension, portal · Liver cirrhosis · Angioplasty, balloon · Portosystemic shunt, transjugular intrahepatic

Abbreviations

$A\text{-}aO_2$	Alveolar–arterial oxygen gradient
ABG	Arterial blood gas
BCS	Budd–Chiari syndrome
CEE	Contrast-enhanced echocardiography
FEV_1	Forced expiratory volume in 1 s
FVC	Forced vital capacity
HPS	Hepatopulmonary syndrome
HV	Hepatic veins
IPVD	Intrapulmonary vascular dilation
IVC	Inferior vena cava
LT	Liver transplantation
$PaCO_2$	Arterial partial pressure of carbon dioxide
PaO_2	Arterial partial pressure of oxygen
TIPS	Transjugular intrahepatic portosystemic shunt

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Introduction

Budd–Chiari syndrome (BCS) is a rare condition characterized by a blocked hepatic venous outflow tract at various levels from small hepatic veins (HV) to the inferior vena cava

(IVC) [1]. Clinical manifestations range widely from asymptomatic to fulminant liver failure [2]. Management follows a stepwise approach, with medical therapy as the first-line treatment, balloon angioplasty of and/or stent placement in the obstructed HV and/or IVC as the second-line treatment, transjugular intrahepatic portosystemic shunt (TIPS) creation as the third-line treatment, and liver transplantation (LT) as the last-line treatment [1, 3].

Hepatopulmonary syndrome (HPS) is a pulmonary complication of liver disease and/or portal hypertension [4]. It is characterized by a defect in arterial oxygenation caused by ventilation–perfusion mismatch resulting from increased capillary flow at the level of the alveoli due to intrapulmonary vascular dilation (IPVD), which includes diffuse or localized dilation of capillaries and, less commonly, arteriovenous communications [4]. Therefore, the diagnosis of HPS requires, in the setting of liver disease and/or portal hypertension, the demonstration of IPVD (i.e., right to left shunting) and arterial oxygenation defects (i.e., increased alveolar–arterial oxygen gradient (A-aO2) or decreased arterial partial pressure of oxygen (PaO_2)), preferably by contrast-enhanced echocardiography (CEE) and arterial blood gas (ABG) analysis, respectively [4]. HPS occurs mostly in cirrhotic patients (12–32%) but can also occur in patients with pre- and post-hepatic portal hypertension (2–28%) [5–7]. It is associated with detrimental effects on the patient's functional status, quality of life, and survival [8]. However, the treatment options for HPS are extremely limited, as only LT has been established to be effective [9, 10].

HPS occurs in a substantial portion (28%) of patients with BCS [11]. Balloon angioplasty has been shown to be able to reverse HPS in patients with BCS [11]. The mechanism is unknown but may be related to portal decompression. This may also explain the favorable outcomes of TIPS creation for HPS in patients with cirrhosis and idiopathic portal hypertension [12]. The purpose of this study was to evaluate changes in arterial oxygenation after portal decompression in BCS patients with HPS.

Materials and methods

Study design

From June 2014 to June 2015, all patients with BCS undergoing balloon angioplasty and TIPS creation at our institution were eligible for inclusion in this study. The exclusion criteria were age < 18 years, intrinsic cardiopulmonary disease (i.e., intracardiac shunting, QT interval > 50% of the RR interval, forced expiratory volume in 1 s (FEV_1) < 70% of the predicted value, forced vital capacity (FVC) < 70% of the predicted value, FEV_1/FVC < 70% of the predicted value, and pulmonary artery systolic pressure > 35 mmHg), active infection, documented malignancy, pregnancy, or previous shunt therapy. Before the procedure, all patients underwent ABG analysis while in an upright position and breathing room air, CEE, pulmonary function tests, and plain chest radiographs. The diagnosis of HPS was established by an elevated A-aO_2 (i.e., ≥ 15 mmHg (≥ 20 mmHg if age ≥ 65 years)) and a positive CEE result [4, 13, 14]. The severity of HPS was classified as very severe (PaO_2 < 50 mmHg), severe (PaO_2 50–59 mmHg), moderate (PaO_2 60–79 mmHg), or mild (PaO_2 ≥ 80 mmHg) [4, 13, 14]. This prospective study was approved by our institutional review board. All patients provided written informed consent for study participation.

CEE procedure

The technique used to perform CEE has been described in detail elsewhere [15]. Briefly, 10 mL of agitated saline was injected through an intravenous line placed in the right upper extremity, and transthoracic two-dimensional echocardiography in the parasternal four-chamber view was used to detect the appearance of microbubbles. The result was regarded as positive if any microbubbles appeared in the left heart chambers 3–6 heartbeats after they initially appeared in the right heart chambers after three injections.

Balloon angioplasty and TIPS creation procedure

For patients with short-segment or membranous obstruction of the HV, IVC, or both, balloon angioplasty was performed. The occluded segment was traversed with a hydrophilic guidewire (Terumo) and a multipurpose catheter (Cordis) via transjugular access. If the occluded segment could not be traversed, a Rösch-Uchida liver access set (Cook) was introduced, and the HV or IVC distal to the occluded segment was punctured. When necessary, a pigtail catheter (Cordis) was placed in the IVC distal to the occluded segment via femoral access to serve as a target for puncture. After the occluded segment was traversed, a 10–20-mm-diameter balloon catheter (Cordis) was introduced, and the balloon was inflated until the circumferential indentation caused by the obstruction disappeared from the balloon contour for 1–2 min. For patients with a long-segment obstruction of the HV with or without IVC obstruction, a TIPS was created. A Rösch-Uchida liver access set (Cook) was introduced into the stump of the HV or the IVC via transjugular access, and the portal vein was punctured. The parenchymal tract was dilated with an 8-mm-diameter balloon catheter (Cordis), and a 10-mm-diameter Fluency Plus stent graft (Bard) was placed to cover the tract. If the portosystemic pressure gradient (PPG) remained > 13 mmHg after TIPS creation, the shunt tract was dilated with a 10-mm-diameter balloon catheter (Cordis).

Follow-up

After the procedure, all patients received long-term warfarin therapy with an initial dose of 1.25 mg that was adjusted in a stepwise manner to achieve the target international normalized ratio of 2–3. ABG analysis was performed with the patient in an upright position and breathing room air at 2–3 days and 1 and 3 months after the procedure. Doppler ultrasound examination, with or without contrast medium, was performed in all patients at 1 and 3 months, or whenever clinically necessary, after the procedure to assess the patency of the HV and IVC or the TIPS. Patients with signs of reobstruction or TIPS dysfunction on Doppler ultrasound underwent angiography with pressure measurements for confirmation.

Statistical analysis

Continuous variables are presented as the mean ± standard deviation and were compared using Student's *t* test. Categorical variables are presented as frequencies (percentages) and were compared using the chi-square test or Fisher's exact test. A two-sided *p* value < .05 was considered indicative of a statistically significant difference. Bonferroni correction was used for multiple comparisons as appropriate. Statistical analyses were performed using SPSS software (version 19.0; SPSS).

Results

Patient population

A total of 28 patients were considered eligible for inclusion in this study (Fig. 1). Among these 28 patients, three (10.7%) were excluded due to the following reasons: inadequate echocardiographic window ($n = 1$), inability to undergo pulmonary function tests ($n = 1$), and documented malignancy ($n = 1$). Of the remaining 25 patients, the CEE results were positive in 14 (56.0%), and 11 (78.6%) of these patients had an elevated A-aO_2. These 11 (44.0%) patients were considered to have HPS, including five (45.5%) with moderate HPS and six (54.5%) with mild HPS. The baseline characteristics of the patients are summarized in Table 1.

Of the 11 patients with HPS, four had short-segment or membranous obstruction of the HV ($n = 2$), IVC ($n = 1$), or both ($n = 1$) and underwent HV ($n = 2$) or IVC ($n = 2$) balloon angioplasty. The remaining seven patients with HPS had long-segment obstruction of the HV with ($n = 5$) or without ($n = 2$) IVC obstruction and underwent TIPS creation. For the 14 patients without HPS, ten had short-segment or membranous obstruction of the HV ($n = 2$), IVC ($n = 5$), or both ($n = 3$) and underwent HV ($n = 2$) or IVC ($n = 2$) balloon angioplasty. The remaining four patients without HPS had long-segment obstruction of the HV with IVC obstruction and underwent TIPS creation.

Procedure details

The procedure was technically successful in 24 patients (Table 2). One patient with HPS had technically unsuccessful TIPS creation due to a very short IVC stump. The subsequent results exclude this technically unsuccessful case. The preprocedural pressure gradient across the obstructed HV or IVC was not significantly different between patients with and without HPS who underwent balloon angioplasty (19.3 ± 3.6 mmHg vs. 17.9 ± 5.6 mmHg; $p = .669$). The preprocedural PPG was also not significantly different between patients with and without HPS who underwent TIPS creation (21.2 ± 4.6 mmHg vs. 21.0 ± 3.2 mmHg; $p = .951$). After balloon angioplasty, the pressure gradient across the obstructed HV or IVC was ≤ 5 mmHg in four (100%) and nine (90.0%) patients with and without HPS, respectively. After TIPS creation, the PPG was ≤ 12 mmHg in four (66.7%) and four (100%) patients with and without HPS, respectively.

No patients were lost to follow-up within 3 months after the procedure. Doppler ultrasound examination did

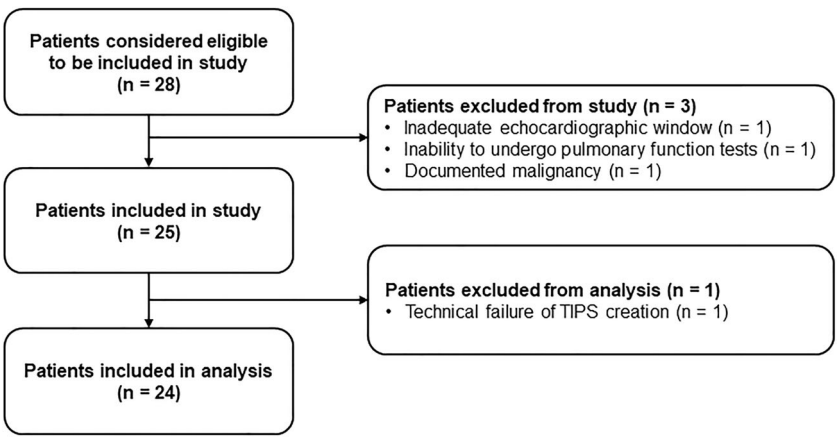

Fig. 1 Flow chart of patient selection

Table 1 Baseline characteristics of the patients

	HPS ($n = 11$)	Non-HPS ($n = 14$)	p value
Age (years)	51.7 ± 17.2	47.2 ± 11.5	.465
Males	6 (54.5)	9 (64.3)	.697
Primary BCS	9 (81.8)	12 (85.7)	> .999
Site of obstruction			.172
HV	5 (45.5)	2 (14.3)	
IVC	1 (9.1)	5 (35.7)	
HV and IVC	5 (45.5)	7 (50.0)	
Liver cirrhosis	5 (45.5)	3 (21.4)	.389
Rotterdam score	.7 ± .5	.9 ± .7	.436
BCS TIPS PI	5.2 ± 1.2	4.9 ± 1.1	.487
MELD score	12.0 ± 3.1	10.4 ± 1.5	.145
Ascites	6 (54.5)	9 (64.3)	.697
Bilirubin (mg/dL)	1.5 ± .8	1.6 ± .5	.584
Albumin (g/L)	32.3 ± 6.7	39.5 ± 3.1	.006
INR	1.4 ± .3	1.2 ± .1	.253
Creatinine (mg/dL)	.9 ± .5	.8 ± .2	.742
Hepatic encephalopathy	0 (0)	1(7.1)	> .999
Variceal bleeding	3 (27.3)	5 (35.7)	> .999
Active alcohol drinker	0 (0)	0 (0)	> .999
Active tobacco smoker	2 (18.2)	2 (14.3)	> .999
Dyspnea	3 (27.3)	2 (14.3)	.625
Cyanosis	1 (9.1)	0 (0)	.440
Digital clubbing	1 (9.1)	0 (0)	.440
Spider angioma	3 (27.3)	1 (7.1)	.288
SaO_2 (%)	96.7 ± 1.7	98.3 ± 1.0	.009
CEE positive	11 (100)	4 (28.6)	.001
ABG analysis			
AaO_2 (mmHg)	27.2 ± 6.3	9.3 ± 11.6	< .001
PaO_2 (mmHg)	81.4 ± 10.2	95.0 ± 11.8	.006
$PaCO_2$ (mmHg)	33.3 ± 6.5	36.8 ± 4.8	.129
Pulmonary function tests			
TLC (% predicted)	93.2 ± 11.7	98.7 ± 10.7	.236
FEV_1 (% predicted)	89.6 ± 19.0	103.4 ± 14.5	.051
FVC (% predicted)	91.6 ± 13.3	101.9 ± 15.3	.090
FEV_1/FVC (% predicted)	80.6 ± 6.9	84.0 ± 6.2	.212
DLCO (mL/min/mmHg)	15.1 ± 4.0	20.0 ± 3.3	.003
DLCO (% predicted)	73.8 ± 25.6	84.5 ± 14.6	.201
Plain chest radiography			
Interstitial markings	1 (9.1)	0 (0)	.440
Plural effusion	3 (27.3)	0 (0)	.072
Portal decompression procedure			.116
Balloon angioplasty	4 (36.4)	10 (71.4)	
TIPS creation	7 (63.6)	4 (28.6)	

HPS hepatopulmonary syndrome, *BCS* Budd–Chiari syndrome, *HV* hepatic veins, *IVC* inferior vena cava, *TIPS* transjugular intrahepatic portosystemic shunt, *PI* prognostic index, *MELD* Model For End-Stage Liver Disease, *INR* international normalized ratio, SaO_2 arterial oxygen saturation, *CEE* contrast-enhanced echocardiography, *ABG* arterial blood gas, AaO_2 alveolar–arterial oxygen gradient, PaO_2 partial arterial pressure of oxygen, $PaCO_2$ partial arterial pressure of carbon dioxide, *TLC* total lung capacity, FEV_1 forced expiratory volume in 1 s, *FVC* forced vital capacity, *DLCO* carbon monoxide diffusing capacity

not detect any signs of reobstruction or TIPS dysfunction in any patient within 3 months after the procedure. Two (8.3%) patients, including one with moderate HPS and one without HPS, had mild hepatic encephalopathy 1 month after TIPS creation. These two patients were treated with lactulose monotherapy. No other procedure-related complications were observed within 3 months after the procedure. No patients received inhaled nitric oxide, methylene blue, antibiotics, tumor necrosis factor-α, vascular endothelial growth factor antagonists, endothelin-1, or interferon-γ within 3 months after the procedure.

Table 2 Procedure details

No./age (years)/sex	HPS	Site of obstruction	WHVP (mmHg)	IVCP (mmHg)	Type of procedure	Technical success	Pressure gradient (mmHg)	
							Preprocedure	Post-procedure
1/40/F	−	HV	24	4	HV balloon angioplasty	+	18*	5*
2/56/M	−	IVC	22	20	IVC balloon angioplasty	+	17†	3†
3/45/M	−	IVC	19	16	IVC balloon angioplasty	+	12†	0†
4/48/F	−	HV & IVC	20	17	IVC balloon angioplasty	+	10†	1†
5/38/F	−	HV & IVC	25	22	IVC balloon angioplasty	+	18†	3†
6/28/M	+	HV	27	3	HV balloon angioplasty	+	24*	4*
7/28/M	−	HV & IVC	−	−	TIPS creation	+	23‡	11‡
8/48/F	+	HV & IVC	−	−	TIPS creation	+	−	−
9/19/F	+	HV & IVC	−	−	TIPS creation	−	21‡	10‡
10/58/M	+	HV & IVC	−	−	TIPS creation	+	25‡	14‡
11/47/M	−	IVC	33	31	IVC balloon angioplasty	+	26†	4†
12/70/M	+	HV	21	5	HV balloon angioplasty	+	16*	3*
13/63/F	+	HV	−	−	TIPS creation	+	19‡	12‡
14/52/M	+	HV & IVC	−	−	TIPS creation	+	27‡	14‡
15/64/M	−	IVC	30	28	IVC balloon angioplasty	+	24†	0†
16/71/M	+	IVC	26	23	IVC balloon angioplasty	+	20†	1†
17/40/M	−	IVC	22	19	IVC balloon angioplasty	+	17†	0†
18/59/F	−	HV & IVC	−	−	TIPS creation	+	17‡	10‡
19/43/F	−	HV & IVC	18	17	IVC balloon angioplasty	+	12†	3†
20/48/F	+	HV & IVC	22	20	IVC balloon angioplasty	+	17†	0†
21/70/F	+	HV	−	−	TIPS creation	+	14‡	5‡
22/42/M	+	HV	−	−	TIPS creation	+	21‡	10‡
23/64/M	−	HV & IVC	−	−	TIPS creation	+	24‡	9‡
24/29/M	−	HV	30	5	HV balloon angioplasty	+	25*	9*
25/44/M	−	HV & IVC	−	−	TIPS creation	+	20‡	12‡

HPS hepatopulmonary syndrome, *HV* hepatic veins, *IVC* inferior vena cava, *TIPS* transjugular intrahepatic portosystemic shunt, *WHVP* wedged hepatic vein pressure, *IVCP* inferior vena cava pressure

*Pressure gradient across the obstructed HV

† Pressure gradient across the obstructed IVC

‡ Portosystemic pressure gradient

ABG analysis

The changes in the A-aO_2, PaO_2, and arterial partial pressure of carbon dioxide ($PaCO_2$) before and after the procedure are summarized in Table 3 and Fig. 2. For patients with HPS, the A-aO_2 remained comparable to baseline 2–3 days after the procedure (-3.2 ± 11.9 mmHg; $p = .412$), significantly improved 1 month after the procedure (-11.7 ± 6.4 mmHg; $p < .001$), and then returned to baseline 3 months after the procedure (-1.3 ± 12.5 mmHg; $p = .757$). Similarly, PaO_2 remained comparable to baseline 2–3 days after the procedure (+4.8 ± 11.6 mmHg; $p = .222$), significantly improved 1 month after the procedure (+14.2 ± 9.8 mmHg; $p = .001$), and then returned to baseline 3 months after the procedure (+3.2 ± 13.6 mmHg; $p = .479$). $PaCO_2$ remained comparable to baseline at all three time points after the procedure (-.8 ± 4.1 mmHg, -1.7 ± 5.8 mmHg, and -1.2 ± 3.9 mmHg; $p = .547$, $p = .390$, and $p = .372$). For patients without HPS, the A-aO_2 (+1.4 ± 8.3 mmHg, +3.5 ± 8.1 mmHg, and +1.3 ± 8.2 mmHg; $p = .543$, $p = .137$, and $p = .565$), PaO_2 (-1.4 ± 9.8 mmHg, -2.5 ± 9.2 mmHg, and +.1 ± 7.8 mmHg; $p = .603$, $p = .340$, and $p = .976$), and $PaCO_2$ (0 ± 4.7 mmHg, -1.3 ± 5.4 mmHg, -1.4 ± 5.0 mmHg; $p = .982$, $p = .387$, and $p = .317$) remained comparable to baseline at all three time points after the procedure.

Table 3 Changes in A-aO_2, PaO_2, and $PaCO_2$ before and after portal decompression

	HPS ($n = 10$)		Non-HPS ($n = 14$)		p value†
	mmHg	p value*	mmHg	p value*	
A-aO_2					
2–3 days	-3.2 ± 11.9	.412	+1.4 ± 8.3	.543	.273
1 month	-11.7 ± 6.4	< .001	+3.5 ± 8.1	.137	< .001‡
3 months	-1.3 ± 12.5	.757	+1.3 ± 8.2	.565	.551
PaO_2					
2–3 days	+4.8 ± 11.6	.222	-1.4 ± 9.8	.603	.170
1 month	+14.2 ± 9.8	.001	-2.5 ± 9.2	.340	< .001‡
3 months	+3.2 ± 13.6	.479	+.1 ± 7.8	.976	.485
$PaCO_2$					
2–3 days	-.8 ± 4.1	.547	0 ± 4.7	.982	.653
1 month	-1.7 ± 5.8	.390	-1.3 ± 5.4	.387	.875
3 months	-1.2 ± 3.9	.372	-1.4 ± 5.0	.317	.913

HPS hepatopulmonary syndrome, *A-aO_2* alveolar–arterial oxygen gradient, *PaO_2* arterial partial pressure of oxygen, *$PaCO_2$* arterial partial pressure of carbon dioxide

*p value based on one-sample Student's t test (test value, 0)

† p value based on independent samples Student's t test (HPS vs. non-HPS)

‡ p value statistically significant after Bonferroni correction ($p < .017$ (.05/3))

Discussion

The present study screened for IPVD and HPS in 25 patients with BCS and then evaluated the changes in arterial oxygenation after portal decompression in 11 BCS patients with HPS and found that (i) IPVD and HPS occurred in 56.0% and 44.0% of BCS patients, respectively and (ii) for BCS patients with HPS, the A-aO_2 and PaO_2 remained comparable to baseline 2–3 days after portal decompression (-3.2 ± 11.9 mmHg and +4.8 ± 11.6 mmHg; $p = .412$ and $p = .222$), significantly improved 1 month after portal decompression (-11.7 ± 6.4 mmHg and +14.2 ± 9.8 mmHg; $p < .001$ and $p = .001$), and then returned to baseline 3 months after portal decompression (-1.3 ± 12.5 mmHg and +3.2 ± 13.6 mmHg; $p = .757$ and $p = .479$).

IPVD is a common occurrence in patients with liver disease and/or portal hypertension [16]. Approximately 60% of cirrhotic patients have IPVD, and depending on the diagnostic criteria employed, more than 90% of them can develop HPS [6, 17]. De et al [11] reported an IPVD rate of 59% and an HPS rate of 28% in a prospective cohort of 29 patients with BCS. While the rate of IPVD in the present study (56.0%) was similar to that reported by De et al [11], the rate of HPS (44.0%) was higher. This is most likely due to differences in the patient populations but may also be attributed to differences in the diagnostic criteria used. De et al [11] employed stricter criteria to define arterial oxygenation defects (i.e., A-aO_2 > 20 mmHg or PaO_2 < 70 mmHg), which led to a lower HPS rate than in this study. In contrast, our current study employed more lenient criteria endorsed by clinical practice guidelines [4, 13, 14].

In the present study, patients with HPS had an improvement of A-aO_2 and PaO_2 1 month after portal decompression. De et al [11] reported an improvement in the A-aO_2 and PaO_2 also in 87% of patients with BCS and HPS 8 weeks after balloon angioplasty. The mechanism by which portal decompression improves arterial oxygenation is unknown. However, it is unlikely due to resolution of the other major complications of portal hypertension (e.g., ascites and variceal bleeding), because the A-aO_2 and PaO_2 remained comparable to baseline after portal decompression for patients without HPS. A possible explanation is that improved ventilation-perfusion matching induced by increased cardiac output after balloon angioplasty and TIPS creation, which may result in increased blood flow to the upper lungs where IPVD is usually less severe [18].

A few case reports have described individuals with recurrence of HPS after LT [19, 20]. In one report, the cause of recurrence was identified to be hepatic venous outflow obstruction, and reversal of HPS was observed after balloon angioplasty [20]. However, in the present

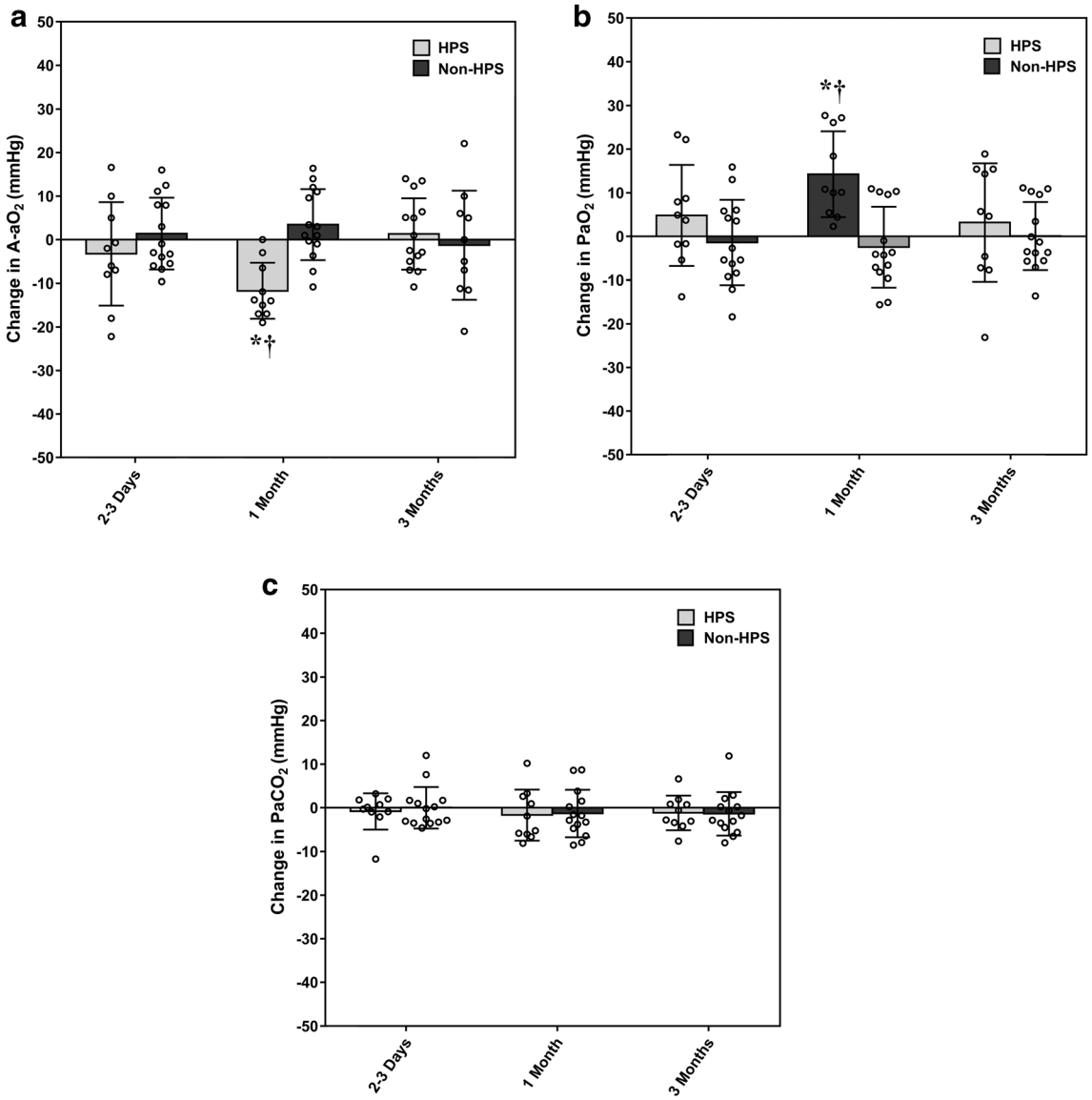

Fig. 2 Scatter dot plot of the changes in **a** A-aO_2, **b** PaO_2, and **c** $PaCO_2$ before and after portal decompression. Bar, mean; error bar, standard deviation.*$p < .05$, based on one-sample Student's t test (test value, 0). †$p < .05$ (Bonferroni correction, $p < .017$ (.05/3)), based on independent samples Student's t test (HPS vs. non-HPS)

study, the A-aO_2 and PaO_2 returned to baseline 3 months after portal decompression despite no reobstruction or TIPS dysfunction. A similar phenomenon has also been observed in patients with cirrhosis and idiopathic portal hypertension treated with TIPS creation for HPS [12]. The most plausible cause may be decreased cardiac output due to gradual acclimation to the increased blood flow to the heart [21].

The present study has several important limitations. First, the sample size was small due to the rarity of BCS. Second, a control group of patients with HPS who did not undergo portal decompression was not available for comparison, because it is unethical to withhold treatment to patients for 3 months. Third, the effects of balloon angioplasty and TIPS creation on arterial oxygenation were not compared in patients with HPS due to small sample size. Last, the degree of IPVD before and after the procedure was not evaluated due to a lack of patients with severe and very severe HPS.

In conclusion, arterial oxygenation transiently improves after portal decompression in BCS patients with HPS. Portal decompression cannot be recommended as a specific therapy to improve arterial oxygenation defect in BCS patients with HPS. However, it may have a role as a temporary bridge to LT in BCS patients with advanced HPS. Further studies are warranted to evaluate the impact of portal decompression on the morbidity and mortality of LT in BCS patients treated for HPS.

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Compliance with ethical standards

Guarantor The scientific guarantor of this publication is Xiao Li.

Conflict of interest The authors declare that they have no conflict of interest.

Statistics and biometry No complex statistical methods were necessary for this paper.

Informed consent Written informed consent was obtained from all subjects (patients) in this study.

Ethical approval Institutional Review Board approval was obtained.

Study subjects or cohorts overlap The subjects who underwent TIPS creation in this cohort have been included in another manuscript submitted elsewhere for publication.

Methodology
- Prospective
- Observational study
- Performed at one institution

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