Original Article

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Surgical treatment and prognosis of type II congenital extrahepatic portosystemic shunts: A single-center experience of 31 cases

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SUMMARY: Congenital extrahepatic portosystemic shunts (CEPS) are rare congenital vascular malformations characterized by an abnormal communication between the hepatic portal venous system and the systemic venous system. Type II CEPS preserves partial portal venous blood flow and can usually be treated with conventional surgery rather than solely relying on liver transplantation. To determine the optimal surgical methods and complication management strategies for type II CEPS patients, we retrospectively analyzed 31 predominantly adult patients with type II CEPS, documenting their surgical approaches and the occurrence of postoperative complications. Five surgical approaches were employed: 11 patients underwent shunt occlusion with 5 cases of complications; 5 patients underwent splenic vessels ligation with 2 cases of complications; 5 patients underwent shunt occlusion combined with splenic artery ligation with 4 cases of complications; 8 patients underwent shunt occlusion combined with distal splenorenal shunt with 3 cases of complications; and 2 patients with lower extremity edema underwent inferior vena cava shunt bypass surgery, with no significant complications observed. In conclusion, surgery centering on the shunt occlusion demonstrates promising therapeutic value and remains the mainstay in the treatment of type II CEPS. Meanwhile, postoperative complications remain a concern, necessitating long-term monitoring and management.

Keywords: congenital extrahepatic portosystemic shunts, portal vein, surgical treatment, prognosis, complication

1. Introduction

Congenital extrahepatic portosystemic shunts (CEPS), also known as Abernethy malformation, is a congenital vascular malformation characterized by an abnormal communication between the extrahepatic portal venous system and the systemic venous system. CEPS has a low global incidence, with fewer than 400 cases reported in the relevant literature to date. CEPS is classified into two types based on the presence or absence of intrahepatic portal venous blood flow (*1,2*). Type I CEPS is characterized by the complete absence of portal venous blood flow to the liver, while type II CEPS is characterized by partial preservation of portal venous blood flow to the liver (*3,4*). To confirm the classification of CEPS patients, balloon occlusion testing is required for those screened by routine imaging examinations to determine the presence of intrahepatic portal venous branches (*5*). For patients diagnosed with type II CEPS, due to the presence of intrahepatic portal venous branches, favorable therapeutic outcomes can usually be achieved through shunt ligation alone (*6*).

Previous findings have documented that some type II CEPS patients may present with severe complications, including hepatic encephalopathy (HE), gastrointestinal bleeding (GIB), pulmonary arterial hypertension (PaHT), and hepatopulmonary syndrome (*7-10*). Some patients with type II CEPS may develop hepatic nodules. Although most are benign, malignant transformation remains possible (*11*). For patients with such severe complications, surgical treatment can alleviate symptoms and slow disease progression to a certain extent. For asymptomatic patients, prophylactic surgical treatment can prevent such severe complications (*12*).

Currently, shunt occlusion remains the primary surgical approach for managing type II CEPS patients, including shunt ligation and endovascular embolization (*13*). Most previous studies have confirmed that shunt occlusion yields favorable recovery outcomes in pediatric-dominant patient populations, but there is a scarcity of literature documenting treatment approaches for adult-dominant populations (*8,10*). This study

aims to summarize our single-center experience with individualized surgical management, focusing on decision-making principles, perioperative outcomes, complication patterns, and long-term follow-up.

2. Patients and Methods

2.1. Study subjects

A total of 31 patients with type II CEPS who were hospitalized in the Department of Hepatobiliary and Pancreatic Surgery at the Chinese People's Liberation Army General Hospital from January 2011 to December 2024 were enrolled in this study. The inclusion criterion was that all patients were confirmed as type II CEPS by balloon occlusion testing, which demonstrated preserved intrahepatic portal venous flow. Patients with type I CEPS (absence of intrahepatic portal perfusion) were excluded.

2.2. Preoperative evaluation and diagnostic confirmation

All patients underwent comprehensive preoperative imaging assessments, including abdominal computed tomography (CT), magnetic resonance imaging (MRI), and/or ultrasonography (US). These examinations served to delineate shunt anatomy and classify shunts according to the Blanc system (*14*), which encompassed categories such as extrahepatic portosystemic shunt (EHPS), end to side like portocaval shunt (ESPC), side to side like portocaval shunt (SSPC), portohepatic shunt (PH), and persistent ductus venosus (PDV). Anatomical variants within this classification, including extrahepatic portorenal (EHPR), extrahepatic porto iliac/caval (EHPC), end to side like portorenal (ESPR), side to side like portorenal (SSPR), and side to side like portoatrial (SSPA), were also documented. In addition, imaging was used to evaluate severity of portal hypertension and to identify associated complications, such as hepatic nodules or gastroesophageal varices.

Laboratory investigations included measurements of liver function parameters (alanine aminotransferase (ALT), aspartate aminotransferase (AST), total bilirubin, albumin, prothrombin time (PT)), blood ammonia levels, and indocyanine green retention 15 min (ICG R15) to evaluate hepatic reserve and operative risk.

Definitive classification of CEPS was established *via* balloon occlusion angiography. Temporary occlusion of the shunt allowed assessment of intrahepatic portal vein patency and portal pressure dynamics, thereby distinguishing type II (partial portal flow preserved) from type I shunts.

2.3. Selection of surgical approaches

Surgical strategy was individualized according to preoperative shunt anatomy, portal hemodynamics, and major clinical manifestations. Preoperative imaging was used to evaluate shunt type, diameter, length, and anatomical relationship to the portal venous system, while intraoperative portal pressure measurement and ultrasonography were used to assess the hemodynamic tolerance of shunt occlusion and to guide final operative strategy. The main clinical considerations included portal hypertension, variceal bleeding risk, hyperammonemia, and lower-extremity symptoms related to abnormal venous drainage. Based on these factors, patients were assigned to one of five surgical approaches (Figure 1).

Shunt occlusion was selected for patients in whom direct interruption of the shunt was considered technically feasible and hemodynamically tolerable. Two options were used. Surgical ligation was preferred in patients with thick or short shunts, or in those requiring additional open surgical procedures. In these patients, absence of obvious congestion in the small intestine and colon after occlusion was used as an intraoperative indicator of acceptable portal venous outflow and tolerable portal pressure elevation. Endovascular embolization was considered in patients with relatively small shunts or in those who were unable to tolerate general anesthesia. After embolization, portal pressure was reassessed to ensure that it did not exceed 25 cmH_2O.

Splenic vessels ligation (ligation of splenic artery and vein) was used mainly in patients with splenic vein–left renal vein shunts accompanied by a splenic vein steal phenomenon identified by intraoperative ultrasonography.

Shunt occlusion combined with splenic artery ligation was considered in patients in whom direct shunt occlusion alone was judged likely to cause excessive postoperative portal hypertension. In these cases, splenic artery ligation was used to reduce portal inflow and improve hemodynamic tolerance after shunt restriction.

Shunt occlusion combined with distal splenorenal shunt (DSRS) or side-to-side anastomosis between the splenic vein and the inferior vena cava (IVC) was reserved for patients with severe portal hypertension suggested by clinical or imaging findings, or with marked varices along the lesser curvature of the stomach and a substantial risk of bleeding identified during intraoperative exploration. In patients with splenic vein shunts, splenic vessels ligation (*i.e.*, shunt occlusion) was performed first, followed by splenorenal shunting. In patients with other shunt types, such as portorenal or gastrorenal shunts, splenic vessels ligation and splenorenal shunting were performed first, followed by shunt occlusion.

IVC shunt bypass surgery was selected for patients with portoiliac shunts who presented with claudication and significant lower-extremity edema. In these patients, an artificial bypass from the shunt to the IVC was created to restore venous return and relieve venous congestion.

Intraoperative changes in portal pressure before and after temporary or definitive shunt occlusion

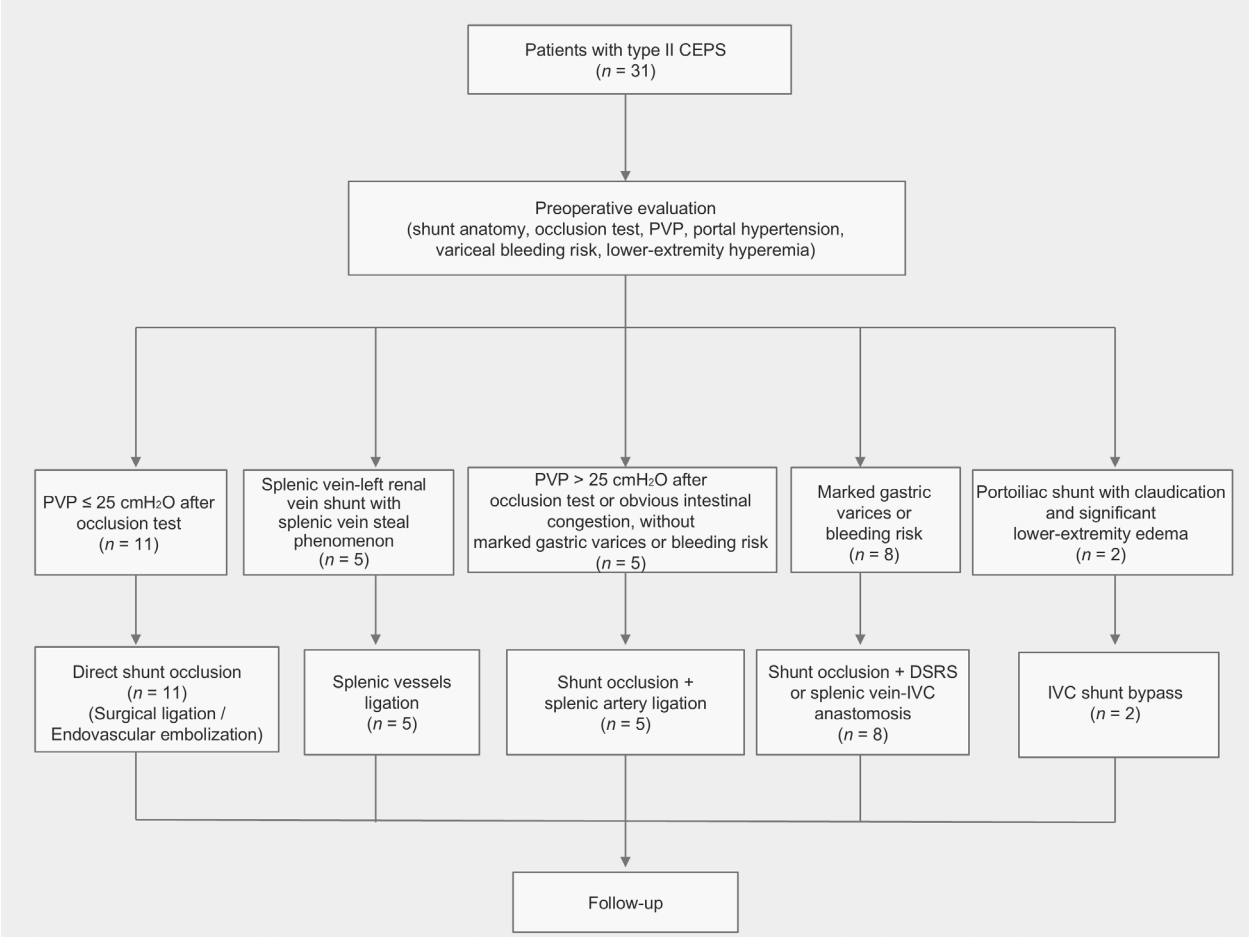

Figure 1. Surgical decision-making flowchart. *Abbreviations*: CEPS, congenital extrahepatic portosystemic shunt; PVP, portal venous pressure; DSRS, distal splenorenal shunt; IVC: inferior vena cava.

were recorded in all applicable patients. Blood flow direction and velocity were monitored by intraoperative ultrasonography to confirm the hemodynamic effect of the procedure. Postoperatively, imaging re-evaluation and laboratory testing were performed to assess recovery and detect complications.

2.4. Study follow-up

Postoperative evaluation included serial imaging (CT/US) and laboratory tests to assess restoration of hepatopetal portal flow, changes in liver volume, and the development of complications, such as thrombosis, recurrent shunts, or variceal progression. Symptom recurrence or new complications prompted further diagnostic work-up and tailored management. Follow-up data were collected until December 2024.

2.5. Statistical methods

SPSS 26.0 software was used to analyze data for each group. Measurement data conforming to normal distribution were expressed as mean ± standard deviation ($\bar{x} \pm s$) and compared using *t*-test; measurement data not conforming to normal distribution were expressed as median and interquartile range (IQR) *M_d* (P25, P75) and compared using the rank sum test. A p value < 0.05 was considered statistically significant. Given the small sample sizes in several subgroups, all statistical analyses were considered exploratory. No adjustment for multiple comparisons was performed because of the descriptive nature of the study.

2.6. Ethical approval

This study was approved by the Ethics Committee of Human Experimentation of the PLA General Hospital (No.S2018-013-01). All patients or their parents (for patients younger than 18 years) signed informed consent forms prior to inclusion in the study. This study was conducted in accordance with the Declaration of Helsinki.

3. Results

3.1. Baseline characteristics of patients

A total of 31 patients were enrolled in this study, including 13 males (42%). The median age at diagnosis was 39 years (range: 2–71 years). The follow-up cutoff

date was December 2024, and detailed baseline data are shown in Table 1.

Among the 31 enrolled patients, six were diagnosed incidentally during physical examination. The main symptoms at admission of the remaining patients were as follows: HE (n = 9, 29%), hyperammonemia (n = 20, 65%), PaHT (n = 4, 13%), hepatic nodules (n = 19, 61%), GIB (n = 7, 23%), abdominal pain (n = 4, 13%), dyspnea (n = 2, 6%), left lower extremity claudication (n = 2, 6%), and hemoptysis (n = 1, 3%). All patients were confirmed as type II CEPS by balloon dilation-occlusion testing. Anatomical types of shunts in all patients were classified according to the 4-category congenital portosystemic shunts (CPS) model proposed by Blanc, and information such as type II CEPS complications and specific surgical approaches is summarized in Table 2.

Among the 20 patients diagnosed with hyperammonemia, the mean blood ammonia level was 69.47 ± 21.60 μmol/L. Nine of these patients had HE before treatment, with a mean blood ammonia level of 81.92 ± 22.67 μmol/L. Lower extremity claudication was caused by a portoiliac shunt. Among the hepatic nodules, 7 were focal nodular hyperplasia (FNH), 8 were cirrhotic nodules (CN), and non-specific hepatic nodules (NSHN) were found in 8 patients on pathological examination.

Liver function test indicators of patients were as follows: the median aspartate aminotransferase (AST) was 24 U/L (IQR: 21.6–41.2), with abnormalities (≥ 40 U/L) in 9 patients. The median alanine aminotransferase (ALT) was 18.2 U/L (IQR: 14.1–26), with abnormalities (≥ 40 U/L) in 3 patients. The median total bilirubin was 21.9 μmol/L (IQR: 13.6–36.9), with abnormalities (> 21 umol/L) in 17 patients. The mean albumin level was 33 ± 5.63 g/L, with abnormalities (< 35 g/L) in 16 patients. Blood ammonia was measured in 30 patients, with a mean value of 54.33 ± 28.17 μmol/L (normal range: 0–32 μmol/L), and 20 patients had abnormalities. Preoperative ICG-R15 was measured in 21 patients, with a mean value of 33.60 ± 14.46%. PT was measured in all patients, with a mean value of 17.36 ± 2.44 s, and abnormalities (> 15 s) were observed in 28 patients

3.2. Treatment approaches and perioperative complication management

Thirty-one patients with type II CEPS underwent surgical intervention, including shunt occlusion (n = 11, 35.5%), splenic vessels ligation (n = 5, 16.1%), shunt occlusion combined with splenic artery ligation (n = 5, 16.1%), shunt occlusion combined with DSRS (n = 8, 25.8%), and IVC shunt bypass (n = 2, 6.4%). A descriptive summary of perioperative outcomes across the five surgical strategies is provided in Table 3. Overall, blood ammonia levels and ICG-R15 decreased significantly after surgery (p = 0.001 and p = 0.04), while portal pressure and liver volume increased significantly (p = 0.008 and p < 0.001, respectively). In contrast, no significant postoperative changes were observed in spleen volume (and p = 0.923). Detailed hemodynamic and volumetric data are presented in Table 4.

Table 1. Baseline Characteristics (n = 31)

Characteristics	n	%
Sex (Male)	13	42
Hepatic encephalopathy	9	29
Incidental finding on examination	6	19
Gastrointestinal bleeding	7	23
Abdominal pain	4	13
Dyspnea	2	6
Left lower limb claudication	2	6
Hemoptysis	1	3

A total of eleven patients received shunt occlusion, including six who underwent surgical ligation and five who underwent endovascular embolization. After treatment, blood ammonia levels decreased from 73.40 ± 22.72 μmol/L to 36.2 ± 20.67 μmol/L, and ICG-R15 decreased from 33.95 ± 19.68% to 21.16 ± 14.47%. Portal pressure increased from 11.45 ± 6.18 cmH_2O to 16.4 ± 4.39 cmH_2O. Liver volume increased from 734.61 ± 227.08 cm^3 to 864.22 ± 247.04 cm^3, whereas spleen volume showed only a limited change (333.475 ± 242.84 cm^3 *vs.* 363.89 ± 243.16 cm^3). When the surgical ligation and endovascular embolization subgroups were descriptively compared, changes in blood ammonia level, liver volume, and spleen volume appeared broadly comparable, although these subgroup observations should be interpreted with caution given the limited sample size.

Of the five patients who underwent splenic vessels ligation, four achieved significant symptom relief after surgery. Blood ammonia levels decreased from 67.58 μmol/L [IQR: 54.61–105.95] to 42.19 μmol/L [IQR: 22.26–100.16]. Portal pressure increased from 19.63 ± 6.9 cmH_2O to 22.12 ± 3.12 cmH_2O. ICG-R15 decreased from 37.13 ± 11.31% *vs.* 23.17 ± 19.17%. Liver volume increased from 673.6 ± 46.78 cm^3 to 768.07 ± 105.08 cm^3.

Among the five patients who underwent shunt occlusion combined with splenic artery ligation, all achieved significant relief of symptoms after surgery. Blood ammonia levels increased from 49.18 μmol/L [IQR: 22.73–51.00] to 56.34 μmol/L [IQR: 12.38-72.14]. Portal pressure increased from 14.1 ± 5.64 cmH_2O to 21.4 ± 1.52 cmH_2O. ICG-R15 increased from 28.9 ± 21.15% to 33.87 ± 24.58%. Liver volume increased from 1000.33 ± 692.06 cm^3 *vs.* 1156.25 ± 683.62 cm^3.

All eight patients who received shunt occlusion combined with DSRS achieved symptom relief. Among the five hyperammonemia patients, blood ammonia levels decreased from 69.31 ± 15.58 μmol/L to 33.72 ± 11.13 μmol/L. Portal pressure decreased from 24.0 cmH_2O [IQR: 15.75–28.0] to 22.0 cmH_2O [IQR: 17.75–31.0]. ICG-R15 decreased from 44.57 ± 8.31% to 25.83 ± 14.15%. Liver volume remained relatively stable from

Table 2. Detailed Information (n = 31)

Case No.	Sex	Age	Fistula Classification and Anatomy (Blanc's Classification)	CPS Pattern	Type II CEPS Complications	Management
1	F	2	Superior Mesenteric Vein - Inferior Mesenteric Vein - Left Iliac Vein	EHPS	Hematochezia, NSHN	Open surgical ligation of the anomalous shunt + Splenic artery ligation
2	F	3	Common Trunk of SMV & SV - Left Renal Vein, Portal Vein arising from the trunk	ESPR	FNH, RNH	Open surgical ligation of the anomalous shunt
3	M	10	Common Trunk of SMV & SV - IVC, Portal Vein arising from the trunk	SSPC	PaHT, NSHN, Accessory Spleen	Open surgical ligation of the anomalous shunt
4	M	15	Main Portal Vein - Right Atrium	SSPA	PaHT, FNH, Upper Abdominal Pain	Open surgical ligation of the anomalous shunt + Splenic artery ligation
5	M	15	Inferior Mesenteric Vein - Iliac segment of IVC	EHPC	lower extremity edema	Two-stage shunt occlusion, Subsequent IVC shunt bypass surgery
6	M	18	Portal Vein - Right Hepatic Vein shunt	PH	PaHT	DSA-guided embolization of the porto-hepatic shunt
7	M	19	Main Portal Vein - IVC	SSPC	FNH	Laparoscopic shunt flow restriction, Subsequent transhepatic arterial embolization
8	F	21	Main Portal Vein - IVC	ESPC	FNH, NSHN, Congenital Heart Disease	Open Portal vein ligation + Splenic artery ligation
9	F	27	Common Trunk of SMV & SV - Left Renal Vein, Main Portal Vein - Left Renal Vein	ESPR	Multiple FNH	Open surgical shunt flow restriction
10	M	31	Main Portal Vein - IVC	SSPC	Hematemesis due to Cirrhosis	Open shunt ligation + Splenic artery + Gastric coronary vein ligation, DSRS
11	F	32	Splenic Vein - Left Renal Vein	EHPR	Melena due to Cirrhosis, CN, Cavernous Transformation of Portal Vein, and Splenomegaly	Laparoscopic Splenic vessels ligation, Subsequent endovascular embolization
12	F	33	Splenic Vein - Left Renal Vein	EHPR	CTPV, Cirrhosis	Open splenic vessels ligation + Shunt vessel - Left Renal Vein shunt
13	M	35	Common Trunk of SMV & SV - Left Renal Vein	EHPR	Hepatic Adenoma, FNH, CN	Open surgical shunt narrowing + Liver Nodule Resection
14	M	35	Inferior Mesenteric Vein - Iliac Vein	EHPC	lower extremity edema	Open partial shunt occlusion, IVC shunt bypass surgery
15	F	37	Common Trunk of SV & SMV - Esophagogastric Variceal Shunt	EHPS	Fatigue, Autoimmune Cirrhosis, CN	Shunt vessel occlusion + Splenic artery ligation + DSRS

Abbreviations: CPS, congenital portosystemic shunts; SMV, superior mesenteric vein; SV, splenic vein; IMV, inferior mesenteric vein; FNH, focal nodular hyperplasia; CN, cirrhotic nodules; NSHN, non-specific hepatic nodules; RNH, regenerative nodular hyperplasia; PaHT, pulmonary arterial hypertension; HPS, hepatopulmonary syndrome; HE hepatic encephalopathy; CTPV, cavernous transformation of portal vein; DSRS, distal splenorenal shunt; DSA, digital subtraction angiography; IVC, inferior vena cava.

Table 2. Detailed Information (n = 31) (continued)

Case No.	Sex	Age	Fistula Classification and Anatomy (Blanc's Classification)	CPS Pattern	Type II CEPS Complications	Management
16	F	38	Superior Mesenteric Vein - IVC near Right Iliac Vein	EHPS	CTPV, NSHN, Hematemesis due to Cirrhosis, HPS	Open surgical ligation of the anomalous shunt
17	F	43	Splenic Vein - Left Renal Vein	EHPR	PaHT, Cirrhosis	Splenic vessels ligation
18	F	46	Main Portal Vein - Left Renal Vein	SSPR	Hematemesis due to Cirrhosis, NSHN, CN	Open portal vein ligation + Splenic artery ligation + DSRS
19	F	47	Gastric Coronary Vein - Left Renal Vein	EHPR	Melena due to Cirrhosis, NSHN	Open shunt ligation + Splenic artery ligation + DSRS, Subsequent DSA-guided splenic embolization
20	F	52	Superior Mesenteric Vein - IVC (near Right Iliac Vein)	EHPC	Hematochezia, CN	Open shunt ligation + Splenic artery ligation
21	F	54	Common Trunk of SV & SMV - Esophagogastric Variceal Shunt	EHPS	CTPV, FNH, Cirrhosis, Splenomegaly	Splenic vessels ligation + DSRS
22	F	55	Common Trunk of SV & SMV - Left Renal Vein	ESPR	HE	Balloon occlusion of splenorenal shunt
23	M	55	Splenic Vein - Left Renal Vein	EHPR	HE, Cirrhosis	Balloon occlusion of splenorenal shunt, Subsequent splenic vessels ligation
24	M	56	Splenic Vein - Left Renal Vein	EHPR	HE, Cirrhosis, NSHN	Splenic vessels ligation
25	F	56	Gastric Coronary Vein - Left Renal Vein	EHPR	Asymptomatic initially, jaundice developed one year later.	DSA-guided shunt occlusion
26	M	57	Splenic Vein - Left Renal Vein	EHPR	HE, Cirrhosis, CN	Splenic vessels ligation
27	F	61	Inferior Mesenteric Vein - IVC	EHPC	HE, CN	Laparoscopic occlusion of varices, DSRS
28	M	63	Portal Vein - IVC , Gastric Coronary Vein - Left Renal Vein	ESPC, EHPC	HE, NSHN	Open shunt ligation, Splenic artery ligation, Subsequent gastric coronary vein embolization & shunt occlusion
29	M	64	Splenic Vein - Left Renal Vein	EHPR	IVC Stenosis, HE, Cirrhosis, CN	DSA-guided porto-hepatic shunt embolization
30	F	65	Portal Vein - IVC	SSPC	Abdominal Pain, Cirrhosis	Open shunt ligation + Splenic artery ligation + DSRS
31	F	71	Portal Vein - Left Renal Vein	ESPR	HE	Laparoscopic coronary vein ligation + Splenic artery ligation

Abbreviations: CPS, congenital portosystemic shunts; SMV, superior mesenteric vein; SV, splenic vein; IMV, inferior mesenteric vein; FNH, focal nodular hyperplasia; CN, cirrhotic nodules; NSHN, non-specific hepatic nodules; RNH, regenerative nodular hyperplasia; PaHT, pulmonary arterial hypertension; HPS, hepatopulmonary syndrome; HE hepatic encephalopathy; CTPV, cavernous transformation of portal vein; DSRS, distal splenorenal shunt; DSA, digital subtraction angiography; IVC, inferior vena cava.

Table 3. Efficacy of five surgical procedure types

Characteristics	Shunt Occlusion (*n* = 11)	Splenic vessels ligation (*n* = 5)	Shunt Occlusion + Splenic Artery Ligation (*n* = 5)	Shunt Occlusion + DSRS (*n* = 8)	IVC Shunt Bypass surgery (*n* = 2)
Procedure Type					
Surgical	6	5	5	8	2
Endovascular	5	0	0	0	0
Clinical Indication & Outcome					
HE	3 (All improved)	3 (All improved)	1 (All improved)	1 (All improved)	-
PaHT	2 (All improved)	1 (1 Death)	1 (All improved)	-	-
GIB	1 (All improved)	1 (All improved)	2 (All improved)	3 (2 improved)	-
Abdominal Pain	2 (All improved)	-	1 (All improved)	1 (All improved)	-
Jaundice	1 (1 Death)	-	-	-	-
Claudication	-	-	-	-	2 (All improved)
Prophylactic Treatment	2	-	-	3	-
Complications	5 (1 Death)	2 (1 Death)	4	3	0

Abbreviations: HE, hepatic encephalopathy; PaHT, pulmonary arterial hypertension; GIB, gastrointestinal bleeding; DSRS, distal splenorenal shunt; IVC, inferior vena cava.

Table 4. Hemodynamic and volumetric changes before and after surgery

Procedure	Before Surgery	After Surgery	*p* value
Overall (*n* = 31, 100%)			
Blood ammonia (μmol/L) [hyperammonemic patients, *n* = 20]	68.38 [53.25–78.93]	32.87 [21.40–53.78]	0.001*
Portal pressure (cmH_2O) [*n* = 22]	15.75 [10.5–19.25]	21.00 [15.0–22.5]	0.008*
ICG-R15 (%)[*n* = 17]	35.73 ± 14.68	27.81 ± 18.38	0.04*
Liver volume (cm^3)[*n* = 25]	811.17 ± 315.94	915.07 ± 338.76	< 0.001*
Spleen volume (cm^3)[*n* = 13]	367.31 ± 264.14	369.65 ± 231.61	0.923
Shunt occlusion (*n* = 11, 35.5%)			
Blood ammonia (μmol/L) [hyperammonemic patients, *n* = 7]	73.40 ± 22.72	36.20 ± 20.67	< 0.001*
Portal pressure (cmH_2O) [*n* = 8]	11.45 ± 6.18	16.40 ± 4.39	0.015*
ICG-R15 (%)[*n* = 6]	33.67 ± 18.39	26.48 ± 20.9	0.207
Liver volume (cm^3) [*n* = 9]	734.61 ± 227.08	864.22 ± 247.04	0.024*
Spleen volume (cm^3) [*n* = 8]	333.48 ± 242.84	363.89 ± 243.16	0.19
Splenic vessels ligation (*n* = 5, 16.1%)			
Blood ammonia (μmol/L) [hyperammonemic patients, *n* = 4]	67.58 [54.61–105.95]	42.19 [22.26–100.16]	0.47
Portal pressure (cmH_2O) [*n* = 4]	19.63 ± 6.90	22.12 ± 3.12	0.47
ICG-R15 (%)[*n* = 3]	37.13 ± 11.31	23.17 ± 19.17	0.404
Liver volume (cm^3)[*n* = 3]	673.60 ± 46.78	768.07 ± 105.08	0.376
Shunt occlusion + splenic artery ligation (*n* = 5, 16.1%)			
Blood ammonia (μmol/L) [hyperammonemic patients, *n* = 2]	49.18 [22.73–51.00]	56.34 [12.38-72.14]	0.655
Portal pressure (cmH_2O) [*n* = 5]	14.10 ± 5.64	21.40 ± 1.52	0.044*
ICG-R15 (%)[*n* = 3]	28.90 ± 21.15	33.87 ± 24.58	0.18
Liver volume (cm^3)[*n* = 4]	1000.33 ± 692.06	1156.25 ± 683.62	0.005*
Shunt occlusion + DSRS (*n* = 8, 25.8%)			
Blood ammonia (μmol/L) [hyperammonemic patients, *n* = 5]	69.31 ± 15.58	33.72 ± 11.13	< 0.001*
Portal pressure (cmH_2O) [*n* = 5]	24.0 [15.75–28.0]	22.0 [17.75–31.0]	0.492
ICG-R15 (%)[*n* = 3]	44.57 ± 8.31	25.83 ± 14.15	0.11
Liver volume (cm^3)[*n* = 7]	806.09 ± 161.89	807.16 ± 129.53	0.89

Notes: Data are presented as mean ± SD or median [IQR]. *indicates $p < 0.05$. *Abbreviations*: DSRS, distal splenorenal shunt; ICG-R15, indocyanine green retention rate at 15 min; IQR, interquartile range; IVC, inferior vena cava; SD, standard deviation.

806.09 ± 161.89 cm^3 to 807.16 ± 129.53 cm^3.

Two patients underwent IVC shunt bypass surgery due to lower extremity edema, and their symptoms were effectively relieved after postoperative management and treatment strategy adjustment.

3.3. Perioperative complication management and outcomes

Among the 31 treated patients, 9 experienced postoperative complications of Clavien-Dindo grade III or higher; detailed postoperative grading is shown in Table 5.

Of the patients who underwent shunt occlusion, case 25 (jaundice) had been found to have a splenorenal shunt 1 year earlier and presented at admission with obvious decompensated cirrhosis. Interventional treatment was selected because it was considered less invasive. Although portal venous flow recovered after the procedure, the patient developed sudden cardiopulmonary

Table 5. Clavien-Dindo Complication Classification (*n* = 14)

Grade	Complication	*n*	Management
Grade II			
	Portal System Thrombosis	4	Anticoagulation
	Pulmonary Embolism	1	Anticoagulation
Grade III			
	Portal Hypertension	2	No effective intervention
	New Shunt Formation	4	Interventional occlusion
	Recurrent hepatic encephalopathy	1	Medical therapy
Grade IV			
	Multi-organ Failure	1	No effective intervention
Grade V			
	Acute Myocardial Infarction	1	No effective intervention

arrest on postoperative day 2, and embolic occlusion of a vital organ was considered a possible cause. Four other patients developed postoperative complications: 2 with portal hypertension, 1 with HE, and 1 with portal vein thrombosis. Case 22 (HE) had an elevated ICG-R15 value after interventional treatment and developed portal hypertension symptoms (manifested as ascites) 1 month postoperatively. Case 23 (HE) had a reduced liver volume postoperatively (1002.9→911.4 cm^3) and recurrent HE two months later. After splenic vessels ligation, the ICG-R15 value decreased to 40.1% (48%→40.1%), and the blood ammonia level dropped to 84.9 μmol/L (113.4→84.9 μmol/L). Case 7 (prophylactic treatment) developed portal hypertension-related HE six months postoperatively; after segmental hepatic artery embolization, the blood ammonia level decreased significantly (40.9→10.4 μmol/L) with favorable recovery. Case 13 (Liver Nodule) was found to have shunt thrombosis by postoperative ultrasound; and no significant changes were observed after three months of warfarin treatment.

Among the five patients who underwent splenic vessels ligation, Case 17 (PaHT) had severe pulmonary arterial hypertension (pulmonary artery pressure 122 mmHg) and right heart insufficiency for 3 years before surgery. Preoperative evaluation suggested insufficient operative reserve. Because marked thrombocytopenia secondary to hypersplenism contraindicated interventional treatment and no other alternative was available, the patient underwent surgery and subsequently developed postoperative multi-organ failure. Case 11 (melena) had an elevated ICG-R15 value (5→34%) 2 months postoperatively. Interventional examination revealed a new shunt (splenic vein-left ovarian vein shunt), which was occluded interventially; and the patient recovered well with a decreased ICG-R15 value (34→25.9%).

Among the five patients who underwent shunt occlusion combined with splenic artery ligation, 4 developed postoperative complications: 3 had elevated ICG-R15 values. Case 4 (PaHT) had an increased ICG-R15 value (17.6→23.7%), developed pulmonary embolism at 1-year follow-up, and multiple abdominal collateral circulations at 5-year follow-up. Case 8 (Congenital Heart Disease) developed superior mesenteric vein and main portal vein thrombosis 10 days postoperatively with an elevated ICG-R15 value (15.8→16.0%). After thrombolytic therapy and regular warfarin administration after discharge, the thrombosis disappeared at 1-month follow-up. Case 28 (HE) had an elevated ICG-R15 value (53.3→61.9%); and a new shunt (gastroepiploic vein- IVC shunt) was detected 3 months postoperatively. After interventional occlusion, the portal pressure increased (12.5→20 cmH_2O), and the blood ammonia level decreased (62.13→28.95 μmol/L). Case 1 (a child with a portoiliac shunt and hematochezia) had symptom recurrence 2 years later due to a new abnormally dilated drainage tract. Interventional examination revealed that the previously ligated shunt vessel (inferior mesenteric vein) drained into abnormally dilated pelvic veins.

Among the eight patients who underwent shunt occlusion combined with DSRS, three developed complications: Case 18 (hematochezia) had elevated portal pressure (24→30 cmH_2O) postoperatively, recurrent hematochezia due to gastrorenal shunt 2 years later, and multiple hepatic nodules 4 years later. Case 10 (GIB) had decreased portal pressure (28→14 cmH_2O) postoperatively but developed extensive portal vein and splenic vein thrombosis 10 days later. Anticoagulant therapy was ineffective, and hematemesis, melena, and portal hypertensive gastropathy occurred within 1 year. Case 19 (hematochezia) had decreased portal pressure (21.5→18.5 cmH_2O) postoperatively, developed portal vein thrombosis complicated by hilar and gastric fundus varices, and splenic artery thrombosis within 10 days. After heparin anticoagulation, the thrombosis resolved, and the extent of splenic infarction decreased.

Two patients underwent IVC shunt bypass surgery due to claudication. Case 5 (lower extremity edema) initially underwent a two-stage shunt occlusion. Postoperative indicators improved (decreased blood ammonia, elevated portal pressure), but symptoms did not. Long-term follow-up showed lower extremity flushing, thrombosis, and portal hypertension complications due to the formation of superior mesenteric vein-iliac vein collateral circulation. After

careful consideration, IVC shunt bypass surgery was performed, and symptoms were relieved. Case 14 (lower extremity edema) showed no symptom relief after splenic artery occlusion in the interventional department. Thus, restrictive portacaval shunt combined with IVC shunt bypass surgery was performed, and symptoms were relieved postoperatively. However, the patient had persistently elevated blood ammonia during long-term follow-up.

4. Discussion

In this study, drawing on a relatively large cohort for this rare condition, we described five surgical strategies tailored to different shunt anatomies and clinical scenarios, and evaluated their perioperative and long-term outcomes. These findings may provide practical guidance for the future management of CEPS.

Bernard summarized occlusion portal pressures in 59 children with congenital portosystemic shunts and showed that pressures varied widely after shunt occlusion. They also emphasized that no precise cut-off value could be defined for deciding between one-stage and two-stage closure, and that small bowel tolerance during occlusion was a key intraoperative consideration (*13*). Compared with this approach, in our center we used 25 cmH_2O as a practical reference threshold for procedure selection. In our cohort, shunt occlusion significantly increased portal pressure and liver volume, suggesting improved hepatic inflow. Additionally, no significant differences were found in the changes of blood ammonia level, ICG-R15, liver volume, or spleen volume between surgical ligation and interventional occlusion for shunt occlusion. These results suggest that interventional treatment, a less invasive approach, can be prioritized for type II CEPS patients undergoing shunt occlusion who meet the criteria above.

Relevant literature has indicated that Abernethy malformation is associated with portal hypertension-related complications, with gastroesophageal varices as the main manifestation in reported cases (*15*). According to related studies, treatment of type II CEPS patients requires a balance between "restoring portal perfusion, avoiding excessive postoperative portal hypertension","controlling complications", and individualized combined surgical or interventional decompression regimens can be adopted (*16*). Therefore, we explored surgical approaches for this patient group.

This study identified that some patients with splenorenal shunts exhibited retrograde portal venous flow into the splenic vein. In such patients, the abnormal shunt diverts blood from both the portal venous system and the splenic arterial system. Splenic vessels ligation was therefore performed to interrupt this abnormal flow. Among the patients in this study, five underwent splenic vessels ligation. Postoperatively, symptoms were significantly alleviated in four of these patients. Although no statistically significant differences were observed in blood ammonia levels, portal pressure, ICG-R15, or changes in liver volume, a trend toward improvement was noted.

For type II CEPS patients with high portal pressure, splenic artery ligation may be considered. Its main purpose is to reduce abnormal splenic inflow, thereby alleviating portal hypertension and hypersplenism while preserving as much splenic immune function as possible. Splenic vessels ligation can directly reduce blood flow from the spleen to the portal venous system, thereby helping to lower abnormally elevated portal pressure and relieve portal hypertension (*13*). Among patients undergoing shunt occlusion, some still have high intraoperative portal pressure after shunt ligation. Combining splenic artery ligation with shunt occlusion can stabilize portal pressure and avoid the risk of secondary surgery. In our cohort, postoperative portal pressure and liver volume increased in this subgroup, consistent with improved portal perfusion. This suggests that shunt occlusion combined with splenic artery ligation can effectively regulate portal pressure and hepatic blood perfusion in patients.

DSRS is widely used for surgical decompression of esophagogastric variceal bleeding in cirrhotic patients and pediatric patients with portal hypertension. It maintains significant hepatopetal perfusion of the main portal vein and mesenteric vein after surgery, while effectively shunting splenic blood flow, thereby reducing portal venous system pressure and the risk of variceal bleeding (*17*). In our study, shunt occlusion combined with DSRS or side-to-side splenic vein-IVC anastomosis was used in patients with severe portal hypertension or high-risk gastric varices. This approach appeared more suitable than shunt occlusion with splenic artery ligation alone in patients with established portal hypertension and marked varices.

All eight patients who underwent shunt occlusion combined with DSRS experienced symptom relief, and blood ammonia levels decreased in patients with hyperammonemia, suggesting improved metabolic status after surgery.

In portoiliac variants of type II CEPS, high-flow drainage into the iliac venous system may generate pelvic and lower-limb venous hypertension, leading to refractory edema, venous claudication, flushing, and thrombotic events. In our cohort, two patients underwent IVC shunt bypass surgery because of disabling claudication and persistent lower-extremity symptoms despite prior interventions. Postoperative symptom relief was achieved, although persistent hyperammonemia was observed in long-term follow-up in one patient.

However, postoperative complications and mortality should be interpreted in the context of heterogeneous baseline risk. In our cohort, the most serious adverse outcomes appeared more likely to occur in patients with advanced disease, major comorbidities, and limited

physiologic reserve, as illustrated by Case 17 with severe pulmonary arterial hypertension and longstanding right heart insufficiency, and Case 25 with decompensated cirrhosis at admission. These outcomes should therefore not be attributed to procedural failure alone. This point is important when comparing treatment strategies, because the five procedures were applied to patients with substantially different anatomy, hemodynamics, symptoms, and operative tolerance. Direct comparison of postoperative safety across procedures is therefore not appropriate in this cohort. In addition, grade III complications, including portal hypertension and new shunt formation, could not be clearly linked to specific baseline characteristics or to a particular surgical strategy. These associations remain speculative and require further study.

Previously, surgical treatment for type II CEPS patients mostly involved one-stage/multi-stage shunt ligation (*3,18,19*). In this study, for some patients with excessively elevated portal pressure after shunt ligation, splenic vessels ligation or DSRS was performed (*20,21*). On the basis of successful restoration of portal pressure and hepatic blood flow, the incidence of surgery-related complications was not significantly increased. Nevertheless, postoperative complications were observed across subgroups, including portal hypertension, hepatic encephalopathy, portal vein thrombosis, and new collateral shunt formation. Most complications improved after anticoagulation, re-intervention, or other active management.

This study also has limitations. It is a retrospective study, with follow-up bias in some patients. Additionally, the study mainly focuses on adult and elderly type II CEPS patients, with a small sample size of infants and children. Furthermore, the sample size of several subgroups was small, and the heterogeneity of shunt anatomy, clinical presentation, and surgical strategies limited direct inter-group comparison. Therefore, the findings should be interpreted with caution and regarded as descriptive rather than confirmatory. Despite the small sample size, it is a large-sample study for such a rare disease as type II CEPS. We believe that the new combination of surgical treatment approaches is beneficial for improving patient outcomes, achieving more thorough symptom relief, and reducing recurrence. However, it still needs further research and clinical practice to be improved. In conclusion, surgery centering on shunt occlusion demonstrates promising therapeutic value and remains the mainstay in treatment of type II CEPS.

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