



Case report

Surgical shunt ligation for a congenital extrahepatic portosystemic shunt with pulmonary hypertension: A case report

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ABSTRACT

Introduction and importance: Congenital extrahepatic portosystemic shunt (CEPS) presents with various symptoms due to abnormal communication between the portal venous system and inferior vena cava. And Klippel-Trenaunay-Weber syndrome is another rare congenital disorder characterized by vascular malformations.

Case presentation: A 16-year-old male with Klippel-Trenaunay-Weber syndrome was referred to our hospital for surgical treatment of pulmonary hypertension due to CEPS since childhood. Dyspnea had developed about two years before presentation at our hospital and gradually worsened. Right heart catheterization had revealed pulmonary hypertension and a high cardiac output state associated with a portosystemic shunt. Although pulmonary vasodilators improved the dyspnea, pulmonary hypertension remained. The patient was then referred to our hospital for surgical shunt occlusion. The results of the preoperative and intraoperative shunt occlusion tests were within acceptable limits. Therefore, primary shunt ligation was performed. There were no postoperative complications. Continuous intravenous vasodilator was tapered off four months after discharge. No additional or increased doses of medications were required for four years after surgery.

Clinical discussion: Surgical shunt ligation for CEPS is effective but can cause acute portal hypertension. Primary shunt ligation could be performed without complications according to the criteria for safe shunt occlusion.

Conclusion: Surgical portosystemic shunt ligation could be performed safely and ameliorated pulmonary hypertension. Patients with CEPS, especially young patients, should be referred to a tertiary center while asymptomatic, and early therapeutic intervention is necessary.

1. Introduction

Congenital extrahepatic portosystemic shunt (CEPS) is a rare vascular malformation with communication between the portal venous system and inferior vena cava, also known as an Abernethy malformation [1]. CEPS could cause hepatic encephalopathy, hepatopulmonary syndrome, pulmonary hypertension (PH), and liver lesions [2–4]. Despite the accumulation of cases, the indications and timing of treatment for CEPS remain unclear [3,4]. Klippel-Trenaunay-Weber syndrome (KTWS) is also an uncommon congenital disorder characterized by vascular malformations, varicose veins, and hypertrophy of bone or soft tissues [5,6]. Here, we report the case of a young male with PH due to CEPS and KTWS for whom pharmacotherapy alone was inadequate

and who benefited from surgical shunt ligation. This case report has been reported in line with the SCARE Criteria [7].

2. Presentation of case

A 16-year-old male was referred to our hospital regarding the indication for surgical intervention for PH caused by CEPS.

He had developed hyperammonemia in childhood and was subsequently diagnosed with KTWS and CEPS. Oral administration of lactulose was initiated to prevent hyperammonemia. He then developed without any problems and could normally exercise.

About two years before this presentation, he had felt shortness of breath on exertion that gradually worsened. Five months before this

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presentation, transthoracic echocardiography at another hospital revealed PH. Right heart catheterization revealed a mean pulmonary artery pressure of 46 mmHg and pulmonary artery wedge pressure of 16 mmHg. A high cardiac output state due to a portosystemic shunt met the criteria for portopulmonary hypertension. Although pulmonary vasodilators, including oral macitentan and continuous intravenous epoprostenol, improved shortness of breath on exertion, follow-up right heart catheterization four months later revealed the remaining PH. His disease was not expected to improve with pulmonary vasodilators. Closure of the shunt vessel by catheter intervention was considered; however, the diameter (30 mm) of the vessel was too large to be closed with the devices available in Japan. The patient was referred to our hospital for surgical intervention.

His medications included lactulose, tolvaptan, furosemide, macitentan, and intravenous epoprostenol. His family history was unremarkable. Physical examination revealed hemangiomas in the oral cavity and edematous changes and hemangiomas in the left leg (Fig. 1). The complete blood count and biochemistry tests were within the normal range, except for blood ammonia, 66 $\mu\text{mol/L}$ (normal range, 12 to 39), and total bile acids, 67.5 $\mu\text{mol/L}$ (normal range, 0 to 14.4). Transthoracic echocardiography showed no congenital heart or valvular diseases. Contrast-enhanced computed tomography showed anastomosis of the inferior mesenteric vein (IMV) with the right internal iliac vein and hypoplasia of the intrahepatic and main trunk of the portal vein (Fig. 2A–C). Computed tomography and magnetic resonance imaging with gadolinium-ethoxybenzyl-diethylenetriamine pentaacetic acid (Gd-EOB-DTPA) showed multiple liver nodules, suggesting focal nodular hyperplasia (Fig. 2D, E). An endovascular shunt occlusion test

was performed to determine the surgical indication. The portogram revealed a thin main trunk of the portal vein (Fig. 2F). The direct portal venous pressure was 6 mmHg, which increased to 13 mmHg after balloon occlusion in the IMV.

Based on these findings, a diagnosis of type II CEPS was made. Primary shunt occlusion was determined to be feasible based on previous studies [8,9]. Before the operation, intravenous epoprostenol, which has a strong antiplatelet effect, was replaced with intravenous treprostinil. The operation was performed under general anesthesia. The direct portal venous pressure was 9 mmHg before shunt occlusion. The IMV was exposed and taped at the inflow to the splenic vein (Fig. 3A). When the IMV, the shunt vessel, was clamped, the portal venous pressure increased to 16 mmHg. There was no evidence of intestinal congestion. The IMV was ligated and cut at the inflow into the splenic vein (Fig. 3B). The liver nodule was biopsied and showed no malignant pathological findings.

There were no postoperative complications, including liver failure, thrombosis, or opening of a new portosystemic shunt. Blood ammonia and total bile acid levels were within the normal range. The patient was discharged on postoperative day 15. Four months after surgery, continuous intravenous treprostinil was discontinued. On right heart catheterization three months and one year after surgery, the mean pulmonary artery pressures slightly decreased (Table 1). One year after surgery, contrast-enhanced computed tomography and portogram via the superior mesenteric artery showed that the main trunk of the portal vein was slightly thickened (Fig. 4A, B, E). Some of the liver nodules regressed (Fig. 4C, D). Four years after the surgery, the patient was doing well without requiring any additional or dose increases of oral and

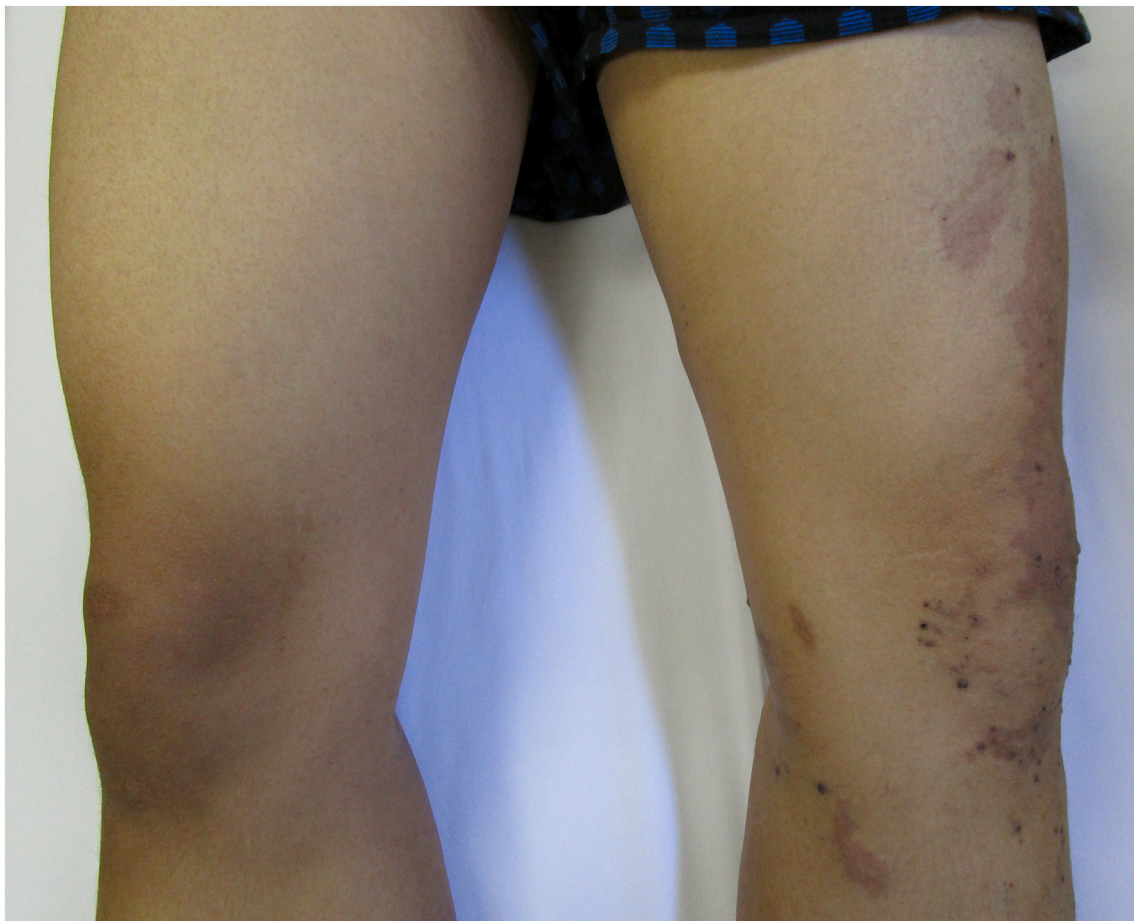


Fig. 1. Clinical photograph of the legs. Edematous changes and hemangiomas are shown in the patient's left leg.

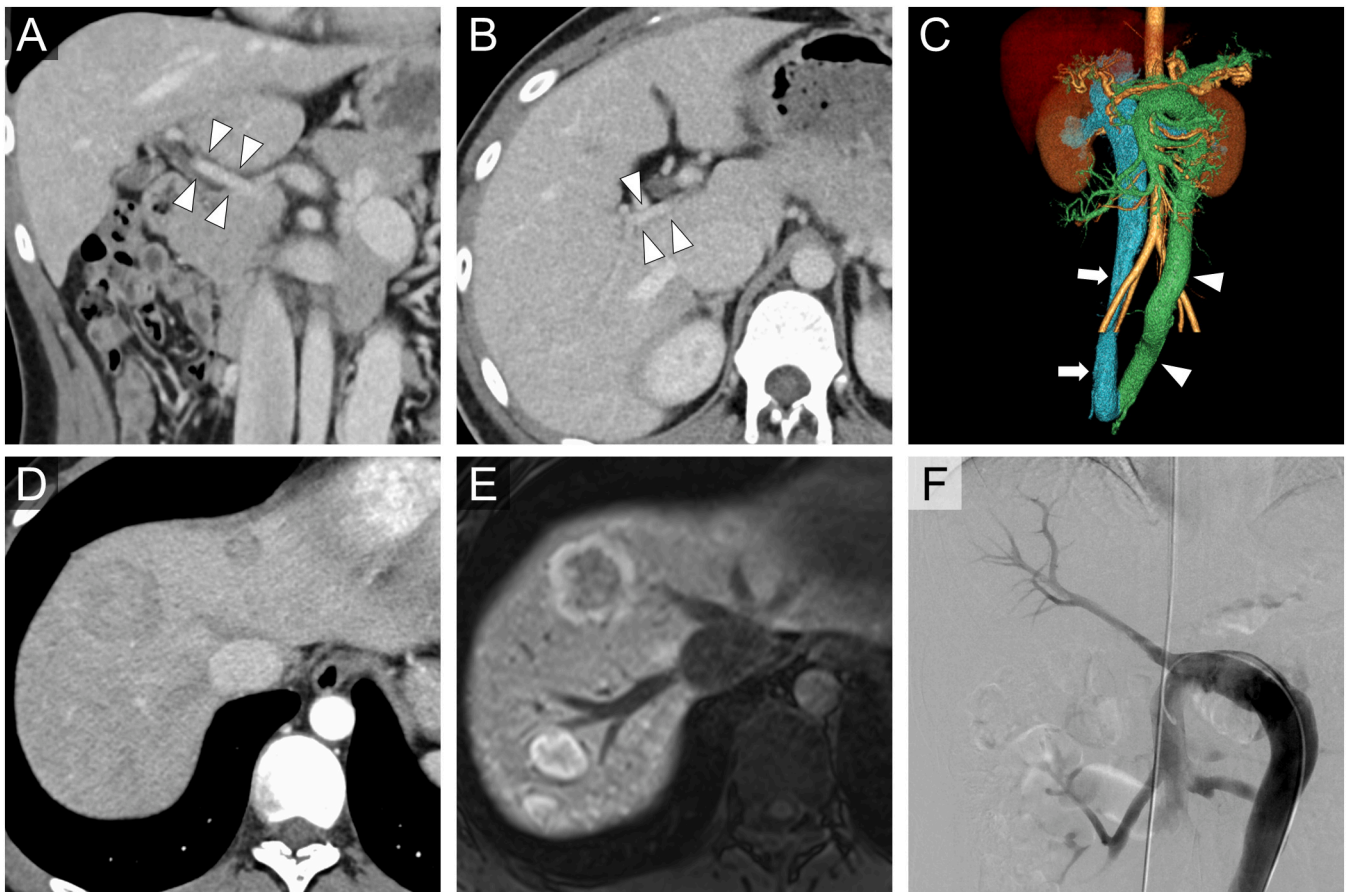


Fig. 2. Preoperative transthoracic echocardiography, abdominal CT, angiography, and MRI.

- A. Coronal CT imaging shows a thin main trunk of the portal vein (arrowhead).
- B. An abdominal CT shows hypoplasia of the right portal vein (arrowhead). The left branch of portal vein is not identified.
- C. A three-dimensional CT shows an abnormal communication between the inferior mesenteric vein (arrowhead) and the right internal iliac vein (arrow).
- D. An abdominal CT shows multiple low-density liver nodules.
- E. A Gd-EOB-DTPA enhanced MRI shows enhancement of multiple liver nodules in the hepatobiliary phase.
- F. A portogram via the portosystemic shunt shows a narrowed portal venous system.

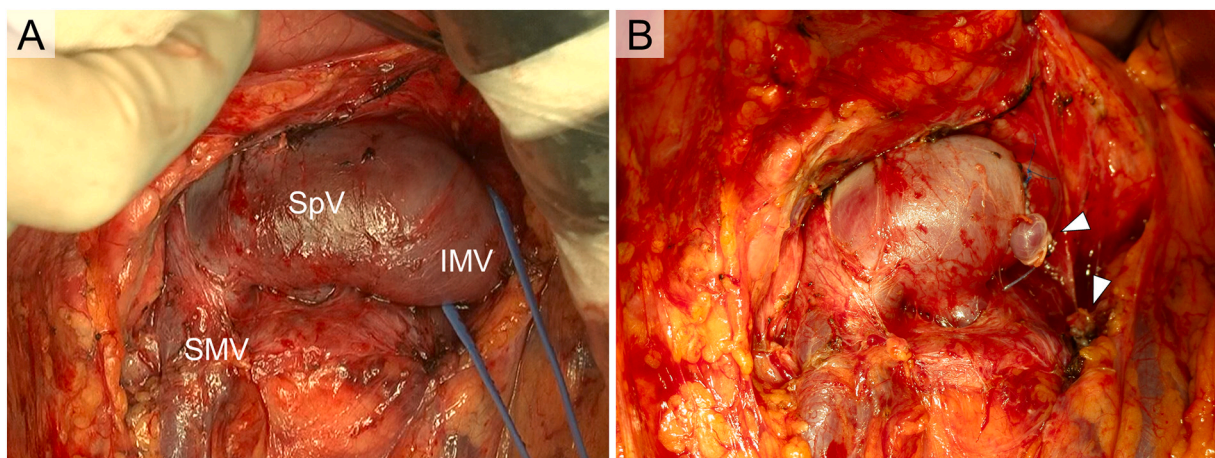


Fig. 3. Intraoperative findings.

- A. The inferior mesenteric vein, the shunt vessel, is taped at its inflow to the splenic vein.
 - B. The inferior mesenteric vein is ligated and cut. Arrowheads indicate the dissected ends of the vein.
- IMV: inferior mesenteric vein; SpV, splenic vein; SMV, superior mesenteric vein.

Table 1
Pre- and postoperative parameters associated with pulmonary hypertension.

	Before surgery	3 months after surgery	1 year after surgery
Systolic/diastolic pulmonary artery pressure (mmHg)	61/25	54/21	52/23
Mean pulmonary artery pressure (mmHg)	43	37	38
Pulmonary capillary wedge pressure (mmHg)	19	15	15
Cardiac output (L/min)	11.5	8.7	8.6
Right atrial pressure (mmHg)	9	7	8
Pulmonary vascular resistance (dyn·s/cm ⁵)	160	202	213
Pulmonary arterial oxygen saturation (%)	83.6	80.2	85.5
Brain natriuretic peptide (pg/mL)	<5.8	8.3	8.3
Blood ammonia (μmol/L)	66	14	10

intravenous medications.

3. Discussion

Surgical shunt ligation was safely performed for CEPS, which is a rare disease with no established standard treatment. This case is also one of the few reports of CEPS associated with KTWS.

CEPS can be divided into two types: type I, in which the intrahepatic portal veins are absent, and type II, in which intrahepatic portal venous flow exists [10]. In type II CEPS, shunt occlusion has been reported to be

effective, but the method has not been thoroughly investigated. Acute portal hypertension is an important complication of shunt occlusion. This is because the intrahepatic portal vein is diminutive, and the portal venous system may not adequately accommodate the increased blood flow. The criteria for safe shunt occlusion to avoid acute portal hypertension have been reported [8,9]. Primary shunt ligation is indicated if the change in portal venous pressure before and after shunt occlusion is less than 10 mmHg and the portal venous pressure after occlusion is less than 25 mmHg. However, if this criterion is not met, staged shunt occlusion is indicated to allow gradual intrahepatic portal system dilation. In this case, preoperative and intraoperative shunt occlusion tests were within acceptable limits, and we determined primary shunt ligation. Intraoperative findings of the intestinal and hepatic conditions were normal, and the postoperative course was good, without complications such as portal hypertension or liver failure.

Another issue is the indication and timing of liver transplantation (LT) for PH caused by CEPS. A Japanese nationwide survey showed that PH improved after LT in all patients [2]. In addition, once PH becomes severe, it is difficult to improve with LT, and high mortality after LT has been reported [12]. However, considering its highly invasive nature, the difficulty in securing donors in Japan, and the adverse effect of lifelong immunosuppressive medication, it is difficult to conclude whether LT should be prioritized over shunt occlusion. In this case, the effect of shunt ligation was not sufficient to normalize PH. Shunt ligation could improve volumetric overload from portosystemic shunts; however, vascular remodeling in the pulmonary arterioles due to prolonged PH is irreversible. Both shunt ligation and LT should be performed before the progression of PH. Patients with a portosystemic shunt should be



Fig. 4. Postoperative transthoracic echocardiography, abdominal CT, angiography, and MRI.
A. Coronal CT imaging shows a thicker main trunk of the portal vein than before surgery (arrowhead).
B. An abdominal CT shows a thicker right portal vein (arrowhead).
C, D. An abdominal CT and a Gd-EOB-DTPA enhanced MR show regression of some of the liver nodules in the hepatobiliary phase.
E. A portogram via the superior mesenteric artery shows a thicker portal venous system.

referred to the tertiary care center as soon as possible, even if asymptomatic.

Many cases of CEPS are associated with one or more congenital malformations, most commonly circulatory abnormalities followed by musculoskeletal abnormalities [3,13]. On the other hand, KTWS is characterized by the coexistence of various vascular malformations, but there have been few reports of CEPS with KTWS [14–16]. In this case, CEPS associated with KTWS caused the PH. CEPS may be one of the vascular malformations associated with KTWS, and physicians should pay attention to this disease.

4. Conclusion

Surgical shunt ligation for CEPS with PH can be safely performed without complications. Patients with CEPS, especially young patients, should be referred to a tertiary center while asymptomatic, and early therapeutic intervention is necessary.

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Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

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CRediT authorship contribution statement

KY and SM wrote the initial draft of the manuscript. YT and YS

contributed to the critical revision of the manuscript. YI supervised the manuscript. HN gave final approval of the manuscript. All authors have read and approved the final manuscript.

Declaration of competing interest

The authors declare that they have no competing interests.

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