

## CASE REPORT

# Role of interventional radiology in the diagnosis and management of congenital extrahepatic portosystemic shunts: Two case reports

Sheetal V Mathai, Victor Kondray<sup>1</sup>, Elias Salloum<sup>2</sup>, Kamlesh Kukreja<sup>3</sup>, Sidhartha Tavri<sup>1</sup>

Department of Radiology, Kasturba Medical College, Mangalore, Manipal Academy of Higher Education, Karnataka, India, <sup>1</sup>Department of Radiology, University Hospitals, Case Western Reserve University School of Medicine, 11100 Euclid Ave, Cleveland, OH 44106, <sup>2</sup>Department of Radiology, Massachusetts General Hospital, 55 Fruit St, Boston, MA 02114, <sup>3</sup>Department of Radiology, Texas Children's Hospital, 6701 Fannin Street, Suite 470, Houston, TX 77030, USA

**Correspondence:** Dr. Sidhartha Tavri, Department of Radiology, University Hospitals, Case Western Reserve University School of Medicine, 11100 Euclid Avenue, BSH 5056, Cleveland, OH 44106, USA. E-mail: Sidhartha.Tavri@UHHospitals.org

## Abstract

Congenital extrahepatic portosystemic shunt (CEPS) is a rare splanchnic venous malformation, wherein the portal venous outflow drains into the systemic venous circulation via a pathologic shunt. CEPS exhibits heterogeneous clinical behavior and angiography is the gold standard for evaluation of the portomesenteric communication to systemic vasculature. The potential severity of complications necessitates shunt closure. Here, we present two cases of CEPS. The first patient presented with an asymptomatic hyperammonemia and was found to have a Type 1 CEPS with absence of intrahepatic portal system. The second patient was asymptomatic and was incidentally found to have a Type 2 CEPS on imaging with normal intrahepatic portal system. Both patients were successfully treated with endovascular occlusion of the CEPS.

**Key words:** Congenital extrahepatic portosystemic shunt; endovascular closure; focal nodular hyperplasia; hyperammonemia; portal hypertension

## Introduction

Congenital extrahepatic portosystemic shunt (CEPS) is a rare splanchnic venous abnormality, wherein the portomesenteric blood drains into the systemic circulation through a shunt, consequently bypassing the hepatic circulation. The severity of complications of CEPS warrants shunt closure. Complications are due to loss of hepatic filtration of toxic metabolites and altered hemodynamics causing high-flow systemic venous circulation.<sup>[1]</sup> Here

we present two cases of CEPS with different clinical presentations and shunt types. Both cases were successfully treated by endovascular techniques.

## Case History

### Case 1

During a routine laboratory workup of a 5-year-old girl status post repair of a tracheoesophageal fistula, the serum

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

**For reprints contact:** reprints@medknow.com

**Cite this article as:** Mathai SV, Kondray V, Salloum E, Kukreja K, Tavri S. Role of interventional radiology in the diagnosis and management of congenital extrahepatic portosystemic shunts: Two case reports. Indian J Radiol Imaging 2019;29:219-22.

### Access this article online

#### Quick Response Code:



**Website:**  
www.ijri.org

**DOI:**  
10.4103/ijri.IJRI\_461\_18

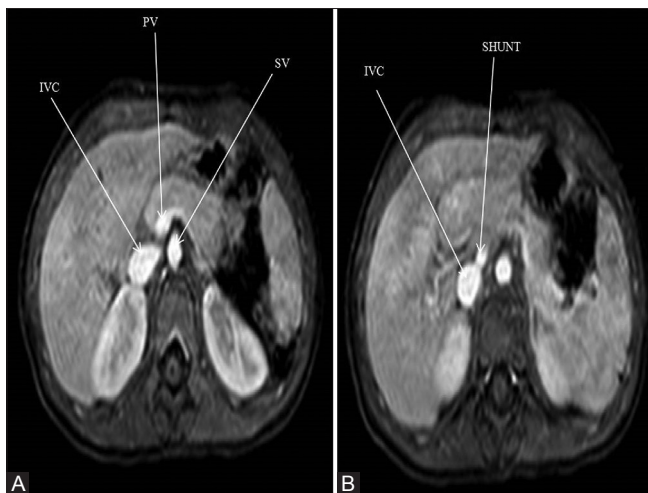
ammonia was found to be elevated to 71 mmol/L. Computed tomography (CT) scan revealed a focal nodular liver lesion and a portosystemic shunt from the portal vein to the inferior vena cava (IVC). Subsequent biopsy confirmed the lesion to be focal nodular hyperplasia. Magnetic resonance imaging/magnetic resonance angiography (MRA) confirmed the shunt [Figure 1A, B] and no significant intrahepatic portal veins. Two weeks later she underwent shunt closure of diagnosed Abernethy Type 1 malformation. An ultrasound and fluoroscopy-guided right transjugular vein approach was utilized. 8 Fr sheath was placed. A 5 Fr Kumpe catheter and 0.035 in. wire were used to select the shunt. Selective portomesenteric venogram demonstrated a patent shunt from portal vein to IVC, enlarged superior mesenteric vein (SMV), and normal caliber splenic vein (SV) [Figure 2A]. Pressures, both in the SMV and SV, were measured to be 7 mmHg. A 20 minute balloon occlusion test demonstrated a rise in pressure to 35 mmHg, and revealed the presence of a diminutive portal vein supplying the right lobe of the liver [Figure 2B]. Following occlusion testing, a 12 × 8 mm Amplatzer plug (Abbott Medical, USA) was deployed into the portosystemic shunt [Figure 3A, B]. Postprocedural ultrasound documented patency of SMV and SV with minimal flow to the IVC through the Amplatzer plug. The patient was observed in the ICU on a heparin drip for 3 days. Follow-up ultrasound revealed complete occlusion of the portosystemic shunt and patency of the superior mesenteric vein. There was normalization of serum ammonia levels on one-month follow-up, and no clinical or ultrasonological evidence of portal hypertension was found.

### Case 2

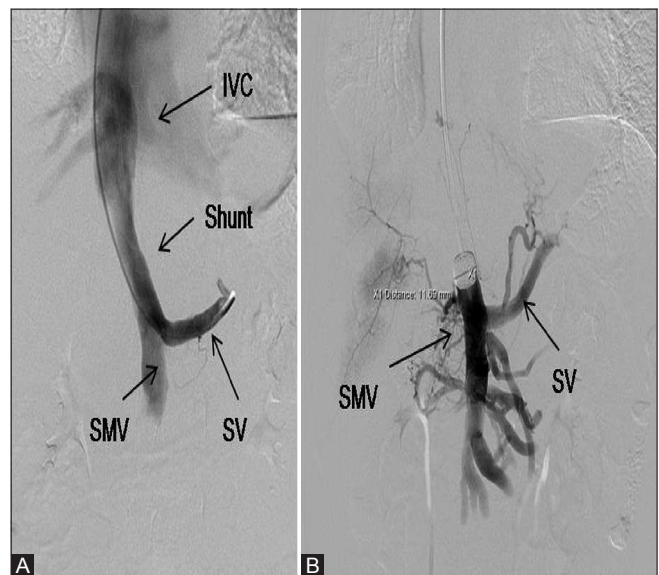
A 14-year-old female presented to the Emergency Department with nonspecific right-sided abdominal pain. Her past medical history was significant for severe scoliosis that was repaired surgically. Routine laboratory parameters were negative. CT scan with contrast revealed an incidental

portosystemic shunt between the SMV and the right internal iliac vein (IIV), consistent with Abernethy Type 2c malformation [Figure 4A]. Gamma-glutamyltransferase, ceruloplasmin, antinucleotide antibody, anti-liver-kidney microsomal, and alpha-1 antitrypsin (M1M1) levels were normal. Antismooth muscle antibody titers were positive, measuring 1:80. Although asymptomatic from the shunt, she was referred to interventional radiology for prophylactic endovascular closure to minimize risk of future complications.

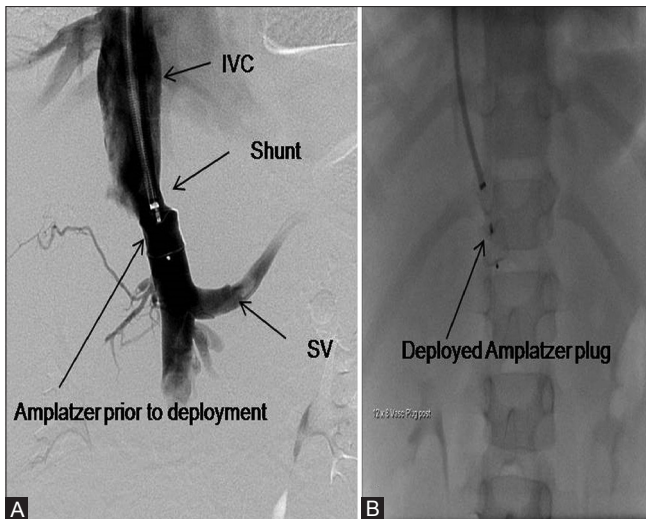
The planning procedure included transjugular liver biopsy and pressure measurements. The biopsy was normal and there was no significant portosystemic gradient. Then, right femoral arterial and venous access was obtained. Inferior mesenteric arteriograms with delayed imaging of venous drainage of the left colon and superior rectum demonstrated drainage through the large shunt into the systemic circulation (drainage from the SMV into the right IIV through the shunt), thus showing reversal of flow [Figures 4B and 5A]. This was repeated after balloon occlusion at the proximal and distal [Figure 5B] ends of the shunt through femoral venous approach. No significant increase in portal pressures was observed post balloon occlusion. Therefore, a one-step shunt closure was performed. Coil embolization of a short segment of the shunt was performed using multiple Concerto coils (Medtronic, USA) closer to the iliac vein in the pelvis with the intent of reversing left colonic venous drainage into inferior mesenteric vein as well as preserving the drainage of rectum and sigmoid colon. Postembolization venogram demonstrated occlusive coil packing with stasis of flow through the shunt [Figure 6A]. The patient was started on an anticoagulation regimen for 3 months. Six months follow-up CT revealed complete resolution of the shunt [Figure 6B].



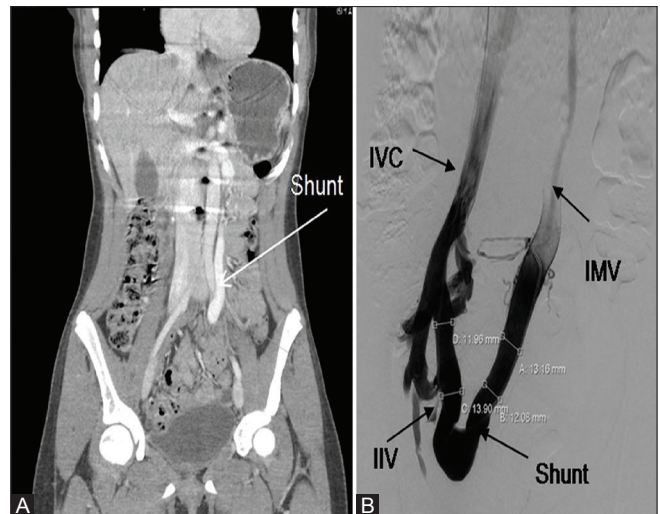
**Figure 1 (A and B):** Axial MRA demonstrating Type 1 portosystemic shunt with portal vein to the IVC (A and B)



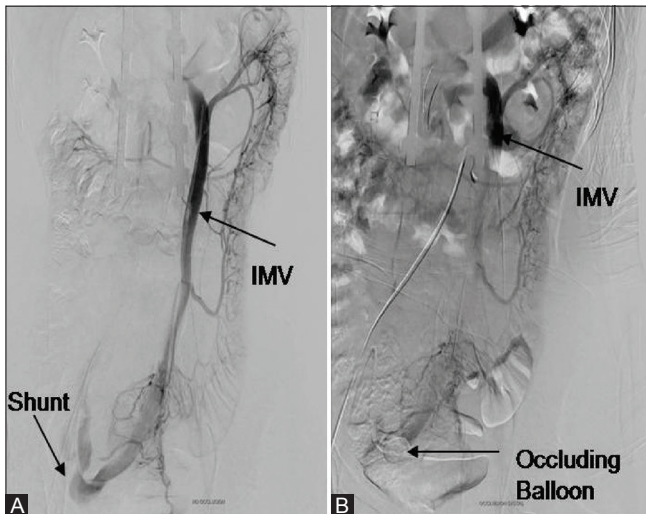
**Figure 2 (A and B):** SV venogram demonstrating a patent shunt to the IVC (A). Balloon occlusion venogram with hypoplastic portal veins (B)



**Figure 3 (A and B):** Balloon occlusion pressure measurements in the proximal shunt demonstrated no significant increased portal pressure (not shown). Venogram prior to Amplatzer plug deployment (A) and fluoroscopic spot image post deployment of Amplatzer plug (B)



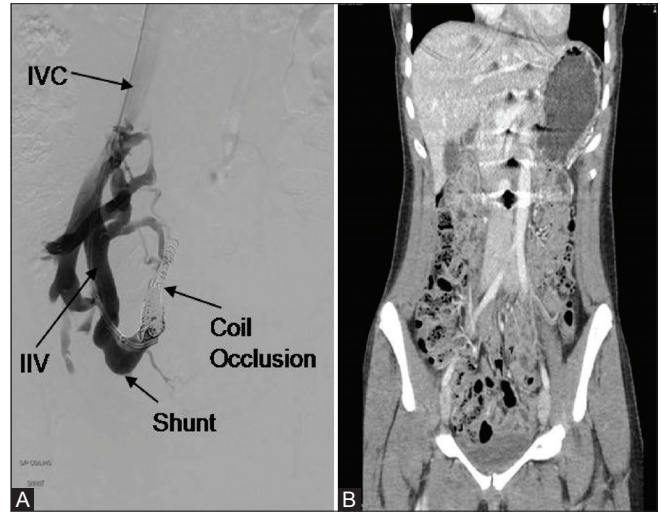
**Figure 4 (A and B):** Coronal (A) CT image showing an enlarged Type 2C portosystemic shunt between SMV and right IIV. Selective inferior mesenteric vein (IMV) venogram demonstrating the IMV with reversed flow to right IIV shunt, Type 11c (B)



**Figure 5 (A and B):** Delayed inferior mesenteric artery (IMA) angiography without balloon occlusion (A) demonstrates venous outflow of the transverse colon through the sigmoid colon drained into an enlarged IMV with patent shunt. IMA angiography with distal balloon occlusion (B) shows colonic drainage can be reversed via shunt

## Discussion

CEPS is classified into two types depending on the absence (Type 1) or presence (Type 2) of the portal trunk and intrahepatic portal system<sup>[2,3]</sup> [Table 1]. The clinical presentation of CEPS is heterogeneous, ranging from asymptomatic cases with incidental imaging findings to cases which present with clinically significant complications. Complications that necessitate closure include hepatic encephalopathy, pulmonary hypertension, pulmonary arteriovenous shunting, and hepatopulmonary syndrome.<sup>[1,4]</sup> CEPS is also associated with cardiac, gastrointestinal, vascular, skeletal, and genitourinary anomalies.<sup>[3-5]</sup>



**Figure 6 (A and B):** Post coil embolization venogram showing shunt occlusion (A). Six months follow-up coronal CT shows complete resolution of the shunt (B)

A multimodality approach is used to delineate the complex anatomy of these vascular malformations. Doppler ultrasonography is an initial noninvasive modality that does not expose patients to sedation or ionizing radiation, and can be used for monitoring.<sup>[1,3]</sup> Shunt course and patency of portal branches is further evaluated by CT or MRA. Conventional angiography is the standard of reference for evaluation of portomesenteric vasculature.<sup>[1]</sup>

Currently no standard treatment guidelines have been formulated due to rarity of CEPS.<sup>[5,6]</sup> In symptomatic cases, shunt closure is mandatory. In asymptomatic cases, there is discrepancy regarding management; some authors advocate early shunt closure while others



**Table 1: Classification of CEPS<sup>[2,3]</sup>**

Type of shunt	Description
Type 1	1a SMV and SV drain separately into a systemic vein
No intrahepatic portal flow	1b SMV and SV join to form a common trunk before draining into a systemic vein
Type 2	2a Origin of shunt from right/left portal vein branches or a patent ductus venosus
Preserved intrahepatic portal flow	2b Origin of shunt from main portal vein including its bifurcation of the confluence of its tributaries
	2c Origin of shunt from SV or mesenteric veins

SMV=superior mesenteric vein, SV=splenic vein

recommend active surveillance.<sup>[4,6-8]</sup> Shunt closure leads to stabilization if not resolution of complications associated with CEPS. Hence, we recommend early prophylactic closure, unless spontaneous closure occurs before the age of 1 year.

In Type 1 shunts, the plasticity of the intrahepatic portal system may enable revascularization following shunt closure, thus reducing the need for liver transplantation—the hitherto gold standard treatment.<sup>[1,6,9]</sup> Type 2 shunts, traditionally treated by surgical closure, are now amenable to endovascular treatment by coil or plug embolization. Treatment method used ultimately depends on the anatomy of the shunt, patient profile, and local expertise. Initially, angiograms are performed to demonstrate shunt anatomy and flow dynamics, which include transhepatic pressures, indirect portosystemic shunt pressure gradients, and direct pre- and post balloon shunt occlusion pressure gradients. These parameters assess the risk of development of portal hypertension post shunt occlusion.<sup>[1,4,6]</sup> If the risk is insignificant, direct embolization is indicated; otherwise, a staged procedure is performed with initial partial occlusion to acclimatize the intrahepatic portal system to increased flow.<sup>[9]</sup>

Interventional radiological techniques are minimally invasive, clinically efficacious, and associated with relatively few risks. Consequently, endovascular therapies have played an increasing role in the diagnostic and clinical management of CEPS.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### References

- Bernard O, Franchi-Abella S, Branchereau S, Pariente D, Gauthier F, Jacquemin E. Congenital portosystemic shunts in children: Recognition, evaluation, and management. *Semin Liver Dis* 2012;32:273-87.
- Srinivasa RN, Beecham Chick JF, Chen N, Gemmete JJ, Saad WA, Dasika NL, *et al.* Pediatric portal interventions. *Semin Intervent Radiol* 2018;35:143-50.
- Alonso-Gamarra E, Parrón M, Pérez A, Prieto C, Hierro L, López-Santamaría M. Clinical and radiologic manifestations of congenital extrahepatic portosystemic shunts: A comprehensive review. *Radiographics* 2011;31:707-72.
- Shah A, Aziz A, Awwad A, Ramjas G, Higashi Y. Incidental radiological diagnosis of asymptomatic Abernethy malformations—two case reports. *BJR Case Reports* 2017;3:20150496. doi: 10.1259/bjrcr.20150496.
- Murray CP, Yoo SJ, Babyn PS. Congenital extrahepatic portosystemic shunts. *Pediatr Radiol* 2003;33:614-20.
- Timpanaro T, Passanisi S, Sauna A, Trombatore C, Pennisi M, Petrillo G, *et al.* Congenital portosystemic shunt: Our experience. *Case Rep Pediatr* 2015;2015:691618. doi: 10.1155/2015/691618.
- Sokollik C, Bandsma RH, Gana JC, van den Heuvel M, Ling SC. Congenital portosystemic shunt: Characterization of a multisystem disease. *J Pediatr Gastroenterol Nutr* 2013;56:675-81.
- Yamagami T, Yoshimatsu R, Matsumoto T, Terayama K, Nishiumra A, Maeda Y, *et al.* Successful embolization using interlocking detachable coils for a congenital extrahepatic portosystemic venous shunt in a child. *J Pediatr Surg* 2007;42:1949-52.
- Elnekave E, Belenky E, Van der Veer L. Noncirrhotic extrahepatic portosystemic shunt causing adult-onset encephalopathy treated with endovascular closure. *Case Rep Radiol Case Rep Radiol* 2015;2015:852853. doi: 10.1155/2015/852853.