The Abernethy malformation—myriad imaging manifestations of a single entity

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Abstract

Abernethy malformation, also known as congenital extrahepatic portosystemic shunts (CEPS) is a rare clinical entity and manifests with different clinical symptoms. CEPS are abnormalities of vascular development where there is shunting of portal blood into the systemic venous system. Multidetector computed tomography (MDCT) is a fast and effective modality for evaluation of CEPS. CT displays all the information desired by the surgeon as well as the clinician including the anatomy of the splenic and superior mesenteric veins, size and site of the shunt, presence or absence of the portal vein radicles, and helps to plan the therapy and even the follow-up of these patients. Contrast-enhanced magnetic resonance imaging (MRI) has also emerged as a promising tool for the evaluation of liver lesions associated with the malformation. The Radiologist should be aware of the various imaging appearances of this entity including its complications. In this article, we describe the imaging appearances of CEPS, their complications, and their imaging appearances on CT and MRI. We have also described various associated anomalies.

Key words: Abernethy malformation; congenital extrahepatic portosystemic shunts; portosystemic shunts

Introduction

Congenital extrahepatic portosystemic shunt (CEPS) is a condition in which portal blood is shunted partially or completely into the systemic circulation via an abnormal communication of the portal system with the systemic circulation. The first account of this malformation was provided by John Abernethy in 1793[1], after whom it is also known as the Abernethy malformation.

The clinical presentation of these patients is varied and these shunts are often unsuspected and picked up either on ultrasound or computed tomography/magnetic resonance imaging (CT/MRI) for evaluation of the varied symptomatology they cause, which includes jaundice, difficulty in breathing, cyanosis, clubbing, and abdominal mass. Multidetector CT (MDCT) is a fast and effective modality for the evaluation of patients with suspected or confirmed portocaval shunts; it displays all the information desired by the surgeon and the clinician including the anatomy of the splenic and superior mesenteric veins (SMV), size and site of the shunt, presence or absence of the portal vein (PV) radicles, and helps to plan the therapy and even the follow-up of these patients. MRI may provide similar information, and though CT has the advantage in speed and spatial resolution; however, MRI scores in the characterization of liver lesions and patients who need long term follow up.

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Anatomy of the Normal Portal Vein

Normally, the venous drainage of the abdomen consists of two separate circulations—the systemic venous drainage and the portal circulation. The PV is formed by the union of the SMV with the splenic vein (SV). The portal system carries blood into the liver.[2] Blood exits the liver through the hepatic veins.

In normal individuals, there are no anatomical connections between the components of the portal system and the systemic or hepatic veins within or outside the liver.[3]

A portosystemic shunt leads to "short circuiting" of all or parts of portal blood, which passes into the systemic circulation without perfusing the liver.

Classification of Congenital Portosystemic Shunts

Congenital portosystemic shunts are classified into intrahepatic and extrahepatic. In the intrahepatic shunts, the shunt is at the level of PV branches after its division whereas in the extrahepatic shunts the anastomoses are established between the tributaries of the portal or mesenteric system or main PV and a systemic vein [Figure 1].[4]

Intrahepatic shunts

These are communications between the branches of the PV and inferior vena cava (IVC). The first case was described by Doehner *et al*. in 1956,[5] and these were further classified by Park *et al*.[6] in 1990, as follows:

- Type 1—Single tube-like vessel connecting the right branch of PV to IVC [Figure 2]
- Type 2—Localized peripheral shunt in which one hepatic segment has communications between the peripheral branches of the PV and the hepatic veins
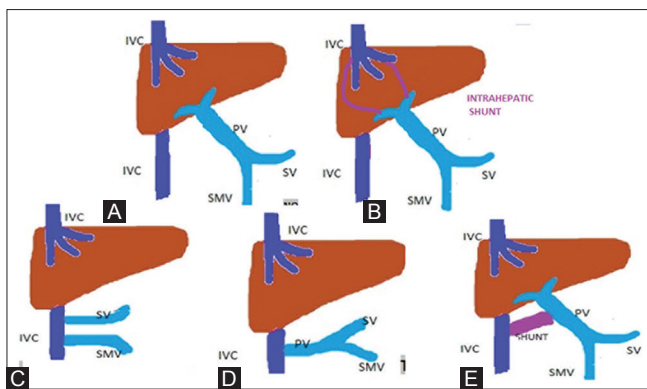

Figure 1 (A-E): Normal portovenous anatomy: (A) Intrahepatic shunt (B). Types of CEPS. Type 1a: Splenic vein and superior mesenteric vein drain directly into the inferior vena cava (IVC) (C). Type 1b: Portal vein drains directly into the IVC (D). Type 2: Shunt between portal vein and IVC (E)

- Type 3—Aneurysmal communication between peripheral PV and hepatic vein
- Type 4—Multiple intrahepatic shunts in both lobes of liver.

Congenital extrahepatic portosystemic shunts

CEPS were classified by Morgan and Superina[7] into the following types [Figure 1]:

- Complete—End to side with absent intrahepatic PV branches
  o SV and SMV drain separately into a systemic vein (type 1a)
  o SV and SMV join to form a common trunk which drains into a systemic vein (type 1b)
- Incomplete—Intrahepatic PV radicles are present, however, there is partial diversion of the portal blood into a systemic vein through a side-to-side shunt (type 2).

Embryological Basis of Congenital Extrahepatic Portosystemic Shunts

In the embryo, the right and left vitelline veins emerge from the yolk sac. At approximately 4 weeks, communications develop between the vitelline veins. They anastomose with each other to form a figure of 8 around the developing duodenum. Selective involution of these veins yields the final configuration of the PV.[8,9]

As the portal vein is formed by the selective involution of the periintestinal vitelline venous loop, abnormal patterns of involution may result in a preduodenal, prebiliary, or duplicated portal vein. Excessive involution may result in an absent portal vein. The absence of the portal vein thus results due to abnormal development in weeks
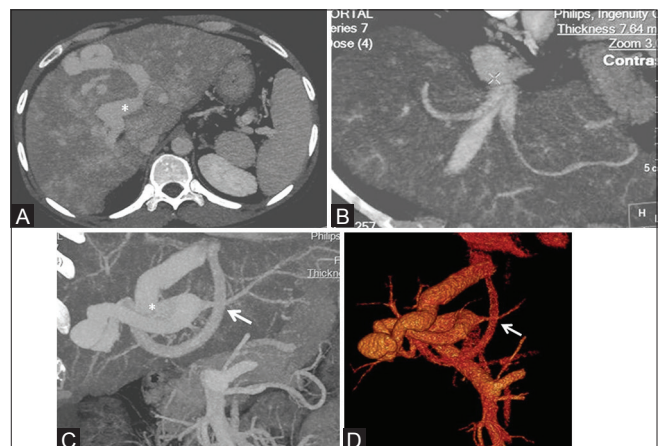

Figure 2 (A-D): An 8-year-old child presenting with cyanosis, CT angiographic MIP images (A) showing a well-defined large tortuous intrahepatic shunt (*) between the main portal vein and the middle hepatic vein. The middle hepatic vein is enlarged (B). Reconstructed coronal MIP (C) and VR images (D) showing another shunt between the left portal vein and left hepatic vein (arrow) and the large intrahepatic shunt

4–10 of gestation.[10,11] Because the vena cava also has a complex development and is derived from several venous channels including the sinus venosus and a portion of the anastomosis between the right and left vitelline veins in the cranial part of the liver,[12] it has been suggested that this may be the embryological basis of development of congenital extrahepatic portosystemic shunts.[13]

Clinical Features of Congenital Extrahepatic Portosystemic Shunts

CEPS may be asymptomatic well into adulthood and many authors have reported seeing adult patients presenting for the first time with clinical manifestations.[14-16]

Clinical features of portosystemic shunts may broadly be divided into:
o Features related to the shunting of portal blood
o Features secondary to associated congenital abnormalities
o Features secondary to hepatic lesions.

Symptoms related to the shunt
Hepatopulmonary syndrome
HPS is characterized by the triad of arterial deoxygenation (a widened PA–a O_2 with or without hypoxemia), intrapulmonary vascular dilatation, and liver disease.[17] It occurs secondary to diversion of vasoactive mediators into the systemic circulation. There is consequent dilatation of the intrapulmonary vessels and AV shunting with resultant hypoxemia as well as an element of ventilation/perfusion mismatch.[6,17]

This syndrome was first described in a patient with Abernethy malformation by Alvarez *et al.*;[18] it has subsequently been reported in a number of children with congenital portosystemic shunts.[2,19,20] Patients frequently present with cyanosis and digital clubbing and are generally investigated for cardiac and pulmonary shunts. Pediatric patients presenting with structurally normal echocardiograms and unexplained cyanosis should be investigated to rule out the possibility of a CEPS with HPS, even if no previous history is present [Figure 3].

Metabolic dysfunction
In patients with congenital portovenous shunts, including patent ductus venosus, blood from the mesenteric circulation bypasses the liver and goes directly into the systemic circulation; thus, toxic compounds which are removed in normal subjects by the liver, pass directly into the systemic circulation. These patients often have hyperammonemia and galactosemia.[2,6,20] Congenital portovenous shunts are often detected early in countries that screen galactose levels in new-borns.[21] Other symptoms related to the shunts/hepatic dysfunction include pulmonary hypertension,[20,22] hyperandrogenism, primary amenorrhea, or signs of virilisation.[23]
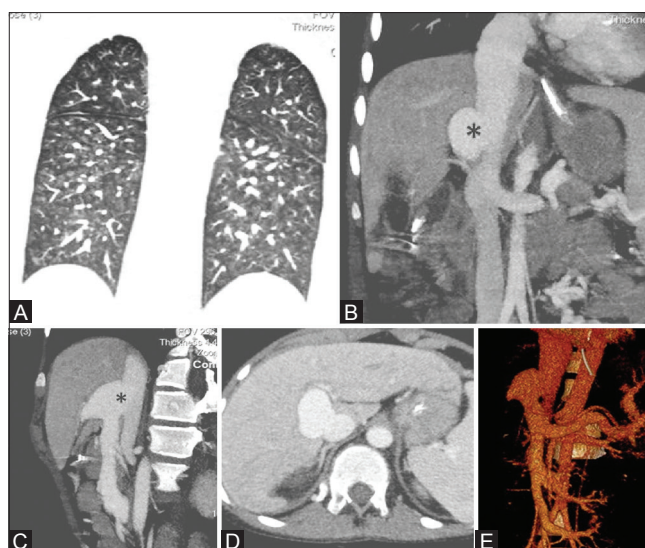

Figure 3 (A-E): A 13-year-old child presenting with altered sensorium, hematemesis, and seizures, Coronal reconstructions showing increased pulmonary vascularity seen as increased interstitial markings (A). Coronal MIP images (B) showing enlarged portal vein draining directly into the IVC through a side-to-side shunt (*). Sagittal MIP image (C) showing the portovenous communication. Axial sections (D) through liver do not reveal any well-defined portal branches. VR image (E) showing the same along with focal prominence of the portal vein

Hepatic encephalopathy
A number of patients present with neurological symptoms and symptoms due to portosystemic encephalopathy and increased blood ammonia levels.[2,6,15,20] These have been recorded as early as 18 months with a mean age of 6 years.[20] However, portosystemic encephalopathy is rarely observed in patients who have CEPS with mild hyperammonemia, and CEPS patients remain almost asymptomatic before suddenly developing hepatic encephalopathy.[24] A variety of explanations have been put forward for the same, including increasing sensitivity of the brain to ammonia or other toxic metabolites with age.[25] It has also been postulated that the shunt ratio may play a role in the occurrence of symptoms.[26] High signal intensity has been observed on T1-weighted MRI images in globus pallidus of several children who had CEPS due to deposition of manganese.[11,27,28] This has also been reported in other patients with hepatic encephalopathy.

Congenital anomalies associated with congenital extrahepatic portosystemic shunts
A number of congenital lesions have been reported in patients with CEPS, as tabulated in Table 1. Patients with type 1 shunt have a female preponderance[13,22] and often have concomitant congenital anomalies. These anomalies are less common in patients in type 2 shunts.[5,6,13] Other anomalies have also been reported in patients with Abernethy malformation which include chromosomal anomalies such as Downs syndrome and structural anomalies of the heart, gastrointestinal, genitourinary, skeletal, and vascular systems.[2,5,6,13,20,29,30]

Table 1: Difference between type 1 and 2 CEPS

	Type 1	Type 2
Sex predilection	Female preponderance	No sex preponderance
Pathology	Congenital absence of PV	PV supply is partially conserved
Associated anomalies	Concomitant congenital anomalies such as congenital heart disease and renal tract anomalies	Fewer associated malformations
Age at presentation	Younger age	Later in life
Portal vein and its communication	The portal vein typically drains into the retrohepatic IVC anywhere between a point just inferior to the hepatic vein confluence to just inferior to the level of the renal veins	The intrahepatic portal vein may be absent or hypoplastic
Type of anastomosis	End-to-side anastomosis	Side-to-side anastomosis

CEPS: Congenital extrahepatic portosystemic shunts, IVC: Inferior vena cava

Cardiac anomalies

A number of cardiac anomalies have been reported in patients with CEPS. These are postulated to develop because there is a close relationship between the development of the heart and the vitelline veins in embryonic life, as described above.[13] It has been proposed that cardiac development may be affected by the systemic diversion of portal venous flow.[29]

The original report by Abernethy[1] included findings of right-sided heart and right aortic arch. A number of authors have reported cases of CEPS with associated cardiac anomalies including atrial septal defect (ASD), ventricular septal defect (VSD), atrioventricular septal defect, aortic stenosis, pulmonary stenosis, coarctation of aorta, and patent ductus arteriosus.[2,5,20,29,30] [Figure 4].

Venous anomalies

A number of venous anomalies are seen in patients with CEPS, which include duplicated SVC,[1] duplicated IVC,[31] left-sided IVC,[9] azygous, and hemiazygous continuation of the IVC.[1,32]

Visceral arterial anomalies

Patients who have Abernethy malformation have also been reported to have arterial anomalies in the upper abdomen. These include enlarged hepatic arteries,[33] which have been postulated to represent a compensatory phenomenon.[2]

Visceral anomalies

A large number of visceral anomalies have been reported in patients with CEPS. All these anomalies with the exception of genitourinary anomalies are more common in patients with type 1 shunts;[22] the various anomalies reported include:
- Hepatobiliary—Congenital biliary atresia, choledochal cysts, congenital hepatic fibrosis, malrotation, annular pancreas, and inhtrahepatic gall bladder.[2,6,9,13,22,31,34]
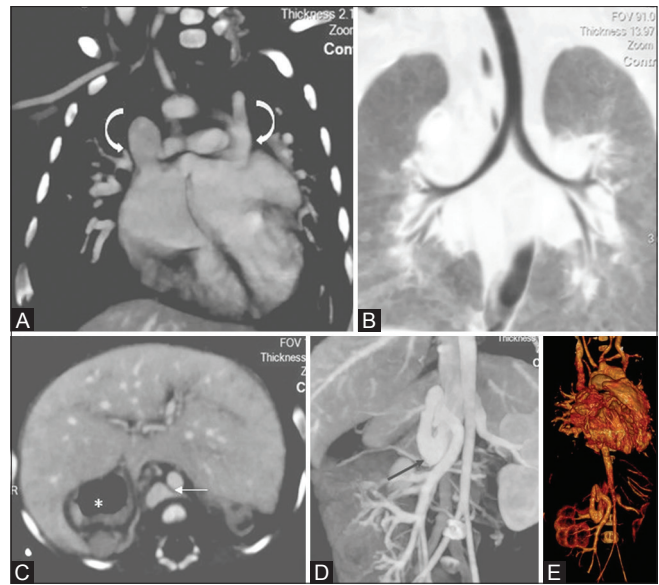- Genitourinary—Cystic dysplasia of the kidneys,

Figure 4 (A-E): A 2-month-old child presenting with severe pulmonary hypertension and cyanosis, Coronal reformatted MIP image (A) showing double SVC (curved arrow) with a small ASD. Coronal MINIP image (B) showing left bronchial isomerism. Axial section through liver (C) shows centrally placed liver with stomach on right side (*) and interrupted IVC (white arrow). Coronal reformatted MIP image (D) showing drainage of superior mesenteric vein through a well-defined shunt (black arrow) into interrupted IVC/azygous vein. No well-defined intrahepatic portal vein is seen. VR image (E) depicting the same

pelviuretic junction obstruction, crossed fused ectopic kidney, and hypospadias.[2,6,22,35]
- Splenic—Polysplenia and splenomegaly with hypersplenism[1,9,13,36] [Figure 4].

Skeletal anomalies

Skeletal anomalies described in CEPS include radial hypoplasia, congenital absence of the first metacarpophalangeal joints, vertebral anomalies, Goldenhars syndrome, maxillary hypoplasia, short fifth fingers.[2,22,37]

Hepatic lesions in patients with congenital extrahepatic portosystemic shunts

Nodular liver lesions have been reported not only in patients with congenitally absent PV but also in many other conditions where patients have disturbances of hepatic circulation.[38,39] Most of these lesions are asymptomatic and are either picked up on imaging done for other indications or seen during further evaluation of a patient of CEPS. Rarely a patient may present with an abdominal mass. It has been suggested that approximately half the patients of CEPS have nodular lesions of some sort in the liver.[6] The increased incidence of these lesions has been attributed to hepatic ischemia with compensatory increase in arterial flow and lack of growth factors and hormones due to reduced/absent portal flow.[2,6]

Nodular hepatic lesions in patients with congenital portosystemic shunts may be single or multiple, and include regenerative nodular hyperplasia,[20,37,40] focal nodular hyperplasia,[20,33,34] Hepatic adenomas/hepatic adenomatosis[16,20] [Figures 5 and 6] and hepatoblastoma.[41,42]

Hepatocellular carcinoma has been reported by a number of authors as an association with the Abernethy malformation.[43-45] Hepatocellular carcinomas/malignant lesions may develop on follow-up of benign nodular lesions in patients with CEPS or may coexist with other such lesions.

Regenerative nodules are usually homogenous with enhancement during arterial phase, on both CT and MRI, however without washout [Figure 6]. "Halo sign" has been described for adenomas where there is a hypodense rim seen surrounding the enhanced lesion. These lesions are usually hyperintense on T1-weighted images.[6,39] Hepatocellular adenomatosis has been reported in patients with Abernethy malformation.[16]

The imaging findings in patients with Abernethy malformation with Hepatocellular carcinoma do not appear to be typical, that is hypervascularity on the arterial phase images with washout on delayed phase. [Figure 7]. Thus, patients who do not have typical findings of a benign lesion, i.e. lack of arterial enhancement, or arterial enhancement without washout, should be closely followed up or biopsied.

Imaging of Congenital Extrahepatic Portosystemic Shunts

The cardinal imaging feature of a CEPS is an abnormal communication between the portal venous system and a systemic vein, either before or after the formation of the PV by union of the SMV and SV.

The first step in the diagnosis of the CEPS is to demonstrate communication between the portal and the systemic venous system. These may be end-to-side shunts where the PV terminates in the IVC (Type I shunts) or side-to-side shunts between the PV and IVC.[13]

The second step entails ruling out the acquired causes of nonvisualization of the PV such as portal cavernoma or PV thrombus. An acute PV thrombus is seen on cross-sectional imaging as a hypodense, nonenhancing PV filled with thrombus. As a rule, patients with congenital portosystemic shunts do not have features of portal hypertension,[6] such as splenomegaly, varices, and collaterals.

In type I shunts, the PV typically drains into the retrohepatic IVC anywhere between a point just inferior to the hepatic vein confluence[37] to just inferior to the level of the renal veins.[46] There are, however, descriptions of an abnormal PV ascending posterior to the liver and draining into the suprahepatic IVC[34] or into the right atrium.[9]

In type 2 shunts, the intrahepatic PV may be absent or hypoplastic, however, even if no PV radicles are seen on imaging, liver biopsy is suggested to confirm the presence or absence of portal venous radicles.[5,6]

In type 1 shunts, the PV or any of its constituents may drain into any Systemic vein, including the left renal vein,[22,47]
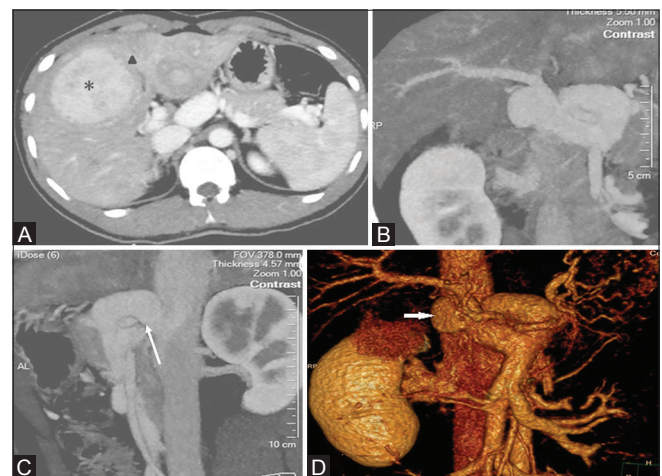

Figure 5 (A-C): An 8-year-old child, presenting with cyanosis, Axial images (A, B) showing multiple well-defined hypodense lesions showing arterial phase enhancement and becoming isodense on delayed images suggestive of adenomas. Reformatted sagittal MIP image (C) showing fistulous communication of main portal vein with IVC (white arrow). No appreciable intrahepatic portal branches are seen

Figure 6 (A-D): A 17-year-old, presenting with fever and loss of appetite, axial MIP image (A) showing multiple arterial phase enhancing lesions (*) (biopsy proven adenoma), one of them showing halo sign (white arrow head). MIP coronal image (B, C) showing aneurysmal dilatation of proximal portal vein. A tortuous dilated shunt is seen between portal vein and IVC (white arrow). Small intrahepatic portal branches are however seen. VRT image (D) shows the same (white arrow showing the shunt)
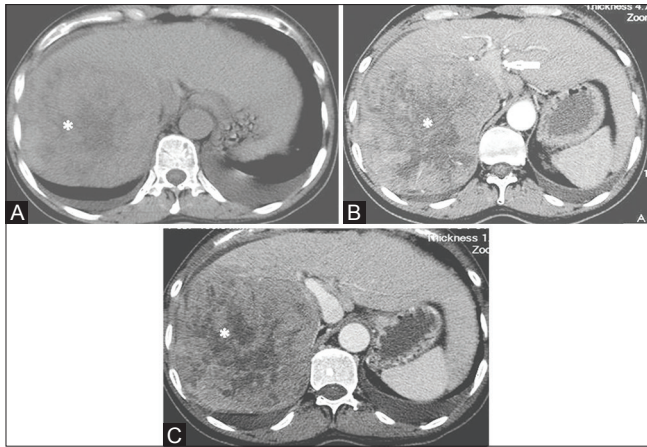
Figure 7 (A-C): A 40-year-old male, presenting with liver mass, axial sections through liver in plain (A) arterial (B) and venous (C) phases show a large arterial phase enhancing lesion showing washout on delayed phase suggestive of hepatocellular carcinoma (*) (biopsy proven). There is a large well-defined fistulous communication seen between the portal vein and IVC (white arrow). The intrahepatic branches of MPV are not well seen

Figure 8 (A-E): An 8-year-old child presented with cyanosis. Axial MIP images (A, B) of thorax reveals aneurysmal dilatation of pulmonary veins (white arrow) with numerous vascular channels likely pulmonary vascular malformations. Coronal and axial MIP images (C, D) show dilatation of splenic vein which unites with superior mesenteric vein and extends into retroperitoneum and then superiorly into the posterior mediastinum and drains into left brachiocephalic vein suggestive of porto systemic shunt (*). The VRT image (E) showing portal vein draining into left brachiocephalic vein (*)

the right renal vein,[22] and intrathoracic vein [Figure 8].[48] Communications have also been reported between the inferior mesenteric vein or its tributaries and left or right internal iliac vein.[5]

Once the portosystemic communication has been demonstrated, the major role of imaging is to diagnose the type of shunt because it may have therapeutic implications. Distinguishing between intra and extrahepatic shunts is dependent on the demonstration of anatomical site of communication.

Imaging Modalities for the Diagnosis of Congenital Extrahepatic Portosystemic Shunts

Doppler ultrasonography
Doppler ultrasonography is a safe and noninvasive modality for the diagnosis of the intrahepatic vasculature and may demonstrate the shunt, including the hemodynamics involved such as the magnitude and direction of flow [Figure 9]. It may pick up congenital shunts preoperatively; however, it may not detect associated anomalies and may also be unable to evaluate the retroperitoneum well, particularly in adult patients.[1,8,49,50] Thus, smaller shunts, particularly type 1a may not be well picked up. Ultrasound may not fully characterize liver lesions seen in these patients. Associated anomalies and findings particularly lung and cardiac anomalies will not be defined on ultrasound.

Multidetector computed tomography angiography
A number of case reports and studies have documented the efficacy of CT in the diagnosis and management of patients with CEPS.[51] CT is a fast, noninvasive modality that elegantly demonstrates anatomy and pathology with
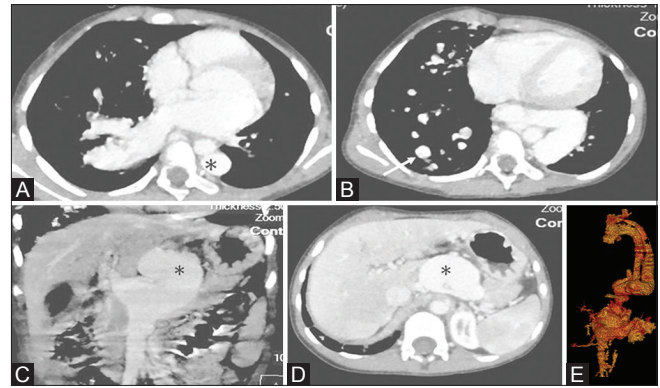
high spatial resolution. Though radiation may be a factor in deciding to choose MRI, the newer machines have a highly reduced radiation dose.[52] The major advantage of CT is that it clearly displays the portal anomaly and type of shunt and helps decide management. CT evaluates associated anomalies particularly in patients with congenital heart disease who require evaluation of pulmonary vasculature, or patients with suspected hepatopulmonary syndrome who require evaluation of the lungs.

Another major advantage of CT is that it helps to detect and characterize hepatic lesions in these patients. It displays the arterial and venous anatomy, including venous invasion if any, and provides an angiographic road map for surgical resection.

Magnetic resonance imaging
MRI has the ability to provide most of the information that CT does, however, it is slower, with need for longer periods of sedation which is a disadvantage in patients with CEPS who may be very young, very hypoxemic (due to hepatopulmonary syndrome) or encephalopathic (due to portosystemic shunting). MRI also has lower spatial resolution than CT and may not show small intrahepatic portal venous radicles in type 2 patients.

MRI, however may be superior, particularly with the advent of hepatobiliary contrast agents in the characterization of hepatic nodules [Figure 10].[39,53] MRI does not expose the patients to ionizing radiation. MRI should be used for serial follow up of hepatic lesions.

Other modalities
Porto Venography—Mesenteric portography has an advantage over visceral arteriography. It helps in identifying

the pressure gradient as well as in delineating the portal anatomy and extrahepatic shunts. However, it is an invasive procedure and is usually performed intraoperatively.[54,55]

Scintigraphy—rectal portal scintigraphy with rectal administration of iodine 123 (^{123}I) iodoamphetamine is a noninvasive quantitative method to determine portal hemodynamic and portal collateral circulation.[56,57]

Management of Patients with congenital extrahepatic portosystemic shunts

A variety of management strategies and therapeutic options are available for patients presenting with CEPS. Alonso-Gamarra *et al*[4] suggested a diagnostic algorithm as outlined below.

Asymptomatic patients/mild metabolic abnormalities should be followed-up with ultrasonography and biochemistry. Patients with liver nodules should also be followed up.

Symptomatic patients, that is those with portosystemic encephalopathy, liver dysfunction, or shunt ratio >60%, were divided into two groups according to the type of shunt—those with type 1 shunts should be transplanted, whereas those with type 2 shunts should be offered shunt closure—either interventional embolization or surgical.[58,59]

Patients with type 1 shunts may not be offered shunt occlusion because it represents the only route for intestinal and splenic blood drainage. Franchi-Abella *et al*.[20] recommended that shunts should definitely be closed whenever patients develop a complication; however, they further suggested that shunts should be closed in asymptomatic patients as well to prevent development of complications at a later date.

There are also other reports that suggest that hypoplastic PV branches are able to expand after the closure of shunt [Figure 11]. Liver transplantation is considered when medical and surgical methods fail especially in patients with complications.

Auxiliary portal orthotopic liver transplant is a recent development where a graft is placed along with native liver. It is useful in reversing fulminant hepatic failure. It is reported to be better than orthotopic liver transplant as lifelong immunosuppression is not required.[60]

Conclusion

Congenital extrahepatic shunts are being diagnosed with increased frequency as imaging techniques become more and more refined, and many are picked up incidentally. Imaging plays a major role in diagnosis, classification, and treatment of these patients. It is useful in pre and post treatment
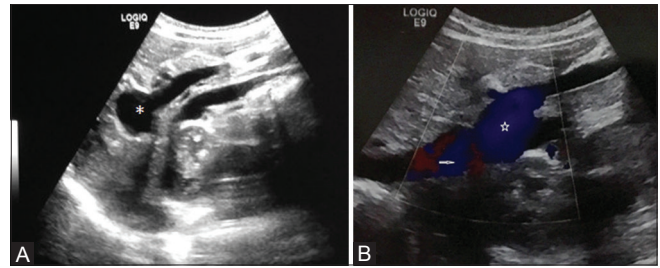

Figure 9 (A and B): Ultrasound images of a 13-year-old child (same patient in Figure 3), (A) shows bulbous dilatation of portal vein (*) and direct shunt with IVC (arrow). Color Doppler image (B) showing the direct communication
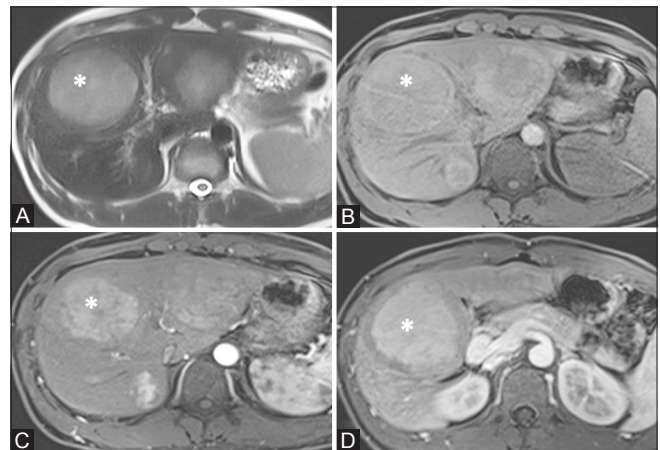

Figure 10 (A-D): MRI of a 17-year-old boy (same patient in Figure 6), Axial imaging in T2WI (A) showing multiple hyperintense lesions and T1WI (B) showing multiple hypointense lesions and post contrast T1 arterial phase image (C) showing arterial enhancement and appearing hyperintense on T1 venous phase image (D) suggestive of multiple hepatic adenomas. Larger one shown as (*)
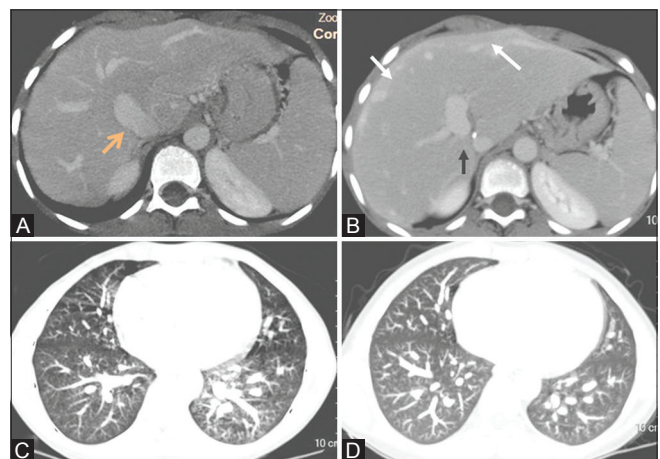

Figure 11 (A-D): Preoperative axial CT (A) shows well-defined shunt between IVC and portal vein (red arrow). Postoperative axial CT (B) shows a surgical staple (black arrow) at site of portosystemic shunt with numerous collaterals in the periphery of liver (white arrow). Pre and post-op axial sections through lung bases (C, D) showing reduction in vascular congestion

followup imaging as well as imaging of its complications. The radiologist holds a pivotal role in diagnosing the type of

Abernethy malformation and evaluating associated visceral anomalies and other complications.

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Conflicts of interest

There are no conflicts of interest.

References

1. Abernethy J. Account of two instances of uncommon formation, in the viscera of the human body. Philos Trans R Soc Lond B Biol Sci 1793;83:59-66.

2. Hu GH, Shen LG, Yang J, Mei JH, Zhu YF. Insight into congenital absence of the portal vein: Is it rare? World J Gastroenterol 2008;14:5969-79.

3. Hales MR, Allan JS, Hall EM. Injection-corrosion studies of normal and cirrhotic livers. Am J Pathol 1959;35:909-41.

4. 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;31:707-22.

5. Doehner GA, Ruzicka FF Jr, Rousselot LM, Hoffman G. The portal venous system: On its pathological roentgen anatomy. Radiology 1956;66:206-17.

6. Park JH, Cha SH, Han JK, Han MC. Intrahepatic portosystemic venous shunt. AJR Am J Roentgenol 1990;155:527-8.

7. Morgan G, Superina R. Congenital absence of the portal vein: Two cases and a proposed classification system for portasystemic vascular anomalies. J Pediatr Surg 1994;29:1239-41.

8. Marks C. Developmental basis of the portal venous system. Am J Surg 1969;117:671-81.

9. Stringer MD. The clinical anatomy of congenital portosystemic venous shunts. Clin Anat 2008; 21:147-57.

10. Komatsu S, Nagino M, Hayakawa N, Yamamoto H, Nimura Y. Congenital absence of portal venous system associated with a large inferior mesenteric-caval shunt: A case report. Hepatogastroenterology 1995;42:286-90.

11. Niwa T, Aida N, Tachibana K, Shinkai M, Ohhama Y, Fujita K, *et al*. Congenital absence of the portal vein: clinical and radiologic findings. J Comput Assist Tomogr 2002;26:681-6.

12. Dickson AD. The development of the ductus venosus in man and the goat. J Anat 1957;91:358-68.

13. Howard ER, Davenport M. Congenital extrahepatic portocaval shunts — the Abernethy malformation. J Pediatr Surg 1997;32:494-7.

14. Oei M, Wessling J. Abernethy malformation — Congenital aplasia of the portal vein a 29-year-old patient. Rofo 2007;179:167-9.

15. Witters P, Maleux G, George C, Delcroix M, Hoffman I, Gewillig M, *et al*. Congenital veno-venous malformations of the liver: Widely variable clinical presentations. J Gastroenterol Hepatol 2008;23(Pt 2):e390-4.

16. Asran MK, Loyer EM, Kaur H, Choi H. Case 177: Congenital absence of the portal vein with hepatic adenomatosis. Radiology 2012;262:364-7.

17. Porres-Aguilar M, Altamirano JT, Torre-Delgadillo A, Carlton MR, Duarte-Rojo A. Portopulmonary hypertension and hepatopulmonary syndrome: A clinician-oriented overview. Eur Respir Rev 2012;21:223-33.

18. Alvarez AE, Ribeiro AF, Hessel G, Baracat J, Ribeiro JD. Abernethy malformation: One of the etiologies of hepatopulmonary syndrome. Pediatr Pulmonol 2002;34:391-4.

19. Tercier S, Delarue A, Rouault F, Roman C, Bréaud J, Petit P. Congenital portocaval fistula associated with hepatopulmonary syndrome: Ligation vs liver transplantation. J Pediatr Surg 2006;41:e1-3.

20. Franchi-Abella S, Branchereau S, Lambert V, Fabre M, Steimberg C, Losay J, *et al*. Complications of congenital portosystemic shunts in children: Therapeutic options and outcomes. J Pediatr Gastroenterol Nutr 2010;51:322-30.

21. Ono H, Mawatari H, Mizoguchi N, Eguchi T, Sakura N. Clinical features and outcome of eight infants with intrahepatic porto-venous shunts detected in neonatal screening for galactosaemia. Acta Paediatr 1998;87:631-4.

22. Murray CP, Yoo SJ, Babyn PS. Congenital extrahepatic portosystemic shunts. Pediatr Radiol 2003;33:614-20.

23. Satoh M, Yokoya S, Hachiya Y, Hachiya M, Fujisawa T, Hoshino K, *et al*. Two hyperandrogenic adolescent girls with congenital portosystemic shunt. Eur J Pediatr 2001;160:307-11.

24. Goo HW. Extrahepatic portosystemic shunt in congenital absence of the portal vein depicted by time-resolved contrast-enhanced MR angiography. Pediatr Radiol 2007;37:706-9.

25. Wakamoto H, Manabe K, Kobayashi H, Hayashi M. Subclinical portal-systemic encephalopathy in a child with congenital absence of the portal vein. Brain Dev 1999;21:425-8.

26. Uchino T, Matsuda I, Endo F. The long-term prognosis of congenital portosystemic venous shunt. J Pediatr 1999;135(Pt 1):254-6.

27. Ikeda S, Sera Y, Yoshida M, Ohshiro H, Uchino S, Seguchi S, *et al*. Reversibility of hyperintense globus pallidus on T1-weighted MRI following surgery for a portosystemic shunt in an 8-year-old girl. Pediatr Radiol 1999;29:449-50.

28. Kumar A, Kumar J, Aggarwal R, Srivastava S. Abernethy malformation with portal vein aneurysm. Diagn Interv Radiol 2008;14:143-6.

29. Massin M, Verloes A, Jamblin P. Cardiac anomalies associated with congenital absence of the portal vein. Cardiol Young 1999;9:522-5.

30. Ratnasamy C, Kurbegov A, Swaminathan S. Cardiac anomalies in the setting of the Abernethy malformation of the portal vein. Cardiol Young 2007;17:212-4.

31. Gocmen R, Akhan O, Talim B. Congenital absence of the portal vein associated with congenital hepatic fibrosis. Pediatr Radiol 2007;37:920-4.

32. Le Borgne J, Paineau J, Hamy A, Dupas B, Lerat F, Raoul S, *et al*. Interruption of the inferior vena cava with azygos termination associated with congenital absence of portal vein. Surg Radiol Anat 2000;22:197-202.

33. De Gaetano AM, Gui B, Macis G, Manfredi R, Di Stasi C. Congenital absence of the portal vein associated with focal nodular hyperplasia in the liver in an adult woman: Imaging and review of the literature. Abdom Imaging 2004;29:455-9.

34. Kinjo T, Aoki H, Sunagawa H, Kinjo S, Muto Y. Congenital absence of the portal vein associated with focal nodular hyperplasia of the liver and congenital choledochal cyst: A case report. J Pediatr Surg 2001;36:622-5.

35. Guariso G, Fiorio S, Altavilla G, Gamba PG, Toffolutti T, Chiesura-Corona M, *et al*. Congenital absence of the portal vein associated with focal nodular hyperplasia of the liver and cystic dysplasia of the kidney. Eur J Pediatr 1998;157:287-90.

36. Appel H, Loddenkemper C, Schirmacher P, Dienes HP, Sieper J, Rudwaleit M, *et al*. Congenital absence of the portal vein with splenomegaly and hypersplenism in a young woman. Digestion 2003;67:105-10.

37. Grazioli L, Alberti D, Olivetti L, Rigamonti W, Codazzi F, Matricardi L, *et al*. Congenital absence of portal vein with nodular regenerative hyperplasia of the liver. Eur Radiol 2000;10:820-5.

38. Kondo F. Benign nodular hepatocellular lesions caused by

abnormal hepatic circulation: Etiological analysis and introduction of a new concept. J Gastroenterol Hepatol 2001;16:1319-28.

39. Brancatelli G, Federle MP, Grazioli L, Golfieri R, Lencioni R. Benign regenerative nodules in Budd-Chiari syndrome and other vascular disorders of the liver: Radiologic–pathologic and clinical correlation. Radiographics 2002;22:847-62.

40. Arana E, Martí-Bonmatí L, Martínez V, Hoyos M, Montes H. Portal vein absence and nodular regenerative hyperplasia of the liver with giant inferior mesenteric vein. Abdom Imaging 1997;22:506-8.

41. Kawano S, Hasegawa S, Urushihara N, Okazaki T, Yoshida A, Kusafuka J, *et al.* Hepatoblastoma with congenital absence of the portal vein—a case report. Eur J Pediatr Surg 2007;17:292-4.

42. Barton JW 3rd, Keller MS. Liver transplantation for hepatoblastoma in a child with congenital absence of the portal vein. Pediatr Radiol 1989;20:113-4.

43. Banz V, Olliff S, Taniere P, Mayer D, Isaac J. Liver tumours in patients with Abernethy malformation. ANZ J Surg 2011;81:640-1.

44. Pichon N, Maisonnette F, Pichon-Lefièvre F, Valleix D, Pillegand B. Hepatocarcinoma with congenital agenesis of the portal vein. Jpn J Clin Oncol 2003;33:314-6.

45. Lundstedt C, Lindell G, Tranberg KG, Svartholm E. Congenital absence of the intrahepatic portion of the portal vein in an adult male resected for hepatocellular carcinoma. Eur Radiol 2001;11:2228-31.

46. Joyce AD, Howard ER. Rare congenital anomaly of the portal vein. Br J Surg 1988;75:1038-9.

47. Kim SZ, Marz PL, Laor T, Teitelbaum J, Jonas MM, Levy HL. Elevated galactose in newborn screening due to congenital absence of the portal vein. Eur J Pediatr 1998;157:608-9.

48. Bonington SC, Hodgson DI, Mehta S, Lynch N, Chalmers N. A congenital venous anomaly, with a portal-systemic shunt into a previously undescribed intra-thoracic vein. Clin Radiol 2002;57:658-60.

49. Koizumi J, Yamashita T, Dowaki S, Tobita K, Shimamura K, Hirabayashi K, *et al.* Hepatobiliary and pancreatic: Hepatic adenoma, focal nodular hyperplasia and congenital absence of the portal vein. J Gastroenterol Hepatol 2006;21:619.

50. Konno K, Ishida H, Uno A, Ohnami Y, Masamune O. Large extrahepatic portosystemic shunt without portal hypertension. Abdom Imaging 1997;22:79-81.

51. Prokop M. Multislice CT angiography. Eur J Radiol 2000;36:86-96.

52. Morgan H, Braunstein D. A computed tomography dose index for infants and young children. Med Phys 2001;28:1818.

53. Trenschel GM, Schubert A, Dries V, Benz-Bohm G. Nodular regenerative hyperplasia of the liver: Case report of a 13-year-old girl and review of the literature. Pediatr Radiol 2000;30:64-8.

54. Krajina A, Lojik M, Chovanec V, Raupach J, Hulek P. Wedged hepatic venography for targeting the portal vein during TIPS: Comparison of carbon dioxide and iodinated contrast agents. Cardiovasc Intervent Radiol 2002;25:171-5.

55. Martinez-Cuesta A, Elduayen B, Vivas I, Delgado C, Gonzalez-Crespo I, Bilbao JI. CO (2) wedged hepatic venography: Technical considerations and comparison with direct and indirect portography with iodinated contrast. Abdom Imaging 2000;25:576-82.

56. Sura PA, Tobias KM, Morandi F, Daniel GB, Echandi RL. Comparison of 99mTcO4(-) trans-splenic portal scintigraphy with per-rectal portal scintigraphy for diagnosis of portosystemic shunts in dogs. Vet Surg 2007;36:654-60.

57. Shiomi S, Kuroki T, Ueda T, Takeda T, Ikeoka N, Nishiguchi S, *et al.* Clinical usefulness of evaluation of portal circulation by per rectal portal scintigraphy with technetium-99m pertechnetate. Am J Gastroenterol 1995;90:460-5.

58. Gallego C, Miralles M, Marín C, Muyor P, González G, García-Hidalgo E. Congenital hepatic shunts. Radiographics 2004;24:755-72.

59. Akahoshi T, Nishizaki T, Wakasugi K, Mastuzaka T, Kume K, Yamamoto I, *et al.* Portal-systemic encephalopathy due to a congenital extrahepatic portosystemic shunt: Three cases and literature review. Hepatogastroenterology 2000;47:1113-6.

60. Inomata Y, Kiuchi T, Kim I, Uemoto S, Egawa H, Asonuma K, *et al.* Auxiliary partial orthotopic living donor liver transplantation as an aid for small-for-size grafts in larger recipients. Transplantation 1999;67:1314-9.