Abernethy malformation: Our experience from a tertiary cardiac care center and review of literature

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ABSTRACT

Abernethy malformation, also called as congenital extrahepatic portosystemic venous shunt, is a rare anomaly involving the portal venous system. Although rare, it is increasingly being reported and is important to diagnose given the adverse clinical consequences in untreated patients. It has myriad of clinical presentations, from being completely asymptomatic to causing hepatic carcinoma, hepatic encephalopathy, severe pulmonary hypertension, and diffuse pulmonary arteriovenous malformation. We describe our experience with five cases in a tertiary pediatric cardiac care center with Abernethy malformation, with review of literature and also discuss possible therapeutic implications.

Keywords: Congenital extrahepatic portosystemic shunt, pulmonary arterial hypertension, pulmonary arteriovenous malformation

INTRODUCTION

Abernethy malformation or congenital extrahepatic portosystemic shunt (CEPS) as the name suggests, was first described by John Abernethy[1] and is an extremely rare condition in which the portal vascular system, which derives blood from abdominal organs drains into systemic circulation bypassing liver through a complete or partial shunt. As a result, toxins from the intestine are bypassed to systemic circulation leading to the clinical manifestation of liver dysfunction, hepatic encephalopathy, and hepatopulmonary syndrome. The clinical importance of recognizing this entity is in its association with cardiac and liver abnormalities. It is also important to identify the presence or absence of hepatic portal venous supply as this may influence treatment options. In this review, we present our experience of five cases of CEPS with varying cardiac presentation two of which had prior interventional cardiac procedure done. We also discuss the embryology, current clinical classification, common presenting features and associated congenital anomalies, and subsequently discuss available diagnostic and therapeutic modalities.

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MATERIALS AND METHODS

Five patients presented with varying clinical presentation and subsequently diagnosed to have Abernethy malformation. Clinical parameters for all five patients were evaluated and are summarized in Table 1. Parameters evaluated included age and sex, clinical presentation, route and drainage of shunt, associated lesions, status of liver, management, and outcome.

All patients underwent, apart from routine investigations, abdominal ultrasound and abdominal computed tomography (CT). Patient 2 and 3 underwent cardiac catheterization and angiography.

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Table 1: Summarizing all five cases with Abernethy malformation with clinical features and outcomes

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age	2 months	19 years	12 years	7 years	4 days
Sex	Female	Male	Male	Female	Female
Abernethy type	Type 2	Type 2	Type 2	Type Ib	Type 2
Presentation	Respiratory failure due to underlying heart disease	Easy fatigability	Easy fatigability and bluish discoloration	Increasing bluish discoloration	Tachypnea and desaturation
Shunt	SV draining into the left renal vein then into hemiazygous vein	PV draining into IVC but with preserved intrahepatic venules	SV draining into the left renal vein then into IVC	SV and SMV forming PVand draining into SVC, through azygous vein with absent intrahepatic venules	SV and SMV forming PV and draining into the liver, and a shunt connecting PV and IVC
Index of suspicion	Desaturation in postoperative period	IncreasedPASPonECHO PDA device closure in early childhood Mild transaminitis	Normal ECHO Contrast ECHO suggestive of PAVM	Normal ECHO Contrast ECHO suggestive of PAVM	Diagnosed antenatally on anomaly scan Post natal ECHO suggestive of PAH Increased ammonia levels
PAVM	Small PAVM in the left upper lobe	None	Diffuse	Diffuse	None
Associated abnormalities	Left isomerism, heterotaxy transitional atriaventricular septal defect	PDA	None	PDA, transaminitis	Transaminitis Elevated serum ammonia
Management	Plan for closure of PAVM and Abernethy malformation on follow up	Closure of abnormal communication with covered stent Planned for cardiac catheterization after 6 months	Closure of abnormal communication with vascular plug	Advised liver biopsy and liver transplantation Lost to follow-up	Started on sildenafil Follow-up

PV: Portal vein, PDA: Patent ductus arteriosus, ECHO: Echocardiography, PASP: Pulmonary arterial systolic pressure, IVC: Inferior vena cava, SV: Splenic vein, SMV: Superior mesentric vein, PAH: Pulmonary arterial hypertension, PAVM: Pulmonary arteriovenous malformation

RESULTS

Patient 1

A 2-month-old female child presented to us with diagnosis of complete atrioventricular canal defect in respiratory failure. In view of her clinical condition, the child was treated with mechanical ventilation. Her initial saturations were 64% which improved on mechanical ventilation to 85%–90%. Echocardiography done revealed left atrial isomerism, interrupted inferior vena cava (IVC) and azygous continuation, transitional atrioventricular canal defect with common atrium, small inlet ventricular septal defect, and bilateral superior vena cava with left superior vena cava draining to the left atrium. Initial persistent low saturations even after ventilation were thought to be due to the persistent left superior vena cava to the left atrium. After initial stabilization, the patient underwent single patch repair of atrial and ventricular septal defect with rerouting of the left superior vena cava to the right atrium. In the immediate postoperative period, the patient continued to have low oxygenation with saturations not going beyond 90% (PaO_2 55). A bubble contrast echocardiography was performed to look for any residual leak, but it demonstrated normal filling of the right atrium and ventricle with filling of the left atrium and ventricle within 2–3 beats raising the possibility of pulmonary arteriovenous malformation. CT of the chest and upper abdomen was done for confirmation of pulmonary arteriovenous malformation as well as to rule out possibility of CEPS as etiology. It revealed small pulmonary arteriovenous malformation in the left upper lobe, Type 2 Abernethy malformation, and polysplenia [Figure 1a-c]. The patient remained stable at the time of discharge with saturations maintaining between 90% and 93% with plan to do interventional closure of Abernethy malformation on follow-up.

Patient 2

A 19-year-old boy presented to us for evaluation with the complaints of easy fatigability. He was a known case of congenital heart disease with patent ductus arteriosus and had undergone device closure for the same at 4 years of age. He was a diagnosed as a case of CEPS at the age of 13 years, when he underwent preanesthetic workup, to repair an accidental tibial fracture, which revealed transaminitis and subsequent ultrasound abdomen revealed the presence of CEPS. Echocardiography done revealed no residual patent ductus arteriosus with device *in situ*. There was significant right atrial and ventricular dilatation with estimated high pulmonary arterial pressure (estimated right ventricular systolic pressure of 90 mmHg) with preserved right ventricular function. Presence of high pulmonary arterial pressures which was initially attributed to late closure of patent ductus arteriosus could

be contributed by the presence of high-flow extrahepatic portosystemic shunt, so he underwent CT of the abdomen to determine the type of shunt. CT demonstrated the presence of Type 2 Abernethy malformation with large window type communication between the portal vein and IVC [Figure 2a]. He was subsequently planned for angiography to assess hemodynamics as well as possible closure of communication. Hemodynamics data revealed pulmonary arterial pressure of 75/43 with the mean of 50 mmHg against systemic pressures of 111/64 with the mean of 80 mmHg. Before attempting to close the extrahepatic portosystemic communication, the mean portal venous pressure was documented to be 13 mmHg with no significant increase postballoon occlusion. Angiography revealed large window type communication with IVC [Figure 2b], so it was decided to use covered stent in the IVC to close the communication. A self-expandable aortic stent graft (Endurant II 36 mm × 36 mm × 49 mm – Medtronic) was used to close the communication. Poststent implantation, there was complete closure of the communication. Within 24 h of stent graft placement, ultrasound of the abdomen was done which revealed collapse of the stent graft and resultant reopening of the communication between the portal vein and IVC. He was again taken to catheterization laboratory and high-pressure balloon dilatation of the collapsed stent graft was attempted which was not successful. He then underwent a stent in stent implantation and a bare Andra Stent size 43 mm mounted on 18 mm × 5 cm Z-Med balloon after 1 month [Figure 2c]. This time there was no flow seen across the communication. At last follow-up, after 3 months, the patient is symptomatically improved with echocardiogram showing preserved ventricular function and estimated right ventricular systolic pressure of 52 mmHg. He is planned for recatheterization in 6 months to assess the impact of closure of extrahepatic shunt on pulmonary hemodynamics.

Patient 3

A 12-year-old boy presented with complaints of bluish discoloration and easy fatigability noticed since 4 years of age. Clinical examination revealed low oxygen saturations (74%) with normal cardiac examination. Echocardiogram done revealed no structural abnormality with bubble contrast demonstrating contrast appearing in the left atrium and ventricle within 2–3 beats suggestive of the presence of pulmonary arteriovenous malformation. Ultrasound abdomen and CT was done which revealed the presence of diffuse pulmonary arteriovenous malformation. A large communicating channel was seen from the distal splenic vein to distal left renal vein causing shunting of blood from the splenic vein to IVC with well-patent main portal vein and its intrahepatic branches (Abernethy Type 2) [Figure 3a and b]. The patient was taken up for angiography with plan to close the abnormal channel with a vascular plug.

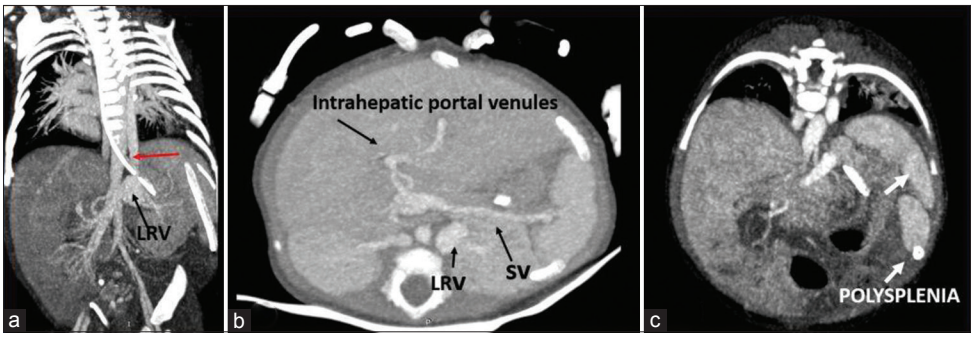

Figure 1: (a) Contrast-enhanced computed tomography abdomen coronal image showing left renal vein draining into Hemiazygous vein (red arrow). (b) Same image in an axial cut showing splenic vein draining into the left renal vein. (c) Oblique coronal cut showing polysplenia (white arrows)

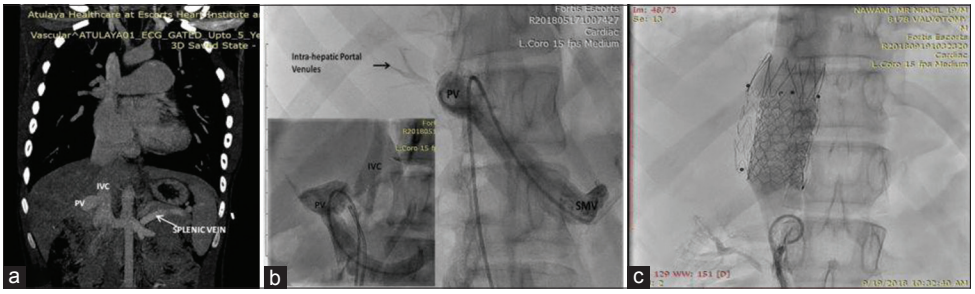

Figure 2: (a) Contrast-enhanced computed tomography abdomen coronal images showing Type 2 Abernathy malformation where the portal vein is seen draining into inferior vena cava. (b) Shows still image from intra-cardiac catheterization, wherein selective injection into the superior mesenteric vein shows filling of the portal vein, and then the portal vein is draining into the inferior vena cava. (c) Shows the abnormal connection being closed with the help of a covered stent

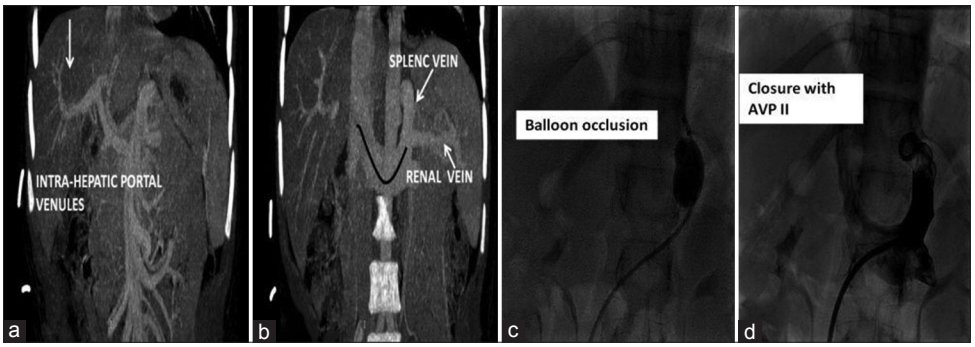

Figure 3: (a and b) Computed tomography abdomen coronal images showing distal splenic vein draining into left renal vein and then into inferior vena cava and also the presence of Intrahepatic portal venules is seen. (c) Showing balloon occlusion and measurement of portal pressure during the closure of malformation. (d) Shows complete closure of the shunt with no residual shunt across the device (amplatzer vascular plug II)

Hemodynamic data revealed normal pulmonary arterial pressures. Portal vein pressure was 12 mmHg with postballoon occlusion of channel pressure of 14 mmHg. IVC angiogram done revealed a large communication measuring 15 mm between the splenic vein and left renal vein. It was closed successfully with Amplatzer vascular plug size 22 mm (Abbott Medical) [Figure 3c and d].

Patient 4

A 7-year-old girl diagnosed as a case of congenital heart disease with patent ductus arteriosus, for which she underwent device closure at the age of 3 years at another hospital now presented with complaints of increasing bluish discoloration. At presentation, she had saturations of 68% with normal cardiac examination. Echocardiography done revealed patent ductus arteriosus device *in situ*; normal biventricular function but dilated left ventricle. Bubble contrast injection done through left brachial vein showed filling of the right atrium and ventricle, followed by filling of the left atrium and ventricle within 2–3 beats suggestive of pulmonary arteriovenous malformation. Subsequently, CT of the lungs confirmed the presence of diffuse pulmonary arteriovenous malformation involving bilateral lungs. The abdomen scan revealed the absence of both intra- and extrahepatic portion of portal vein with large tortuous communication between superior vena cava and splenoportal confluence suggesting diagnosis of Type 1 Abernethy malformation. The patient was advised further workup with pediatric gastroenterology department with liver biopsy and possible liver transplant; however, the patient was lost to follow-up.

Patient 5

A 4-day-old female child, born prematurely at 33 weeks by an elective cesarean section presented with tachypnea soon after birth. Antenatally, the fetus was suspected to have Abernathy malformation on Level II scan. The baby was shifted to neonatal intensive care unit (NICU). In the NICU, the baby developed significant desaturation along with mild tachypnea, for which nasal continuous positive airway pressure (CPAP) ventilation was started. Chest X-ray was normal. Sepsis screen was negative, but in view of clinical sepsis, antibiotics were started empirically. Echocardiography done outside revealed the features of severe pulmonary arterial hypertension (PAH) with patent foramen ovale shunting right to left and suspicion of abnormal pulmonary venous drainage to IVC. In view of the current findings, the child was referred to our center for further evaluation. At presentation, the child was stabilized. Echocardiography revealed normal pulmonary venous drainage and features of PAH. In view of antenatal suspicion of Abernethy malformation, CT pulmonary angiogram was done which confirmed the diagnosis of Abernethy malformation Type 2. Other investigations showed mild elevation of serum ammonia levels. In view of neonatal PAH, sildenafil was started, and the child was weaned off from nasal CPAP. Antibiotics were stopped after blood culture was sterile. The child was subsequently shifted back to neonatal unit with a plan to follow-up the child for pulmonary artery pressures and signs of hepatic dysfunction.

DISCUSSION

Congenital portosystemic shunt is defined by the presence of an atypical communication between the portal venous system and systemic venous system, and subsequently, blood from mesenteric circulation is improperly shunted to systemic circulation bypassing the liver. It can be either intrahepatic or extrahepatic portosystemic shunt (CEPS).

CEPS is a rare condition initially well described in animals, particularly dogs.[2] John Abernethy[1] in his "Account of two instances of uncommon formation in the viscera of the Human Body," in 1793 first described a portal venous malformation which he discovered during a postmortem examination of 10-month-old girl. This malformation was named subsequently as Abernethy malformation and consisted of CEPS.

Embryology

The development of the portal venous system is extremely complex and occurs between the 4th and 10th week of embryonic life.[3] The portal vein is formed from the vitelline vein, a pair of vessels located on the anterior surface of the yolk sac. Aberrations in this process may result in anatomic variation within the portal system. The complicated development of the vena cava, its close relationship with the development of the vitelline vein and the abnormal development of these vessels during this stage may explain the occurrence of this rare congenital extrahepatic portosystemic anastomosis.[4,5]

Classification

Congenital portosystemic shunts are classified into intrahepatic and extrahepatic types in accordance with the presence of connection between branches of the portal vein, after its division, and the hepatic veins or IVC. In CEPS, the anastomosis between the portomesenteric vasculature and systemic veins is observed before the division of the portal vein. The drainage vessel can vary. The most common drainage site is IVC (portocaval shunt), but it can drain in the renal vein, iliac vein, azygous vein, or right atrium.[6]

Morgan and Superina[7] classified CEPS into two types [Figure 4]. In Type 1 CEPS, there is complete diversion of portal blood to systemic circulation with absent intrahepatic portal branches. Type 1 CEPS are further classified into those, in which splenic vein and superior mesenteric vein drain separately into systemic vein (Type 1a) and those in which superior mesenteric and splenic vein form a confluence and drain into systemic vein (Type 1b). Type 2 CEPS intrahepatic portal vein is intact, and there is diversion of blood from portal venous circulation to systemic circulation by some communication. The knowledge of the presence of intrahepatic shunt and portal venous architecture is important to plan for therapeutic intervention needed.

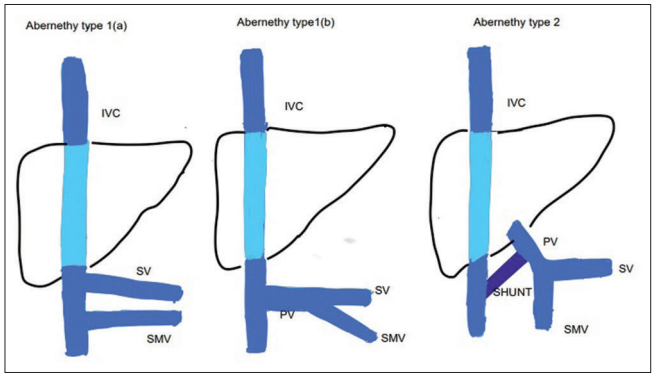

Figure 4: Types of Abernethy malformation. Type 1a – splenic vein and superior mesenteric vein drain separately into inferior vena cava. Type 1b – splenic vein and superior mesenteric vein form a portal vein that drains into inferior vena cava. Type 2 – Shunt between the portal vein and inferior vena cava

Patients with Type 1 shunts have female preponderance and are typically young, have high incidence of associated congenital anomalies. Type 2 shunts have no gender predilection.[3,6]

In addition to anatomic classification, Kobayashi *et al.*[8] proposed a clinical classification in accordance with drainage vessel of the portosystemic shunt. Portal blood flow is classified as Type A (if draining into IVC), Type B (if draining into renal vein), and Type C (if draining into iliac vein via inferior mesenteric vein). Presence of cardiac anomalies is more common with Type A patients, and Type C patients are associated more commonly with lower gastrointestinal bleeding.

In a recent classification put forward by Kanazawa *et al.*,[9] based on visualization of the architecture of the intrahepatic portal system during angiography using shunt occlusion test. They demonstrated that every case diagnosed as CEPS Type 1 showed visible intrahepatic portal system when shunt occlusion test done. They classified CEPS into three types depending on the severity of hypoplasia of the intrahepatic portal system: mild type, moderate type, and severe type and established correlation with histopathological findings. This information has important therapeutic implications whether portal vasculature will accept portal blood flow after shunt occlusion.

Diagnostic evaluation

The major challenge is to suspect and subsequently diagnose this rare malformation as highlighted by all our cases in this series who presented to a pediatric cardiac care unit with both cardiac and pulmonary manifestations. The first case presented with desaturation and respiratory distress and had significant cardiac abnormalities to explain desaturation. Persistence of low oxygenation despite successful surgical correction led to suspicion of pulmonary arteriovenous malformation in view of left isomerism and interrupted IVC and led to diagnosis of Abernethy malformation. Similarly, in the second case, persistence of PAH and transaminitis despite closure of patent ductus arteriosus led to the further evaluation and subsequent diagnosis. The third case presented with cyanosis as the primary manifestation and diagnosis of diffuse pulmonary arteriovenous malformation was investigated further and the fourth case underwent a patent ductus arteriosus device closure in early childhood but presented few years later with unexplained cyanosis. Case 5 highlights the dilemma faced by us, as to the etiology of pulmonary artery hypertension in this newborn. Clinically, there was no obvious cause for primary pulmonary hypertension of newborn. In view of association of Abernethy malformation with PAH, sildenafil was started empirically.

Diagnostic evaluation usually involves the use of multiple imaging modalities. The initial suspicion always arises from an abnormal scan of the abdomen but requires a high index of suspicion on part of the person performing the scan. Since it is not a routine investigation in evaluation of patients with congenital heart disease, it can be completely missed as highlighted by two of our cases who underwent device closure for patent ductus arteriosus, and in other case, routine preoperative ultrasound also missed because it was not suspected. Other investigations such as CT and magnetic resonance imaging (MRI) are helpful in the diagnosis. CT has the advantage of clearly delineating portal anatomy and type of shunt and thus help in deciding the therapeutic management. It also evaluates associated anomalies, particularly in patients with congenital heart disease who require evaluation of pulmonary vasculature and lungs in patients with suspected hepatopulmonary syndrome.[9]

MRI has the ability to get all the information with the advantage of avoiding ionizing radiation but has low spatial resolution than CT and thus may not be able to find small intrahepatic portal venous radicles in patients with Type 2 shunts.[9]

Liver biopsy is necessary in patients with Type 1 shunts because it may reveal small portal vasculature within the portal triads, a finding may not be seen by imaging tests.[10]

Treatment

Currently, there are no formal indications for the timing of treatment. In the presence of complications such as hepatopulmonary shunts and pulmonary hypertension even mild are usually considered absolute indication for treatment.[11,12]

Alonso-Gamarra *et al.*[11] suggested a diagnostic algorithm according to which asymptomatic/mild metabolic abnormality should be followed up with ultrasonography and biochemistry. Symptomatic patients or if shunt ratio is >60% decision is based on type of shunt. Type 1 shunt requires liver transplantation. Type 2 shunts will require closure by either endovascular or surgical means. The capacity of the portal system to accommodate increased flow is prerequisite for endovascular closure, so the presence of the intrahepatic portal venous system has to be documented clearly by either imaging or liver biopsy. Endovascular treatment can be either using detachable coil,[13,14] vascular plug,[14-16] patent ductus arteriosus device,[17] or using aortic endograft.[18] It can be either single-stage or using multiple-stage occlusion technique.[19]

In our series, one patient with Type 1 shunt was advised liver transplant and of the other two with Type 2 shunt underwent endovascular closure, the patient 2 with large shunt between IVC and portal vein underwent closure using aortic endograft reinforced with AndraMed stent, the patient 3 with long communication between the splenic vein and left renal vein was closed using a Amplatzer vascular plug. Limited series of CEPS which have been reported in the literature have been summarized in Table 2.

Table 2: Outcomes of various studies after surgical or endovascular treatment, in Abernethy malformation

Study	Sharma *et al.*, 2018, India[17]	Jain *et al.*, 2018, India[20]	Guneyli *et al.*, 2012, Turkey[16]	Franchi-Abella *et al.*, 2010, France[12]
Number of patients	*n*=5 Male (3), Female (2)	*n*=5 Male (4), female (I)	*n*=I Male	*n*=II Male (S), female (6)
Presentation	PH, HPS	HPS, liver mass	Psychiatric manifestations Hyperammonemia	PH, HPS, HCC, psychomotor retardation
Intervention	2 device closure (Lifetech PDA occluder 16/18 mm and AVP II 22 mm) 1 surgical closure 1 attempted but failed so advised surgical ligation, 1 under evaluation	All underwent surgical ligation of shunt	Device closure AVP II 22 mm	Primary surgical closure=4 Surgical banding - surgical closure=5 Surgical banding - device closure (AVP II)=1
Decision for intervention	Patient symptoms	Shunt occlusion done I. To measure portal pressure before ligation 2. Visualize intrahepatic portal venules	Shunt fraction (shunt fraction was calculated, was >82%, so risk of progression to HE)	Shunt occlusion done I. To measure port al pressure before ligation 2. Visualize intrahepatic portal venules
Follow -up	All treated patients were asymptomatic on follow-up	All improved on follow-up, but gradually	1-year follow-up, complete resolution of symptoms	On follow-up, 1 patient died due to HCC, 1 patient developed portal thrombosis after surgical banding Most patients with PAH had mPAP in range of 30-35 and had to be treated with pulmonary vasodilator therapy

HCC: Hepatocellular carcinoma, HE: Hepatic encephalopathy, HPS: Hepatopulmonary syndrome, PH: Pulmonary hypertension, mPAP: Mean pulmonary artery pressure, PDA: Patent ductus arteriosus, AVP: Amplatzer vascular plug

Another important aspect to consider prior to shunt closure is assessment of portal venous pressure. Franchi-Abella *et al.*[12] proposed a cutoff of 32 mmHg of portal venous pressure postshunt occlusion. In both our cases who underwent endovascular closure portal venous pressure was 12 mmHg and 14 mmHg postshunt occlusion testing.

CONCLUSION

Abernethy malformation or CEPS is a rare vascular malformation, which in the setting of associated congenital heart disease, can mask the clinical manifestation of the malformation as happened in two of our cases first case, persistence of PAH could have been explained by late closure of the patent ductus arteriosus, and in the second case, interruption of IVC could provide an alternate explanation for formation of pulmonary arteriovenous malformation. Hence, in patients presenting with pulmonary arteriovenous malformation, it is mandatory to rule out Abernethy malformation. A very high index of suspicion is necessary as highlighted by our cases with persistent PAH in order not to miss this rare anomaly. In majority of patients if diagnosed early are amenable to treatment to avoid the development of complications. Preprocedure assessment of portal venous system is must with imaging techniques and if required liver biopsy. Although therapeutic options are limited for Type 1 shunts who will require liver transplantation, certain selected patients are amenable to endovascular treatment. While planning endovascular treatment, measurement of portal venous pressure prior and after test occlusion of shunt is mandatory and also need to assess the location of renal vein/hepatic vein in relation to shunt. CT helps in planning and depending on the anatomy choice of vascular plug or stent graft can be decided.

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Conflicts of interest

There are no conflicts of interest.

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