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ABDOMINAL IMAGING

REVIEW

Abernethy malformation: A comprehensive review

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ABSTRACT

Abernethy malformation is a rare condition in which portomesenteric blood bypasses the liver and drains into a systemic vein through a partial or complete shunt. It is categorized into 2 types on the basis of the shunt pattern between the portal vein and a systemic vein. Abernethy malformation is associated with multiple congenital anomalies and acquired complications. A detailed understanding of the anatomy and embryology is a prerequisite to interpret imaging findings. Computed tomography and magnetic resonance angiography can delineate the shunt anatomy and evaluate concomitant malformations. It is essential to differentiate Abernethy malformation from intrahepatic portosystemic shunts and acquired extrahepatic portosystemic shunts. Mild metabolic abnormalities are treated with dietary modifications and medical therapy. Definitive treatment is done in symptomatic patients. Generally, type I Abernethy patients undergo liver transplantation, and type II undergo shunt occlusion by surgery or transcatheter coiling.

Abernethy malformation, also called congenital extrahepatic portosystemic shunt, is a rare condition in which the portosystemic blood drains into systemic circulation, bypassing the liver. John Abernethy first reported it in 1793.[1] It is a very rare anomaly with unknown prevalence; however, there is an increase in the number of cases in the last decade due to improved imaging techniques and awareness. It is categorized into 2 types based on the status of intrahepatic portal venous branches. Abernethy malformation is associated with multiple congenital anomalies and childhood complications.[2] The decision about the timing and mode of intervention is based on its types and associated abnormalities. The various available diagnostic modalities include ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and conventional angiography. In this article, we review the anatomy, embryology, classification, clinical findings, imaging appearances, differential diagnosis, and concomitant abnormalities of this rare anomaly. We also discuss the common interpretive pitfalls and various therapeutic options.

Anatomy and embryology

The portal vein (PV) is formed by the union of the splenic vein (SV) and the superior mesenteric vein (SMV). It measures approximately 9-12 mm in diameter and 8 cm in length in adults. It drains the splanchnic circulation and divides into 2 intrahepatic branches named right portal vein (RPV) and left portal vein (LPV). The RPV further divides into right anterior portal vein (RAPV) and right posterior portal vein (RPPV). The RAPV supplies segments V and VIII, while the RPPV supplies segments VI and VII. The LPV perfuses segments II, III, and IV. Segment I (caudate lobe) is supplied by the branches of both RPV and LPV. These segmental branches undergo a series of ramifications, ultimately leading to small venules which drain into hepatic sinusoids.[3]

The embryology of the portovenous system is a very complex process. The development of the portovenous system occurs in close relation with the umbilical venous system. There are 2 vitelline veins, which arise from the yolk sac, travel along the third part of the duodenum, and drain into primitive hepatic sinusoids. These vitelline veins are connected by 3 transverse communicating channels located at the level of transverse hepatic fissure (cranial most), dorsal (middle), and ventral (caudal most) to the duodenum. During the process of embryonic

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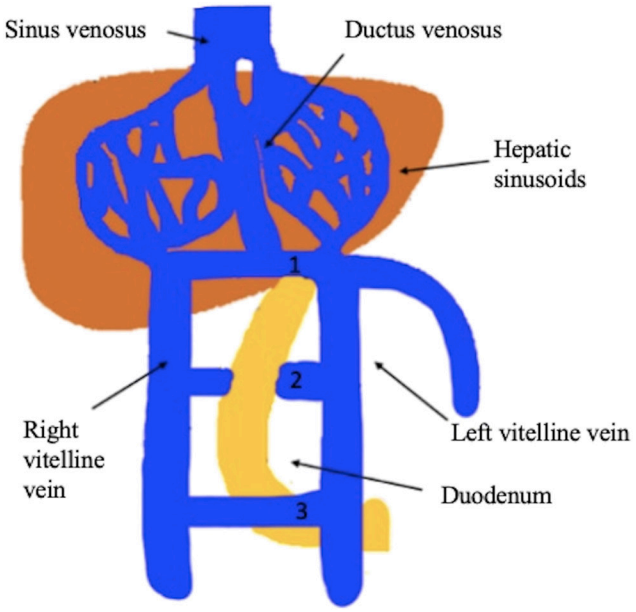


Figure 1. Embryologic development of the portal vein (PV). The ultimate PV system originates from selective involution of the anastomotic network between the right and left vitelline veins, which are located around the duodenum. 1, superior anastomotic communication; 2, middle anastomotic communication; 3, inferior anastomotic communication.

development, there occurs complex intricate involution of these transverse communicating channels resulting in the fully developed adult portal system (Figure 1).

If the vitelline veins fail to establish anastomosis with hepatic sinusoids, type I Abernethy malformation results. The persistent right vitelline vein results in an abnormal shunt between the PV and the retro hepatic inferior vena cava (IVC). Similarly, the persistence of the left vitelline vein leads to an abnormal shunt between the PV and the suprahepatic IVC or right atrium.

Main points

- Type I Abernethy is characterized by atretic intrahepatic portal venous branches and complete extrahepatic shunting of portal blood into a systemic vein, while type II is characterized by hypoplastic intrahepatic portal venous branches with partial extrahepatic shunting of portal blood into a systemic vein.
- Abernethy malformation is associated with multiple congenital malformations and acquired complications.
- It is essential to differentiate Abernethy malformation from congenital intrahepatic and acquired extrahepatic shunts.
- Multipurpose catheters should be considered when typical catheters fail to identify the RAV or when cranially oriented RAVs are identified but cannot be sampled with typical catheters.

There is another network of cardinal veins in the proximity of the PV which is the precursor of IVC. In the embryonic stage, there are multiple anastomotic channels between vitelline and sub-cardinal veins. These anastomotic channels undergo involution with fetal development, and any abnormal persistence of these channels results in type II Abernethy malformation.[4,5]

Classification

The most commonly used clinical classification was proposed by Morgan and Superina. It divides the Abernethy malformation into 2 types: type I and type II. Type I refers to the total aplasia of intrahepatic portal venous branches with complete extrahepatic shunting of portovenous blood into systemic veins. Type I is further divided into type Ia (SMV and SV drain separately into a systemic vein) and type Ib (SMV and SV unite to form a common vein before draining into a systemic vein). Type II Abernethy refers to hypoplastic intrahepatic portal venous branches with partial extrahepatic shunting of portovenous blood into a systemic vein (Figure 2).[1,6]

Lautz et al.[7] further classified type II into IIa (right or left PV drains into a systemic vein) and IIb (main PV drains into a systemic vein). Kobayashi et al.[8] divided Abernethy malformation into 3 types based on the site of the shunt, which are type A (the PV drains into the IVC), type B (the PV drains into the renal vein), and type C (the PV drains into the iliac vein). Another classification was proposed by Kanazawa et al.[9] based on the degree of intrahepatic portal hypoplasia on venography. He classified Abernethy malformation into 3 types: (1) mild type,

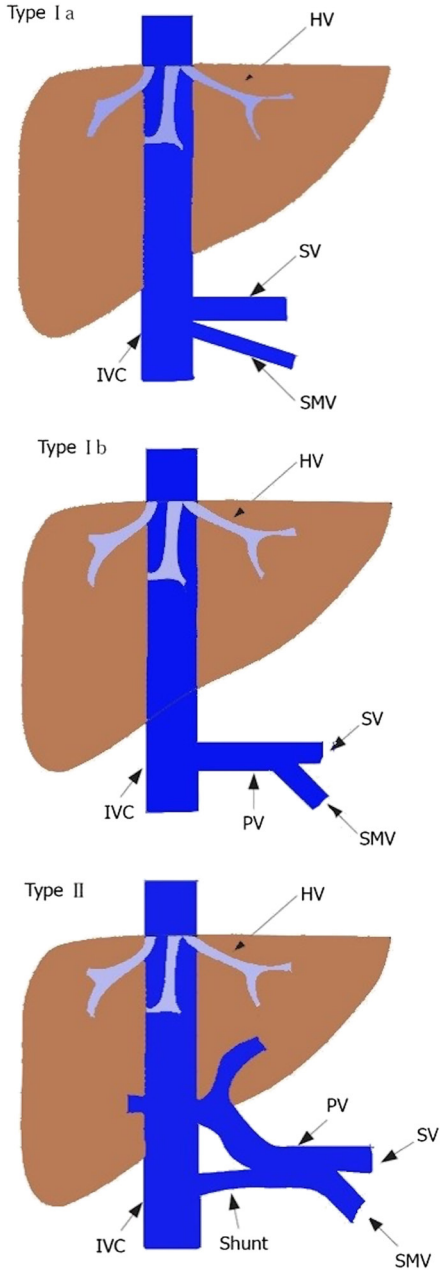


Figure 2. Different types of Abernethy malformation. Type 1 represents end-to-side shunting of PV into IVC bypassing the liver. It is further classified into type 1a (the SV and SMV drain separately) and 1b (both veins drain together after forming a common trunk). Intrahepatic portal venous branches are absent. Type 2 represents side-to-side anastomosis of PV and systemic vein. Intrahepatic portal venous branches are present but hypoplastic. PV, portal vein; SV, splenic vein; SMV, superior mesenteric vein; HV, hepatic veins; IVC, inferior vena cava.

Table. Congenital anomalies and childhood complications related with Abernethy malformation
Cardiovascular system
Atrial or ventricular or atrioventricular septal defect
Patent foramen ovale or patent ductus arteriosus
Transposition of the great arteries
Tetralogy of Fallot
Coarctation of the aorta
Dextrocardia
Hepatobiliary system
Biliary atresia
Congenital choledochal cyst
Caroli's disease
Congenital hepatic fibrosis
Intrahepatic gallbladder
Focal nodular hyperplasia
Hepatic adenoma
Nodular regenerative hyperplasia
Hemangioma
Hepatoblastoma
Hepatocellular carcinoma
Gastrointestinal system
Intestinal malrotation
Esophageal atresia
Anal atresia
Tracheoesophageal fistula
Bronchopulmonary system
Bronchomalacia
Laryngomalacia
Bronchial stenosis
Tracheal diverticulum
Lobar pulmonary sequestration
Vascular system
Agenesis or double IVC
Azygos/hemiazygos continuation of the inferior vena cava
Double aortic arch
Pulmonary artery stenosis
Urogenital system
Vesicourethral reflux
Renal agenesis
Hydronephrosis
Varicocele
Crossed fused ectopia
Skeletal system
Scoliosis
Sacral anomalies
Hip dysplasia
Hemivertebra
Facial dysmorphism
Metabolic disorders
Hyperammonemia
Hypergalactosemia
Hypothyroxinemia
Shunt complications
Portopulmonary hypertension
Portopulmonary syndrome
Portosystemic encephalopathy
Miscellaneous
Polysplenia/asplenia
Situs inversus/heterotaxia

defined as well-visualized intrahepatic portal radicles showing uniform distribution; (2) moderate type, defined as moderate visualization of intrahepatic portal radicles; and (3) severe type, defined as very poor or no visualization of intrahepatic portal radicles. The purpose of these classifications is to describe the shunts, thereby guiding management decisions. Sometimes, it is difficult to distinguish between type 1 and type 2 on imaging, and a liver biopsy serves as a problem-solving tool.

Concomitant congenital and acquired abnormalities

Multiple congenital anomalies are seen in association with Abernethy malformation. Cardiac anomalies are the most common and include atrial septal defect (ASD), ventricular septal defect, patent foramen ovale, patent ductus arteriosus, and tetralogy of Fallot. The association of cardiac anomalies with Abernethy malformation suggests either a common damage to heart and portal structures early in the embryonic life, or an adaptive response of the heart to overcome the hyperdynamic effects of the portosystemic shunt. Common hepatobiliary anomalies include congenital choledochal cyst, biliary atresia, congenital hepatic fibrosis, and intrahepatic gall bladder.[10,11]

Patients with Abernethy malformation show myriad hepatic pathologies ranging from fatty liver to hepatic malignancies. The altered hemodynamics is the key contributor to these changes. It has been seen that absent or decreased portal venous supply leads to hypertrophy of the hepatic artery and upregulation of arterial flow. The increased arterial blood supply brings more hepatic growth factors which predisposes to the development of multiple benign and malignant hepatic lesions. Studies have shown increased incidence of hepatic adenoma, hepatoblastoma, focal nodular hyperplasia (FNH), and hepatocellular carcinoma (HCC) in Abernethy patients.[12] Besides these hemodynamic changes, multiple mutations in the beta-catenin pathway are also known to contribute to hepatic malignancies.[13]

Patients with Abernethy malformation are also prone to primary pulmonary hypertension (PPH) and portopulmonary syndrome (PPS). It has been postulated that PPH develops due to repetitive pulmonary embolism from mesenteric circulation,

and PPS results from shunting of vasodilatory mediators from the PV to a systemic vein.[14] The common congenital and acquired abnormalities are summarized in the Table.

Clinical features

Abernethy malformation can present with a broad spectrum of clinical manifestations. Usually, it is diagnosed during childhood in the presence of hypergalactosemia, cholestasis, failure to thrive, psychomotor delay, or other congenital defects. Sometimes, it is incidentally found during the workup of abdominal pain, abnormal liver function tests, portopulmonary syndrome, portopulmonary hypertension, or portosystemic encephalopathy. Currently, prenatal diagnosis of Abernethy malformation is increasingly being reported. The common findings on antenatal ultrasound include abnormal portosystemic communication or an enlarged umbilical vein.[15,16] Rarely, the patient may present with lower gastrointestinal bleeding or hematuria, due to rectal varices secondary to the portosystemic shunt draining portal blood into the iliac vein via an inferior mesenteric vein.[17,18] To date, approximately 323 cases have been reported (182 type I and 141 type II).[11,19,20] There is no clear sex preponderance. The age of presentation is also variable. Although up to 70% of the cases are diagnosed before 18 years or less, the age of presentation can vary from prenatal diagnosis to as old as 61 years for type I and 76 years for type II. Congenital anomalies are more commonly seen in type I as compared to type II. There is no geographical predilection and cases have been reported worldwide. The resolution of symptoms has been seen after liver transplantation or endovascular intervention at 1-year follow up, but no data are available on the prognosis and long-term outcomes of various interventions.

Diagnostic modalities for evaluation of Abernethy malformation

Ultrasonography is the initial investigation as it is non-invasive and radiation-free. The classical US findings include failure to visualize intrahepatic portal venous branches and hypertrophy of the hepatic artery. US rules out the acquired causes of non-visualized PV, like PV thrombosis. It also displays the focal hepatic lesions, which are frequently associated with Abernethy malformation. Contrast-enhanced US (CEUS) improves diagnostic accuracy in evaluating focal hepatic lesions. The limitations of US include operator dependency and failure to delineate the extrahepatic shunts accurately.[21]

CT and MRI are of great importance for diagnosis and pre-operative planning. Both CT and magnetic resonance angiography can accurately detect the shunt anatomy

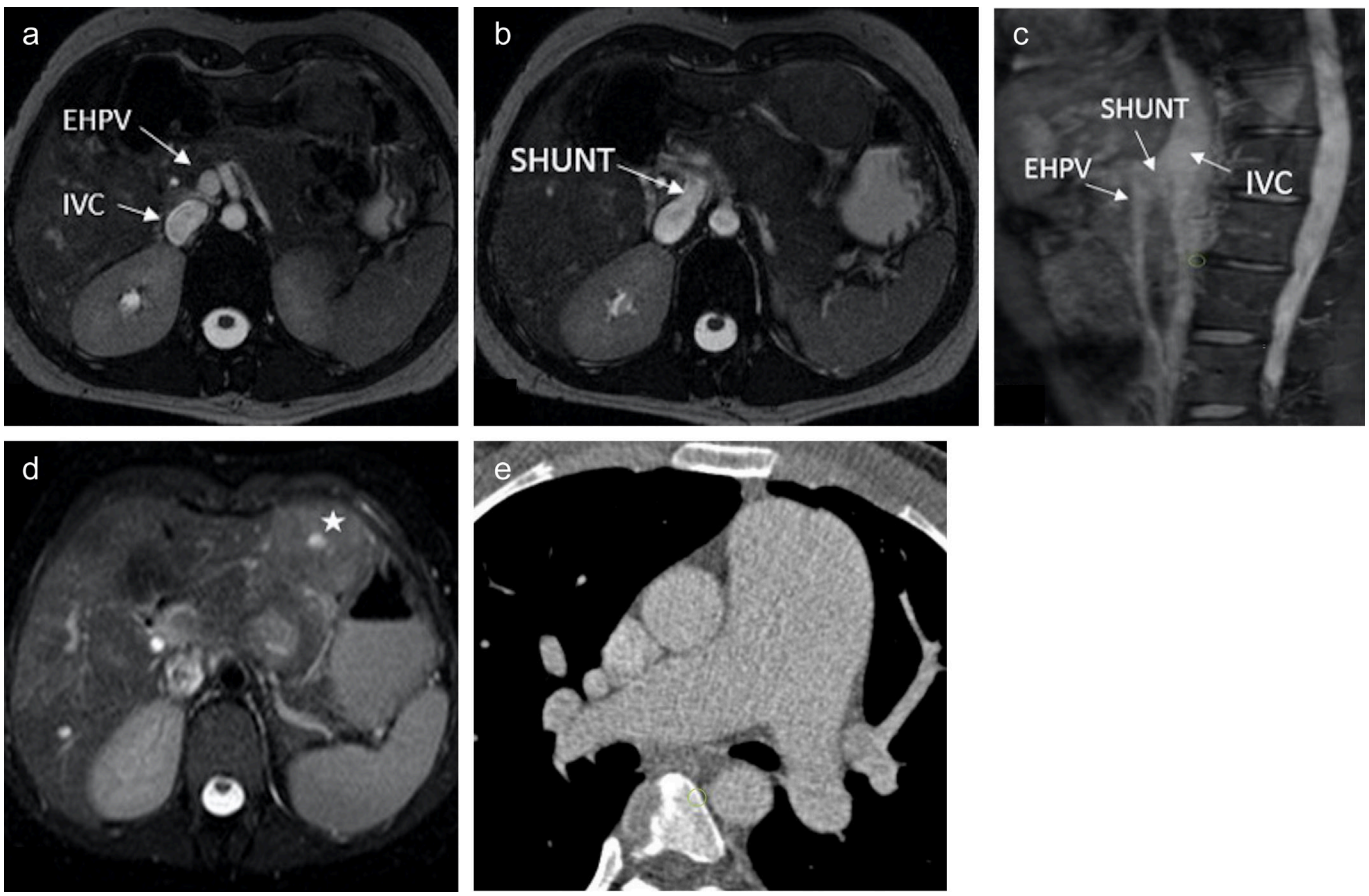


Figure 3. a-e. Type 1b Abernethy malformation with hepatic focal nodular hyperplasia and pulmonary hypertension. Consecutive axial balanced turbo field echo magnetic resonance (MR) images from inferior **(a)** to superior **(b)** show the EHPV draining into the retro hepatic IVC. No intrahepatic PV branches are seen. Sagittal MR angiography image **(c)** shows a shunt directly connecting EHPV to retro hepatic IVC. T2-weighted axial MR image **(d)** shows multiple hyperintense lesions (*star*), with hyperintense central scar in both lobes consistent with focal nodular hyperplasia. Axial contrast-enhanced computed tomography (CECT) image **(e)** of the same patient shows dilated pulmonary infundibulum (34 mm in diameter) suggesting pulmonary artery hypertension. EHPV, extrahepatic portal vein; IVC, inferior vena cava.

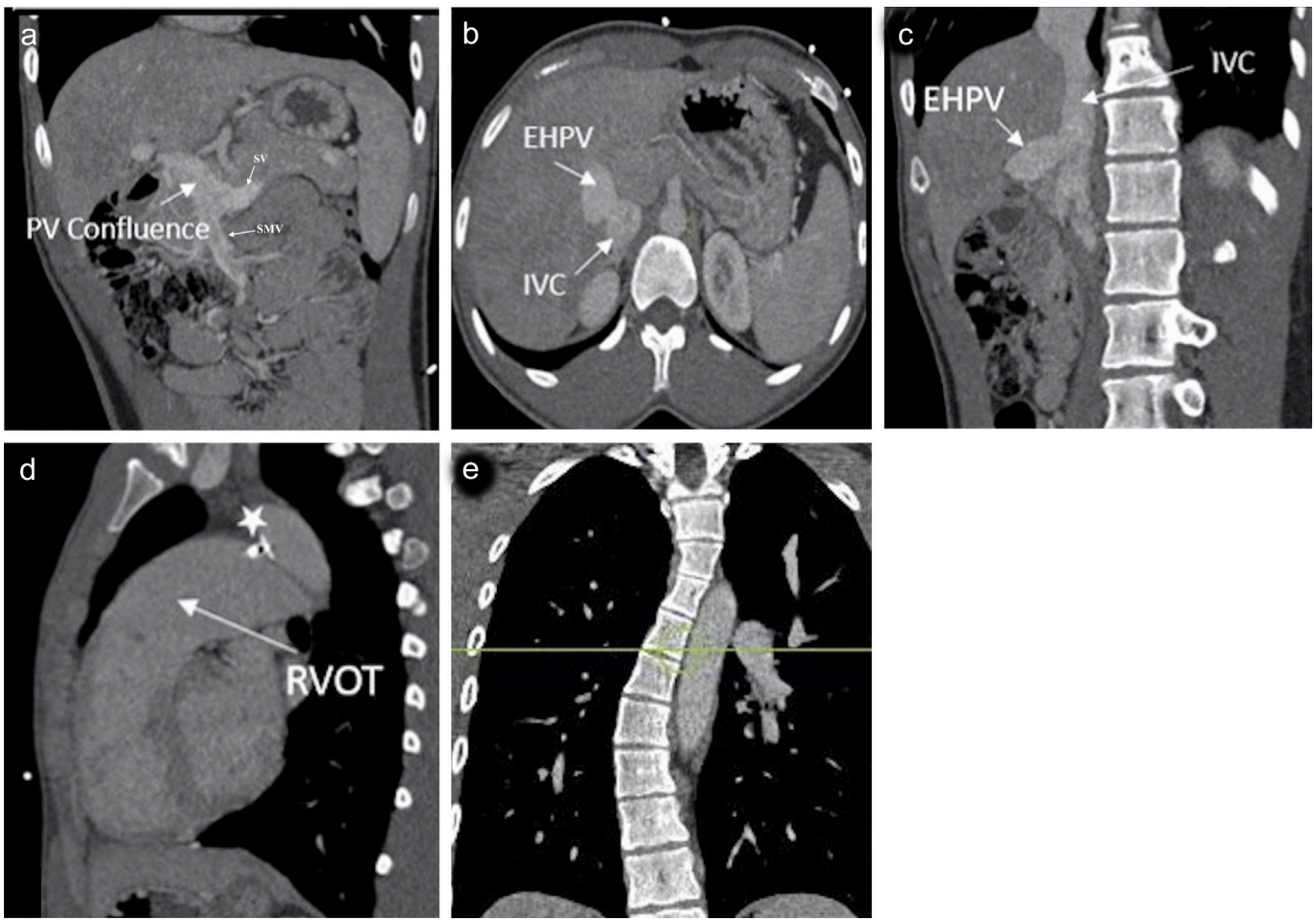
and characterize various coexisting hepatic and extrahepatic lesions. The advantages of CT include superior spatial resolution and fast scanning. The entire scan can be completed in a single breath-hold. Dose reduction algorithms can significantly reduce the patient dose. The disadvantages of CT include exposure to iodinated contrast and ionizing radiation.[22] MRI, due to high soft tissue contrast, is an excellent modality for evaluating various hepatic and extrahepatic abnormalities associated with Abernethy malformation. There is no exposure to ionizing radiations. However, MRI is limited by higher cost, availability, and the need for sedation or anesthesia in children and patients with MRI non-compatible devices.

Angiography is used when the results of other techniques are inconclusive. Angiography techniques include indirect mesenteric portovenography and percutaneous transhepatic portography. The advantages of angiography include the calculation of pressure gradients across shunts and the simultaneous therapeutic occlusion of selective shunts. Liver biopsy is used to differentiate between severe hypoplasia and total aplasia. Total aplasia is characterized by the absence of portal venules on histopathology. The biopsy can also characterize various hepatic lesions.[23]

Imaging features

The major role of imaging in Abernethy malformation is to demonstrate the extrahepatic portosystemic communication and to diagnose the type of shunt. It is essential in distinguishing between intra- and extrahepatic shunts, as it may have therapeutic implications. Type I Abernethy malformation is characterized by aplasia of intrahepatic portal venous branches and complete extrahepatic shunting of portal venous blood into a systemic vein. Imaging shows an end-to-side shunt between the PV and the IVC, which could be of 2 types: the SV and the SMV drain separately into IVC (type Ia), or both the SV and the SMV form a common trunk (PV), which drains into the IVC (type Ib) (Figures 3 and 4). Usually, the PV drains into retro-hepatic IVC anywhere between a point just inferior to the hepatic venous confluence and superior to the level of renal veins. However, there are case reports demonstrating PV drainage into the suprahepatic IVC and right atrium. Intrahepatic portal venous branches are absent in type I Abernethy malformation.[3,5,6]

Type II Abernethy malformation is characterized by hypoplastic intrahepatic portal venous branches with partial extrahepatic drainage of portal venous blood into a systemic vein. Imaging shows a side-to-side shunt between the PV, including its tributaries, and systemic veins. PV or its tributaries (SMV and SV) may drain into any systemic vein including IVC, left or right



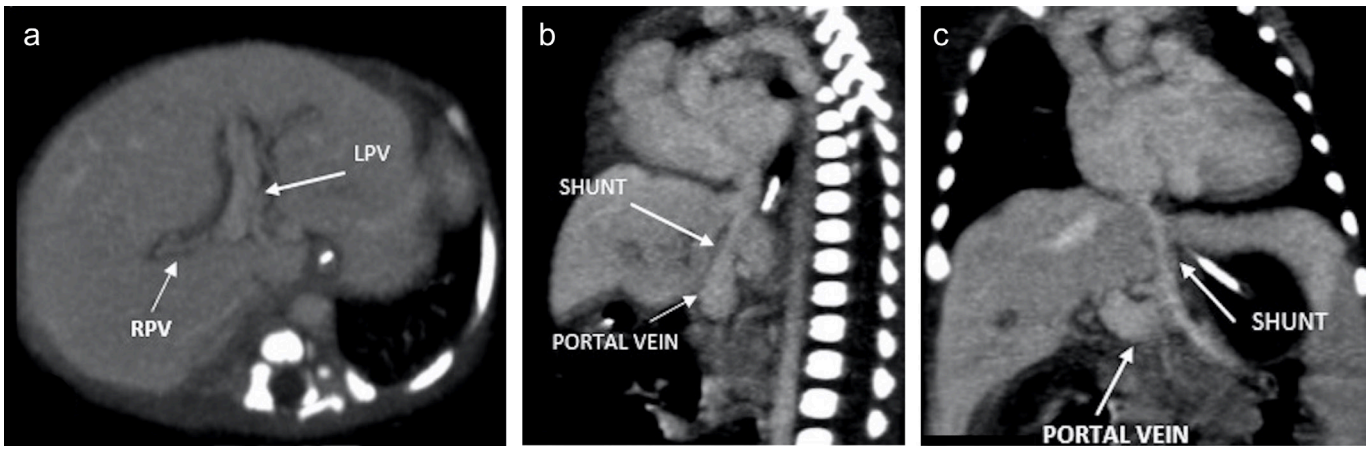


Figure 5. a-c. Type 2 Abernethy malformation. Axial CECT image **(a)** shows hypoplastic right portal venous branch and normal left branch. Sagittal CECT image **(b)** shows an extrahepatic shunt between PV and suprahepatic IVC. Coronal CECT image **(c)** shows extrahepatic shunt between PV and IVC. RPV, right branch of PV; LPV, left branch of PV.

renal vein, inferior mesenteric vein, and left or right internal iliac veins (Figure 5). In case of interrupted IVC with azygos or hemiazygos continuation (i.e., left isomerism), the PV or its tributaries drain into the azygos or hemiazygos veins. Intrahepatic branches are seen in type II Abernethy malformation, but they are hypoplastic[3,5,6] (Figure 6).

Once the portosystemic communication is demonstrated, it is equally important to diagnose the other congenital abnormalities associated with Abernethy malformation. Hepatic lesions are the most common abdominal findings and include hepatic adenoma, FNH, and HCC. These lesions show typical and diagnostic appearance on MRI. Therefore, whenever seen on US, a triple-phase MRI with hepatobiliary-specific MRI contrast agents should be done. Similarly, an echocardiography should be done in every patient to rule out cardiac anomalies.

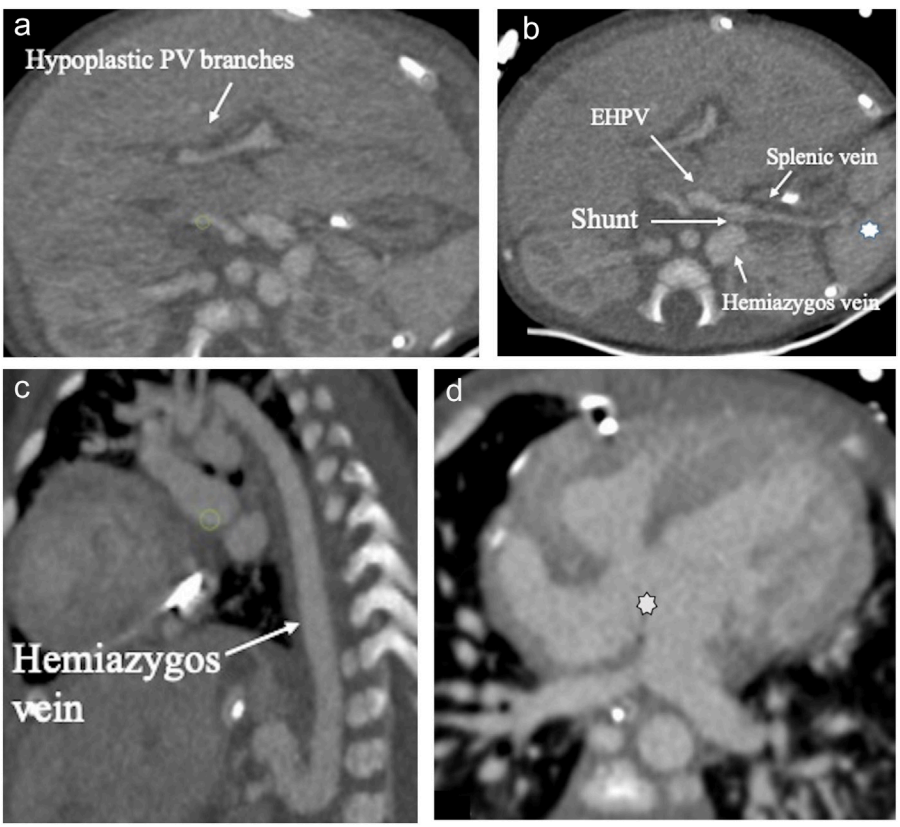


Figure 6. a-d. Type 2 Abernethy malformation with left-sided isomerism and atrial septal defect. Axial CECT image **(a)** shows hypoplastic intrahepatic portal venous branches. Axial CECT image **(b)** shows shunt between splenic vein and hemiazygos vein. Polysplenia (*asterisk*) is also noted. Sagittal CECT image **(c)** shows intrathoracic course of hemiazygos vein. Axial CECT image **(d)** shows atrial septal defect (*asterisk*). PV, portal vein; EHPV, extrahepatic portal vein.

Differentials and common pitfalls

The common differential diagnosis of Abernethy malformation is acquired extrahepatic portosystemic shunt, usually seen in patients with cirrhosis. PV thrombosis is another close mimicker. In chronic thrombosis, the PV is replaced by a thin fibrotic cord, which may simulate type II Abernethy malformation. Both cirrhosis and PV thrombosis show secondary signs of portal hypertension like splenomegaly, ascites, and collateral channels. These signs are absent in Abernethy malformation and help in distinguishing these entities (Figure 7). Furthermore, Abernethy malformations must be distinguished from the intrahepatic portosystemic shunt. Intrahepatic portosystemic shunts can be congenital or acquired (trauma, portal hypertension). Park et al.[24] have described 4 types of intrahepatic portosystemic shunts, and the most common type is a single vessel connecting the RPV to the IVC. Ductus venosus is another congenital intrahepatic portosystemic shunt. It generally closes between 2 and 18 days of life in term infants but may remain patent in some cases. The main difference between the intrahepatic and extrahepatic shunts is the site of origin. Extrahepatic shunts arise from the main PV, and intrahepatic shunts originate from the portal venous branches.

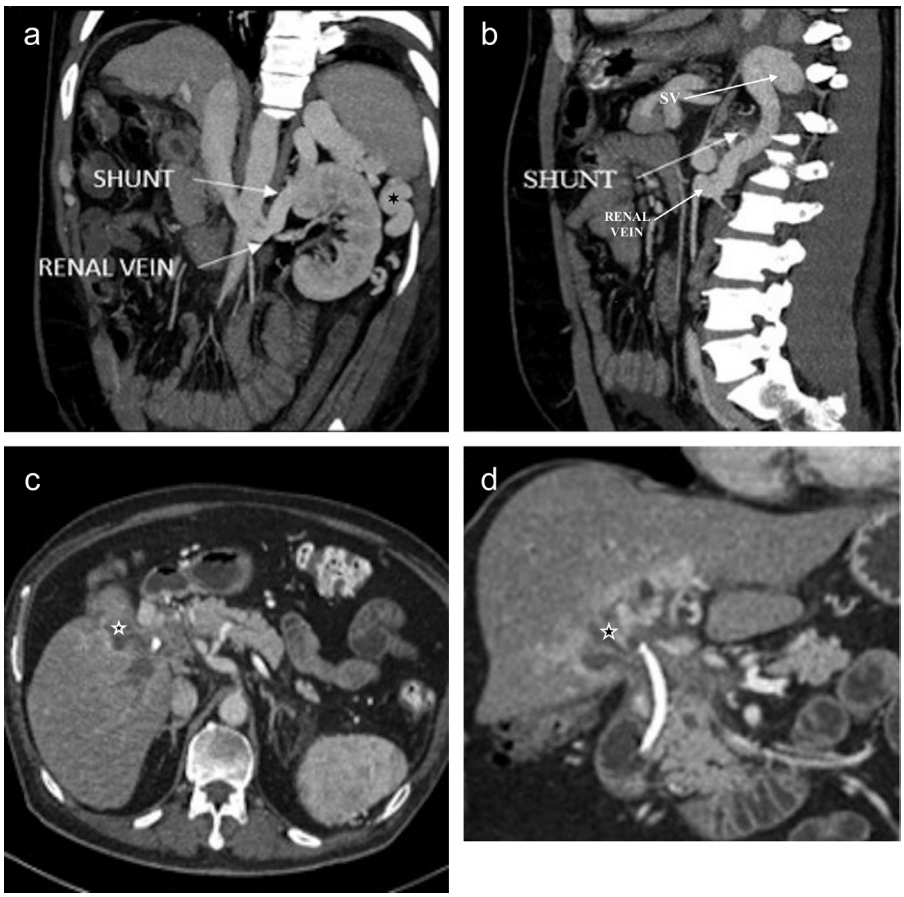


Figure 7. a-d. Acquired portosystemic shunt. Oblique coronal and sagittal maximum intensity projection CECT images **(a, b)** show multiple collaterals (*asterisk*) at splenic hilum. A large portosystemic shunt is seen between splenic and left renal vein. Axial and coronal CECT images **(c, d)** show absence of the intrahepatic portal vein and collateral channels in the porta hepatis (*asterisk*); findings consistent with PV obstruction with secondary collateralization. The shunt seen between the splenic and left renal vein is an acquired shunt due to portal hypertension, and not a congenital portosystemic shunt. These collateral vessels and other secondary signs of portal hypertension, such as perisplenic collaterals, differentiate PV thrombosis from Abernethy malformation.

It is essential to distinguish Abernethy malformation from intrahepatic shunts and acquired extrahepatic shunts because they have different treatment strategies.

Treatment

Management of Abernethy malformation depends on its type, related complications, and the associated anomalies. There is no universal approach, and the treatment strategy is decided on a case to case basis. Generally, type I Abernethy patients need liver transplantation, whereas type II Abernethy can be treated by shunt closure.[25,26] All symptomatic patients must be treated. The treatment of asymptomatic patients is based on the shunt ratio, which is calculated by Doppler ultrasound. If the shunt fraction (ratio of shunt flow volume to total portal flow volume) is greater than 60%, the chances of hepatic encephalopathy are very high and hence treatment is advised. If the shunt fraction is less than 60%, follow-up is recommended.[27,28] The various indications for liver transplantation in Abernethy malformation include hepatic encephalopathy, liver tumor like hepatoblastoma or HCC, and associated biliary anomalies like biliary atresia.[29-32] Multiple authors have proposed different algorithms for shunt closure in type II Abernethy malformation. Kanazawa et al.[9] have recommended interventional closure of the portosystemic shunt based on portal venous pressure after the balloon occlusion test of the shunt. According to them, the balloon occlusion of shunt should be followed by measurement of portal venous pressure (PVP). If the PVP is <25 mmHg, a single-step occlusion of the shunt should be done; if PVP is >30 mmHg, the patient is a candidate for liver transplantation or a two-step closure of shunt. The two-step closure includes surgical banding of the shunt followed by shunt closure a few months later. If the PVP is between 25 and 30 mmHg, other factors are taken into account. Blanc et al.[33] have proposed another algorithm for shunt closure. According to them, single-step shunt closure should be performed if the PVP is less than 32 mmHg after the balloon occlusion of shunt. If the PVP is more than 32 mmHg, then an alternative 2-step procedure is recommended. Mild metabolic abnormalities are treated with dietary modifications and medical therapy. If any hepatic lesion is noted on follow-up, a dedicated triple-phase study should be done with hepatobiliary MRI-specific agents like gadobenate dimeglumine (Gd-BOPTA) a gadoxetic acid (Gd-EOB-DTPA). These hepatobiliary agents can characterize the lesion, notably FNH and HCC, avoiding unnecessary liver biopsies. The associated anomalies also affect the therapeutic approach. For example, in a patient with pulmonary hypertension with type II Abernethy malformation and ASD, the treatment strategy is decided on the basis of balloon occlusion tests. If the pulmonary pressure decreases after balloon occlusion of ASD, the ASD is repaired first, followed by occlusion of the portosystemic shunt later.

Conclusion

Abernethy malformation is a rare anomaly with multiple clinical associations. Most often, children present with dyspnea, encephalopathy, and abdominal complaints. The purpose of imaging is to identify and classify the shunt, with identification of accompanying anomalies. It is essential to distinguish this entity from intrahepatic shunts and acquired extrahepatic shunts. Careful monitoring is recommended if the patients are asymptomatic or have mild metabolic abnormalities. Any complication warrants appropriate therapeutic intervention. Radiologists must be familiar with imaging features of this rare anomaly for early diagnosis and therapeutic guidance, leading to better patient outcome.

Conflict of interest disclosure

The authors declared no conflicts of interest.

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