PICTORIAL REVIEW

Hepatic vascular shunts: embryology and imaging appearances

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ABSTRACT. The purpose of this pictorial review is to understand the embryological basis of the development of congenital hepatic vascular shunts and to review the multimodality imaging appearances of congenital and acquired hepatic vascular shunts. Hepatic vascular shunts are commonly seen in imaging. Familiarity with their characteristic appearances is important in order to accurately characterise these shunts and diagnose the underlying disorders.

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Intrahepatic vascular shunts are common; all intrahepatic vessels are associated with the formation of shunts. These include arteriportal (AP), portosystemic and arteriosystemic venous shunts. Rarely, systemic venous (*i.e.* hepatic vein to hepatic vein) shunts and portal-to-portal communications are seen [1]. The unique dual blood supply of the liver (approximately 75% portal venous and 25% arterial) means there is a compensatory relationship between the two sources. When vascular compromise occurs, it causes a macroscopic opening of the shunts between the hepatic arteries, the portal veins and the hepatic veins through physiological anastomoses, which become abnormally enlarged [1–3]. The purpose of this pictorial review is to illustrate the imaging appearances of hepatic vascular shunts on multidetector CT (MDCT) with sonographic and conventional angiographic correlation.

Embryology

In the fifth week of intrauterine life, there are three major paired veins in the abdomen: the vitelline or omphalomesenteric veins, the umbilical veins and the

cardinal veins. The vitelline veins form the hepatic sinusoids. The left vitelline vein subsequently involutes and the blood is diverted into the right vitelline vein, which enlarges and forms the hepatocardiac portion of the inferior vena cava (IVC). Derivatives of the vitelline veins also form the hepatic and portal vein. The entire right umbilical vein and a portion of the left umbilical vein also degenerate. The persistent portion of the left umbilical vein connects to the right vitelline vein via the ductus venosus (Figure 1). The hepatic artery arises from the aorta and follows the mesentery [4, 5].

In antenatal circulation, oxygenated blood from the placenta returns to the foetus via the umbilical vein and flows through the ductus venosus into the IVC (Figure 2a). At birth, there is functional closure of the umbilical arteries, veins and the ductus venosus. Following atrophy, these form the medial umbilical ligaments, the ligamentum teres and the ligamentum venosum, respectively. Post-natal anatomy consists of inflow into the liver through the hepatic artery and portal vein, with the outflow through the hepatic veins (Figure 2b) [5].

There is potential for intrahepatic shunts to develop when the ductus venosus remains patent, from failure of right upper vitelline vein regression, from a varix of the portal vein connecting to the hepatic/systemic venous circulation, from focal persistence of the most superior aspect of the vitelline veins or from arteriovenous malformations or neoplasms [6, 7].

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Figure 1. Schematic images showing development of the hepatic venous system, which starts to develop in the fifth week of intrauterine life. Vitelline veins form the hepatic sinusoids. Left umbilical vein connects to the right vitelline vein via the ductus venosus. Anastomotic network around the duodenum forms the portal vein. LCV, left cardinal vein; RCV, right cardinal vein; RUV, right umbilical vein; LUV, left umbilical vein; SC, superior communication; MC, middle communication; IC, inferior communication.

Figure 2. Hepatic venous system changes at birth. (a) Schematic diagram showing antenatal circulation with direct communication between the umbilical vein and inferior vena cava (IVC) through the ductus venosus. (b) Schematic diagram showing post-natal circulation with inflow into liver through the hepatic artery and portal vein, and outflow through the hepatic veins.

Figure 3. A 23-year-old male with Osler-Weber-Rendu syndrome. (a) Axial arterial-phase CT image showing multiple arteriportal shunts in both lobes of liver (arrows). (b) Selective coeliac arteriogram showing multiple arteriportal shunts (arrows).

Figure 4. A 55-year-old female with known hepatocellular carcinoma. Selective hepatic arteriogram showing a hypervascular tumour in the right lobe (arrow) with arterioportal shunting. Note the early opacification of portal vein, which shows a "thread and streak" sign from tumour thrombus (double arrow).

Classification

The hepatic vascular shunts can be divided into AP, portosystemic, arteriosystemic, systemic-venous, and portal-to-portal shunts.

Imaging features of hepatic vascular shunts

Arterioportal shunts

AP shunts are seen in congenital vascular malformations, including those identified in patients with hereditary haemorrhagic telangiectasia (HHT), as well as in benign and malignant hepatic neoplasms, cirrhosis, tumour thrombus and trauma. On colour Doppler imaging, these shunts display high-velocity, low-resistance arterial waveforms with arterialised portal venous flow. On CT and MRI, early enhancement of the portal vein is often seen in the segmental portal venous branch close to the shunt, as well as within the main portal vein. Evidence of reversed flow in the portal vein may be inferred by non-opacification of the splenic vein and the superior mesenteric vein on the same arterial phase. In cases of tumoural AP shunts, there is transient wedge-shaped enhancement peripheral to the tumour [3, 8].

HHT, or Osler-Weber-Rendu (OWR) syndrome, is an autosomal dominant disorder known to involve virtually every organ of the body, with hepatic involvement in 8–31% of cases. Hepatic manifestations include AP shunts (Figure 3), arteriosystemic shunts (most common type), portovenous shunts, telangiectases, large confluent vascular masses, perfusion abnormalities, fibrosis and cirrhosis [9, 10]. Patients with symptomatic liver disease (high-output liver failure or portal hypertension) and atypical cirrhosis ultimately require transplantation [10]. Among hepatic tumours, AP shunts are most commonly identified in patients with hepatocellular carcinoma (HCC). They are also commonly seen in cirrhosis, especially after a portocaval shunt operation [4, 8]. These shunts are typically wedge-shaped, with or without internal linear branching structures in the arterial phase, and are iso- to slightly hyperattenuating during the portal venous phase. However, a follow-up radiological study may be required to exclude the possibility of a coexistent tumour [11]. Tumour thrombi are seen most commonly in patients with HCC (Figure 4) and to a lesser extent in patients with cholangiocarcinoma or metastases, and also cause AP shunting. Trauma-related AP shunts may occur secondary to penetrating trauma, and can be caused by a liver biopsy (Figure 5) or by percutaneous biliary catheterisation. Radiological imaging-guided intervention, such as embolisation, is performed if there is rapid development of life-threatening portal hypertension or high-output cardiac failure. If embolisation fails, surgery is often necessary [2, 8].

Portosystemic venous shunts

Portosystemic venous (PV) shunts are relatively rare, and are seen between a portal venous branch and a hepatic vein or the IVC. Postulated congenital causes include persistence of a communication between the vitelline veins and the right horn of the sinus venosus or rupture of a portal vein aneurysm into a hepatic vein [4, 12, 13]. Park et al [13] have described four morphological types of portosystemic shunts. Type I shunt, the most common type, is a single large shunt connecting the right portal vein to the IVC (Figure 6). Type II shunt is either single or multiple, and communicates between the portal and hepatic veins in one hepatic segment (Figure 7). Type III shunt is a small connection between a peripheral portal and hepatic vein through an aneurismal varix (Figure 8). Type IV shunts are multiple communications between the peripheral portal and hepatic veins, and are seen diffusely in both lobes (Figure 9). A patent ductus venosus connecting the left portal vein to the IVC could be considered the fifth type of portosystemic shunt (Figure 10) [4]. Embolisation or surgery is performed if the shunt is symptomatic.

Acquired causes of PV shunts include portal hypertension with or without associated HCC, iatrogenic (from transjugular intrahepatic portosystemic shunt, TIPS; Figure 11), surgical portocaval shunt (Figure 12), surgical mesentericocaval shunt and trauma [14]. PV shunts appear as anechoic tubular cystic channels on greyscale sonography, with a pulsatile spectral Doppler waveform in the portal and splenic veins [4, 15]. CT and MRI can confirm the diagnosis [4].

Figure 5. A 65-year-old man with a large arterioportal shunt after percutaneous liver biopsy. (a) Axial colour Doppler image of the right lobe of liver at the site of cystic lesion seen on a greyscale sonogram (not shown), showing increased colour flow (arrow) at the site of percutaneous liver biopsy. (b) Axial spectral Doppler image of the right portal vein, showing arterialisation with hepatofugal flow due to large arterioportal shunt in the right lobe. (c) Anterior volume-rendered CT image confirming presence of a large arterioportal shunt (arrow) in the right lobe. (d) Selective right hepatic artery angiogram showing a large arterioportal shunt. Note the enlarged portal vein (double arrow) due to shunting into the right portal vein (arrow).

Figure 6. A 17-year-old male with fragile X syndrome and Park Type I portosystemic shunt. (a) Oblique sagittal greyscale sonogram showing a large shunt (arrowhead) between the right portal vein (arrow) and the inferior vena cava (IVC; double arrow). (b) Contrast-enhanced CT axial and (c) coronal images showing a large shunt (black arrow) between the right portal vein (arrow) and the IVC (double arrow).

(a)

(b)

Figure 7. A 76-year-old female with a history of alcohol abuse and Park Type II portosystemic shunt. (a) Axial contrast-enhanced CT image showing a small shunt (white arrow) involving portal venous branch. (b) Another image 5 mm superior to this level showing the shunt (black arrow) connecting to the right hepatic vein. Bilateral pleural effusions, left larger than right, are also seen.

(a)

(b)

Figure 8. A 50-year-old male scheduled for liver transplantation for end-stage liver disease secondary to hepatitis C with Park Type III portosystemic shunt. Contrast-enhanced CT (a) axial and (b) sagittal images showing a shunt (white arrows) connecting the main portal vein to the inferior vena cava (black arrows) via a small varix (arrowhead).

(a)

(b)

Figure 9. A 48-year-old male with cirrhosis and Park Type IV portosystemic shunts. Axial contrast-enhanced CT images showing small shunts (arrows) between the portal vein and hepatic veins in (a) the left lobe and (b) the right lobe. Small right pleural effusion is also present.

Figure 10. A 46-year-old female with a history of heavy alcohol use, transient ischaemic attacks, pulmonary arteriovenous malformations and a large portal vein to inferior vena cava (IVC) shunt (patent ductus venosus). Contrast-enhanced CT coronal image showing a large shunt (double arrow) originating from the bifurcation of the portal vein communicating with the IVC (arrow). Multiple hepatocellular carcinomas were also identified on the same CT examination (not shown).

Figure 11. A 48-year-old male with transjugular intrahepatic portosystemic shunt (TIPS) performed for cirrhosis and portal hypertension. Angiogram showing a patent TIPS shunt in the liver between the right hepatic vein and the right portal vein. Coil embolisation of the varices was performed. The proximal end of the stent extends into the right atrium in this patient, who was not a suitable candidate for liver transplantation. Such extension into the right atrium in a potential liver transplantation candidate may make subsequent liver transplantation difficult.

Figure 12. A 50-year-old male with a surgically created portocaval shunt for cirrhosis and portal hypertension. (a) Oblique coronal colour Doppler sonogram showing a shunt (arrow) seen connecting the inferior vena cava (IVC) to the main portal vein (PV). (b) Axial contrast-enhanced CT image showing the shunt (arrow). Multiple foci of hepatocellular carcinomas are also present.

Arteriosystemic venous shunts

Arteriosystemic venous (AV) shunts are communications between the hepatic artery and the hepatic veins. These can be seen in vascular malformations such as HHT, as well as in benign and malignant liver lesions including infantile haemangioendothelioma, haemangioma and HCC [1]. Infantile haemangioendothelioma (Figure 13) usually manifests clinically before 6 months

Figure 13. A 32-week-old foetus with a vascular liver mass. Post-natal resection revealed infantile haemangioendothelioma. Axial colour Doppler image of the foetal abdomen shows a heterogeneous mass in the foetal liver with increased vascularity from arteriosystemic venous shunts.

of age with high-output congestive failure, hepatomegaly, anaemia, thrombocytopenia, respiratory distress, haemorrhage or jaundice. Haemangiomas in the skin and associated organs (haemangiomatosis) are also commonly identified. When AV shunting is present, high-peak Doppler shifts and a low resistive index in the feeding arteries may be seen, as well as hyperpulsatility of the portal vein and a pulsatile pattern in the draining veins. There is also aortic tapering at the branching point of the coeliac artery. Asymptomatic patients are closely followed up without treatment. Standard treatment for infantile haemangioendothelioma is corticosteroids. If there is no clinical improvement, interferon treatment should be attempted. Embolisation or surgery is reserved for patients in whom congestive heart failure remains even after a reasonable trial of medical therapy. Chemotherapy and radiation therapy have also been used [4].

Systemic-to-systemic venous shunts

Systemic-to-systemic shunts are more often seen in patients with chronic hepatic venous congestion or Budd-Chiari syndrome. When Budd-Chiari syndrome occurs, there is obstruction of the hepatic venous outflow in the absence of right heart failure or constrictive pericarditis, and patients present with hepatomegaly, ascites and abdominal pain. Hepatic venous shunts (Figure 14) are a specific diagnostic criterion for the diagnosis. These are either intrahepatic, which communicate with systemic vessels via subcapsular collaterals, or from an occluded hepatic vein to a non-occluded hepatic vein or to the caudate lobe veins. Rarely, shunts are seen from the hepatic veins to the suprahepatic IVC

(a)

(b)

(c)

(d)

Figure 14. A 24-year-old female with Budd-Chiari syndrome. (a) Sagittal colour Doppler image showing narrowing in the inferior vena cava (IVC; arrows) with aliasing consistent with stenosis. (b) Oblique transverse colour Doppler image through the liver showing hepatic vein to hepatic vein collaterals (arrows). (c) Venogram through caudate lobe vein (white arrow) showing intrahepatic venovenous collaterals with occlusion (arrowhead) of the terminal right hepatic vein (black arrow). Also note truncation (double arrow) of the intrahepatic segment of the right hepatic vein, as evidenced by non-opacification of the side branches. (d) Right hepatic venogram showing a narrowed and truncated hepatic vein (black arrow) with intrahepatic venous collateral (white arrowhead) drainage through the caudate lobe (white arrow) and an accessory right hepatic vein (black arrowhead).

Figure 15. A 54-year-old male with cirrhosis, portal hypertension and main portal vein to umbilical vein shunt. Axial contrast-enhanced CT image showing a collateral from the main portal vein to the patent umbilical vein (arrow). Moderate ascites and small splenic infarcts are also seen.

close to the right atrium. Therapeutic interventions include thrombolytic therapy, angioplasty with or without stenting, TIPS, surgical shunting and liver transplantation [16]. Systemic-to-systemic venous shunts have also been identified in patients with HHT, and in HCC following tumour extension into the hepatic veins [1].

Portal-to-portal venous shunts

Intrahepatic portal-to-portal venous (PPV) shunts are rare, but may be seen in cirrhosis. Recognition of these shunts is important as they can mimic HCC [17]. Extrahepatic PPV collaterals (Figure 15) and portal vein cavernomas are commonly seen with portal vein thrombosis [1].

Conclusion

Congenital and acquired hepatic vascular shunts are frequently seen on cross-sectional imaging examinations, particularly on sonography and MDCT. Awareness of the various types of hepatic vascular shunts can assist the radiologist in establishing an accurate diagnosis and in guiding appropriate patient management.

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