Received:
27 December 2014

Revised:
13 May 2015

Accepted:
27 May 2015

doi: 10.1259/bjr.20150008

Cite this article as:

Arora A, Sarin SK. Multimodality imaging of primary extrahepatic portal vein obstruction (EHPVO): what every radiologist should know. *Br J Radiol* 2015; **88**: 20150008.

PICTORIAL REVIEW

Multimodality imaging of primary extrahepatic portal vein obstruction (EHPVO): what every radiologist should know

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ABSTRACT

Portal vein thrombosis (PVT) is a frequent complication of liver cirrhosis, but it can also occur as a primary vascular disorder amid absent liver disease. Extrahepatic portal vein obstruction (EHPVO) refers to the obstruction of the extrahepatic portal vein with or without involvement of the intrahepatic portal vein branches, splenic and/or superior mesenteric vein. It is a distinct disorder that excludes PVT occurring in concurrence with liver cirrhosis or hepatocellular carcinoma. The term “EHPVO” implies chronicity and is principally reserved for a long-standing condition characterized by cavernous transformation of the portal vein. The most characteristic imaging manifestation is the formation of portoportal collaterals (*via* the venous plexi of Petren and Saint) that allow hepatopetal flow. However, this collateral circulation is insufficient resulting in clinically significant pre-hepatic portal hypertension, wherein the liver function and structure remain preserved until late. Although the long-term (more than 10 years) survival with controlled variceal bleeding is up to 100%, affected individuals have an impaired quality of life owing to portal cavernoma cholangiopathy, hypersplenism, neurocognitive dysfunction and growth retardation. Imaging diagnosis is not always straightforward as the collaterals can also present as a tumour-like solid mass that can be inadvertently biopsied. Moreover, EHPVO has its implications for the biliary tree, arterial circulation, liver/splenic volumes and stiffness, which merit proper understanding but have not been so well described in literature. In this review, we present the complete spectrum of the vascular, biliary and visceral changes with a particular emphasis on what our medical/surgical hepatology colleagues need to know from us in the pre-operative and post-operative settings.

Extrahepatic portal vein obstruction (EHPVO) refers to the obstruction of the extrahepatic portal vein with or without involvement of the intrahepatic branches, splenic vein (SV) and/or superior mesenteric vein (SMV). The term EHPVO implies chronicity and is principally reserved for a long-standing condition characterized by cavernous transformation of the portal vein. It is a distinct (primary) vascular disorder that excludes acute or chronic portal vein thrombosis (PVT) occurring in concurrence with liver cirrhosis or hepatocellular carcinoma.^{1,2} Along with obliterative portal venopathy (OPV), EHPVO constitutes an important cause of non-cirrhotic (pre-hepatic) portal hypertension (NCPH), wherein the liver function and structure remain preserved until late. It has been proposed that an infection or a prothrombotic event occurring early in life (in a genetically predisposed individual) precipitates thrombosis of the main portal vein leading to EHPVO. By contrast, repetitive microthrombotic events occurring late in life, which involve smaller intrahepatic portal venous branches, are responsible for OPV.¹

EHPVO is primarily a disorder of children and young adults and is the most common cause of paediatric portal hypertension (PHT) in developing countries. Also, it is the most common cause of gastrointestinal bleed in children and adolescents (68–84%). Whereas non-cirrhotic non-tumoral PVT in the Western world constitutes the second most frequent cause of PHT in adults, it is responsible for only 11% of cases of paediatric PHT.¹ The aetiology of EHPVO differs in paediatric and adult populations (Table 1), but hypercoagulable states are most commonly incriminated. Nevertheless, the vast majority of cases (up to 70%) may remain idiopathic despite thorough laboratory and clinical workup.^{1,2}

Patients with EHPVO typically present in the first two decades with symptomatic PHT most commonly in the form of (well-tolerated) episodes of upper gastrointestinal bleed. The long-term (more than 10 years) survival with controlled variceal bleeding is as high as 100%;¹ however, affected individuals have an impaired quality of life owing to biliary complications (portal cavernoma cholangiopathy), hypersplenism

Table 1. Aetiology of extrahepatic portal vein obstruction

Children	Adults
Infections Omphalitis Neonatal umbilical sepsis Intra-abdominal infections Post umbilical catheterization	Prothrombotic state Myeloproliferative disorders (e.g. polycythaemia rubra vera, thrombocytosis, myelofibrosis etc.) Protein-C deficiency Protein-S deficiency Antithrombin-III deficiency Antiphospholipid syndrome Anticardiolipin antibody Factor-V Leiden deficiency Hyperhomocysteinaemia Paroxysmal nocturnal haemoglobinuria
Trauma Umbilical vein cannulation Childhood abdominal trauma	Trauma and surgery Abdominal surgery (splenectomy, pancreatic surgery etc.)
Congenital anomaly Congenital portal vein stenosis Portal vein atresia/agenesis	Local inflammatory conditions Pancreatitis Liver abscess
Prothrombotic state Prothrombin gene (G20210A) mutation Methylene tetrahydrofolate reductase gene mutation (C677T) Protein-C deficiency Protein-S deficiency Factor-V Leiden deficiency Antithrombin-III deficiency Antiphospholipid syndrome Anticardiolipin antibody	Miscellaneous Pregnancy Oral contraceptive use Post liver transplant
Idiopathic	Idiopathic

(thrombocytopenia, sepsis owing to leukopenia, anaemia etc.), neurocognitive dysfunction owing to subclinical hepatic encephalopathy and growth retardation.^{1,2}

Figure 1. Pictorial depiction of extrahepatic portal vein obstruction illustrating the venous, arterial, biliary and visceral changes.

Figure 2. Pictorial illustration of portal cavernoma formation in extrahepatic portal venous obstruction, which is composed of two peribiliary venous plexi: the paracholedochal plexus of Petren (running parallel to the bile duct) and the epicholedochal plexus of Saint, located on the surface of the bile duct wall.

Herein, we discuss the spectrum of vascular, biliary and visceral manifestations of EHPVO (Figure 1) that one should be aware of, highlighting the role of each imaging modality, with a particular emphasis on what our medical/surgical hepatology colleagues need to know from us in both the pre-operative as well as the post-operative settings.

Figure 3. (a, b) Ultrasonography and colour Doppler revealing serpiginous portoportal collaterals replacing the portal vein (cavernoma formation). (c) Spectral Doppler shows monophasic hepatopetal flow. (d) Attendant gall-bladder varices are also noted.

Figure 4. (a) Pseudotumoral extrahepatic portal vein obstruction wherein the thrombosed portal vein and marked periportal fibrosis (arrows) simulate a solid juxta-hilar mass. (b) Colour Doppler confirms the vascular nature of the juxta-hilar soft tissue by displaying serpiginous portoportal collaterals (arrows). There is attendant mild biliary dilatation (arrowhead) in keeping with portal biliopathy.

VENOUS CHANGES

EHPVO is characterized by cavernous transformation of the portal vein (portal cavernoma), which substitutes for the thrombosed portal venous system. Portal cavernoma formation is nothing but the formation of extensive portoportal collaterals that attempt to preserve hepatopetal flow (from the splanchnic veins to the intrahepatic portal veins). These portoportal collaterals are formed *via* the two well-formed venous plexi of the bile ducts: paracholedochal and epicholedochal plexi of Petren and Saint, respectively, which dilate in an attempt to bypass the thrombosed portal vein (Figure 2).^{3,4} Development of this portoportal collateral circulation is, however, insufficient to bypass the entire spleno-mesenteric inflow resulting in development of PHT, which is characterized by the formation of portosystemic shunts

Figure 5. (a-d) Axial contrast-enhanced CT delineating the portal cavernoma (arrows). Additional to the extrahepatic portal venous system involvement, there is involvement of the intrahepatic branches (black arrowheads). Note signs of non-cirrhotic portal hypertension: splenic enlargement (asterisk) and gastro-oesophageal varices (white arrowhead).

Figure 6. (a, b) Coronal contrast-enhanced CT and maximum intensity projection showing extrahepatic portal vein obstruction (white arrows) with spared superior mesenteric vein (dotted white arrow) and splenic vein (dotted black arrow). Note the dilated left gastric vein (black arrowhead) supplying the gastric varices (white arrowhead). (c, d) Coronal contrast-enhanced MRI delineating the peribiliary collaterals (arrows) and their relationship with the biliary tree.

(primarily *via* the left gastric vein and the perisplenic veins) and splenic enlargement.¹⁻⁵ Since PHT occurs in the presence of a functionally and morphologically normal liver, it is termed NCPH.^{1,2}

On greyscale ultrasound portal cavernoma is seen as a “sponge-like” mass of serpentine vessels in the hepatoduodenal ligament, porta hepatis and/or peripancreatic region with variable extension into the liver hilum (Figure 3a,b).¹ On spectral Doppler, the portoportal collaterals show monophasic hepatopetal flow with loss of normal respiratory undulations (Figure 3c). Gallbladder varices are commonly present (in 30–50% of patients) and can be evaluated well on ultrasound–Doppler (Figure 3d). EHPVO can at times simulate a solid mass at the hepatic hilum and the

Figure 7. (a, b) Axial arterial and portal venous phase CT displaying pseudotumoral extrahepatic portal vein obstruction simulating a solid soft tissue mass lesion (arrows).

Figure 8. Portal cavernoma cholangiopathy. (a) Thick-slab two-dimensional MR cholangiopancreatography showing an ectatic common duct with extrinsic indentations (arrowheads), luminal compromise and upstream biliary dilatation. (b) Coronal CT minimum intensity projection showing paracholedochal collaterals indenting the biliary tree (arrowheads). (c) Coronal CT maximum intensity projection showing cavernomatous transformation of the portal vein (arrowheads).

use of colour Doppler is crucial to establish its vascular nature (Figure 4). Ultrasound–Doppler has a high sensitivity and specificity (>95%) for establishing EHPVO; however, the exclusion of other possible causes of portal vein obstruction and evaluation of the exact extent of vascular involvement warrants contrast-enhanced CT or MRI.¹

CT/MRI displays changes in the splanchnic circulation with high precision and allows assessment of the exact extent of obstruction of the portosplenomesenteric axis (Figure 5).^{5,6} Post-processing tools such as maximum intensity projection, volume rendering and surface shaded display are especially useful to delineate the network of portoportal and portosystemic

collaterals and study their relationship with the bile ducts (Figure 6). Additionally, apart from confirming the diagnosis, cross-sectional imaging allows exclusion of tumoral PVT and other possible causes of portal vein obstruction (e.g. chronic pancreatitis etc.).

A potential pitfall on imaging is the “tumour-like cavernoma”, wherein a tumour-like solid mass is seen at the hepatic hilum with non-visualization of individual collateral channels (Figure 7). This has been attributed to abundant fibrosis around individual periductal veins.^{3,5} One has to be aware of such a presentation as an inadvertent biopsy can precipitate profuse bleeding.

Table 2. Types of portal cavernoma cholangiopathy (PCC)

	Varicoid PCC	Fibrotic PCC	Mixed PCC
Definition	Biliary obstruction by large paracholedochal collaterals	Biliary obstruction caused by ischaemia of the bile duct wall	Extrinsic compression + mural ischaemic fibrosis
Peribiliary venous plexus responsible	Paracholedochal venous plexus of Petren	Epicholedochal venous plexus of Saint	Plexi of Petren and Saint
Aetiopathogenesis	Extrinsic mechanical compression of the biliary tract	Ischaemia and fibrous scarring of the bile duct wall	Extrinsic compression ± bile duct ischaemia and fibrosis
Imaging	“Wavy” or undulating contour of the bile ducts Smooth multiple extrinsic/peribiliary collateral channels Paracholedochal varices enhance best on the portal venous phase	Localized stricture with upstream dilatation Densely enhancing thickened bile duct wall causing luminal narrowing Progressive delayed enhancement connoting intramural fibrosis	Irregular bile duct contours with multifocal areas of narrowing and dilatation Wall thickening at the level of the narrowed portion does not show delayed increased enhancement
Clinical significance	May be reversible with decompression of the splanchnic venous system (surgical portosystemic shunt creation)	Does not revert following portosystemic shunt surgery	Variable response

Figure 9. Varicoid portal cavernoma cholangiopathy (PCC): (a, b) thick-slab two-dimensional (2D) MR cholangiopancreatography (MRCP) and contrast-enhanced MRI showing a “wavy” contour (arrowheads) of the bile duct owing to paracholedochal varicosities (arrows). Fibrotic PCC: (c, d) thick-slab 2D MRCP and contrast-enhanced CT showing smooth annular enhancing bile duct wall thickening (arrows) with luminal compromise (arrowheads) and upstream biliary dilatation.

BILIARY CHANGES

Owing to intricate anatomic contact between the cavernoma and the biliary tree, biliary changes are seen in 70–100% cases of EHPVO. This may involve the extrahepatic/intrahepatic bile ducts and/or the gallbladder and is referred to as portal biliopathy or portal cavernoma cholangiopathy (PCC). However, only a small patient population is clinically symptomatic (5–18%). Postulated mechanisms of PCC include extrinsic mechanical compression or ischaemic insult with resultant fibrosis of the biliary tree. The spectrum of biliary changes that constitute PCC includes extrinsic indentations (Figure 8),

Figure 10. (a) Thick-slab two-dimensional MR cholangiopancreatography in a patient with extrahepatic portal vein obstruction and portal cavernoma cholangiopathy (PCC) showing choledocholithiasis (arrow) and hepaticolithiasis (arrowhead). (b) Coronal contrast-enhanced CT showing liver abscesses in a patient with PCC and severe cholangitis.

Figure 11. (a) Endoscopic retrograde cholangiopancreatography showing bile duct narrowing in a patient with extrahepatic portal vein obstruction. (b) Post-biliary stenting (arrow).

luminal narrowing $\pm$ upstream dilatation, bile duct thickening, angulation/displacement of the extrahepatic duct, choledocholithiasis and hepatic lithiasis (owing to cholestasis).^{3–6} The suprapancreatic common duct is the most commonly involved, and the stenosis can be either short segment (<25 mm) or long segment (>25 mm).⁵

The imaging manifestations (based upon the aetiopathogenesis, type of narrowing and implications for therapeutic planning) can be categorized into three subtypes: varicoid, fibrotic and mixed PCC (Table 2) (Figure 9).^{3,4} It is important to look for allied complications (Figure 10), which include cholangitis, liver abscess(es), choledocholithiasis, hepaticolithiasis etc.

Whilst ultrasound and CT can allow visualization of PCC changes, MR cholangiopancreatography remains the modality of choice.^{1–5} Endoscopic retrograde cholangiopancreatography is reserved only for those requiring therapeutic intervention (Figure 11). Endoscopic ultrasonography is not routinely employed and is generally recommended only when differentiation between fibrous collateral tuft, stones and tumours is not possible with other imaging modalities.

VISCERAL CHANGES

Hepatic changes

EHPVO is primarily a pre-hepatic disorder, and the liver size, architecture, volume and echotexture remain normal.¹ Relative decrease in portal perfusion at the liver periphery with

Figure 12. Axial contrast-enhanced CT showing decreased perfusion at the liver periphery (arrowheads). The central liver in contrast stays better perfused by the portoportal collaterals.

Figure 13. (a) Advanced extrahepatic portal vein obstruction (EHPVO) with liver contour undulations (arrows) simulating cirrhosis. (b) Liver cirrhosis (white arrows) accompanied with chronic portal vein thrombosis (black arrow). Hepatic vein catheterization and assessment of the hepatic venous pressure gradient can help differentiate advanced EHPVO from liver cirrhosis.

compensatory increase in arterial perfusion can be seen as the portal collateral vessels better perfuse the central liver (Figure 12).⁶ Long-standing compromised portal perfusion can cause parenchymal extinction and smooth hepatic atrophy. Decreased perfusion at the periphery may lead to subcapsular atrophy ensuing nodular liver contours simulating cirrhosis. In confounding cases, hepatic haemodynamic studies by an interventionalist (Figure 13) can help to differentiate the two by revealing an elevated wedged hepatic venous pressure in cirrhosis (which remains normal in EHPVO owing to pre-sinusoidal PHT). Alternatively, liver biopsy can be employed.¹

Splenic changes

Patients with EHPVO have a hyperdynamic circulation attributed to elevated levels of nitrous oxide, with notably increased splenic blood flow and consequent moderate to massive splenomegaly (average size, 11 cm below the costal margin).¹ The presence of intrasplenic siderotic nodules (Gamna–Gandy bodies) on imaging connotes long-standing PHT (Figure 14). Moreover, the splenic stiffness increases,⁷ and a value >42.8 kPa [ultrasound transient elastography (FibroScan®; Echosens™, Paris, France)] predicts variceal bleed with fairly good accuracy [sensitivity (88%) and specificity (94%)].

Figure 14. (a) Coronal CT showing massive splenic enlargement with multiple calcified Gamna–Gandy bodies (arrows). (b) Coronal fast imaging employing steady-state acquisition sequence MRI showing “blooming” of these fibrosiderotic nodules (arrows)—stigmata of long-standing portal hypertension.

ARTERIAL CHANGES

The “splenic hyperkinetic state” also promotes development of splenic artery aneurysms, which are often large at presentation (Figure 15). Their presence, number and dimension should be documented as those >2 cm are at high risk of rupture and should be treated surgically or with laparoscopic ligation or percutaneous embolization.⁸

TREATMENT

EHPVO warrants a multidisciplinary approach that includes the management of variceal bleed, portal biliopathy and massive splenomegaly.^{1,2,8–10} Therapeutic options range from conservative medical therapy, endoscopic variceal ligation, endotherapy (biliary stenting $\pm$ sphincterotomy, stone extraction etc.) to surgical intervention, that is, formation of portosystemic shunt and/or splenectomy. Patients with tight symptomatic biliary strictures may require hepaticoenterostomy to relieve biliary obstruction. Liver transplantation is reserved for those with life-threatening complications not manageable by any of the afore-said measures.

Figure 15. (a) Axial CT maximum intensity projection (MIP) showing splenic artery hypertrophy (arrowheads) denoting splenic hyperkinetic circulation, with an aneurysm (arrow) at the splenic hilum. (b) Coronal CT MIP in a different patient with extrahepatic portal vein obstruction showing splenic artery dilatation (arrowhead) with multiple aneurysms (arrows) giving a bunch-of-grapes appearance.

Figure 16. Types of surgical portosystemic shunts. (a) Proximal spleno-renal shunt following splenectomy in which the proximal end of the cut splenic vein (SV) is anastomosed to the left renal vein (LRV). It also treats the painfully enlarged spleen/hypersplenism and is preferred in adult patients. (b) Distal spleno-renal shunt: SV is detached from the portal vein (PV) and reattached to the LRV. Spleen is preserved (hypersplenism/ massive splenic enlargement is not tackled). (c) Mesocaval shunt: a prosthetic or native vein is used as a conduit between the superior mesenteric vein (SMV) below the pancreas and the inferior vena cava (IVC). (d) Rex shunt: an autologous vein graft (internal jugular vein) is used to bypass the mesenteric blood from the SMV to the intrahepatic left portal vein (LPV) branch. PC, portal cavernoma.

Decompression of the portomesenteric axis can be achieved by surgical portosystemic shunting typically by spleno-renal or mesentericocaval anastomosis or Rex shunt (mesenterico-left portal bypass) (Figure 16).^{1,2,10} In children, the Rex shunt is preferred since it is more physiological and restores hepatopetal flow to the liver; however, it can be created only if the left portal vein branch and SMV are patent. The Rex shunt has been shown to correct liver dysfunction, coagulation parameters and improve growth potential in paediatric patients.^{1,2}

Surgical portosystemic shunts (SPSSs) are extremely useful in patients with failed medical therapy, hypersplenism, massive

variceal bleeding or those with ectopic varices (colonic, jejunal etc.) not amenable to treatment. SPSSs apart from controlling variceal bleed also relieve portal biliopathy, improve growth retardation and hypersplenism. Owing to their long-term advantages, they have become the initial procedure of choice in paediatric EHVPO.^{1,2,10}

Pre-operative imaging for surgical portosystemic shunts

As the aforementioned shunt procedures are often rendered unfeasible owing to concurrent SV/SMV and/or intrahepatic PVT, CT or MR portography is required to visualize and

Figure 17. (a, b) Axial contrast-enhanced CT maximum intensity projections showing an unusually dilated gastroepiploic vein (arrowheads) that can be used for shunting in case no other suitable channel is available.

Figure 18. (a, b) Greyscale ultrasound and colour Doppler of the left (LT) kidney showing a patent proximal splenorenal shunt (PSRS). The left renal vein (LRV) and inferior vena cava (IVC) are patent. (c) Coronal CT maximum intensity projection delineating the patent shunt (arrow) draining into the left renal vein.

document the patency/involvement of the entire splanchnic axis. Involvement of each vessel needs to be explicitly highlighted in the report. Additionally, renal vein and inferior vena cava patency should be assessed and anatomic variants, if any, should be reported as a pre-operative roadmap for the surgeon. Patients with concomitant SV/SMV thrombosis often warrant alternate anastomosis with other suitable portal varix (e.g. the gastroepiploic vein), and these should be carefully looked for (Figure 17). For a surgical anastomosis to be successful and to satisfactorily decompress the portal system, the shunt should be of sufficient size (at least 10 mm in diameter).

Post-surgical assessment

Although post-surgical regression of gastro-oesophageal varices and congestive gastropathy (on endoscopy) are indirect signs that suggest shunt patency, nevertheless direct visualization of the shunt vessel is advocated. This can be performed using ultrasound–Doppler; however, CT or MRI allows superior delineation of the anastomotic channel (Figure 18). Moreover, potential post-surgical complications

such as renal vein thrombosis etc. (Figure 19) can be readily picked up on cross-sectional imaging.

ROLE OF RADIOLOGICAL INTERVENTIONS

The role of radiological interventions in patients with EHPVO is increasing. Patients with refractory massive variceal bleeding can be subjected to transjugular intra-hepatic portosystemic shunt,^{9,10} albeit the procedure is technically challenging and can preclude a future Rex shunt.² Portal biliopathy patients with cholangitis/jaundice might benefit from transhepatic biliary drainage or placement of an internal–external biliary drain, until the portal venous system is decompressed by SPSS creation or hepaticoenterostomy. Partial splenic embolization can be employed to decrease splenic size and alleviate hypersplenism, while preserving the immunological function *via* the spared splenic tissue. Splenic artery aneurysms can be subjected to percutaneous embolization. Lastly, thrombosed portosystemic surgical shunts can be subjected to chemical or mechanical thrombectomy to preclude repeat surgery.⁹

Figure 19. (a, b) Axial and coronal contrast-enhanced CT showing renal devascularization and atrophy (arrows) in a patient with post-operative shunt occlusion and left renal vein thrombosis.

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