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Learning objectives

To illustrate the classification of porto-systemic shunts (PSS), the role of imaging in diagnosis of vascular anomalies and eventually associated liver focal lesions, such as benign neoplasms or hepatocellular carcinoma (HCC). This in order to recognize malformations, correlate nodular liver lesions associated to perfusion anomalies and obtain a prognostic stratification of patients.

Furthermore, to define the contribution of radiological assessment in definition of therapeutic strategies on the basis of residual portal circulation.

In this poster we will analyse the value of contrast-enhanced (CE) MDCT and MRI primarily in the definition of PSS and characterization of liver focal lesions respectively.

Our aim is to make radiologists aware of the importance of correct identification, classification of PSS and interpretation of related diseases, in order to refer patients to an appropriate treatment.
**Background**

In PSS, porto-mesenteric blood drains into systemic circulation partially or completely bypassing the liver. These anomalies could be acquired or congenital.

**Acquired shunts** are the most common, their major causes being portal hypertension or trauma. In the first case, increased pressure of portal blood causes normally existing but insignificant porto-systemic anastomoses to enlarge. More than 20 such pathways have been described, from gastro-esophageal, para-esophageal, para-umbilical and spleno-renal to inferior mesenteric district.

**Congenital shunts** are a rare consequence of abnormal involution of vitelline veins and their primitive anastomoses. The development of portal venous system is extremely complex and occurs between 4th and 10th week of embryonic life. Initially, right and left vitelline veins emerge from yolk sac, cross septum transversum, and directly drain into sinus venosus (the primitive heart). Three cross communicating vessels develop between the two vitelline veins: the subhepatic (cranio-ventral duodenal), intermediate (dorsal duodenal) and caudal (ventral duodenal), respectively. They anastomose each other forming the figure of an "8" around developing duodenum. Selective involution of these vessels leads to the final configuration of portal vein (PV) that remains posterior to duodenum (Fig. 1a, b, c, d). It is clearly understandable how several abnormalities in the anatomy of portal system might arise due to errors in the development process described. Abnormal patterns of involution and persistence can result in pre-duodenal, pre-biliary or duplicated PV, as well as excessive involution may hesitate in PV absence [1].

Associated anomalies can involve posterior venous system leading to azygos continuation of inferior vena cava (IVC).
Congenital PSS are classified into intrahepatic or extrahepatic. In the first, connections are created between branches of PV, after its division, and IVC or hepatic veins (HV). In the second, anastomoses are established outside the liver between porto-mesenteric vasculature, before PV division, and a systemic vein. Both categories can be further divided into various subtypes.

**Intrahepatic shunts** are classified by Park et al. [2] into:

- **type 1**, a single large vessel connecting right PV to IVC
- **type 2**, one or more communications between peripheral branches of PV and HV, within one hepatic segment
- **type 3**, an aneurysmal communication between peripheral PV and HV
- **type 4**, multiple communications between PV and HV, distributed in both lobes

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**Fig. 1**

First two varieties are the most common [3]. Although rare, persistent patent ductus venosus (DV) could be considered another type of intrahepatic PSS. In the fetus, DV arises from posterior aspect of left PV branch, opposed to the opening of umbilical vein, and drains into HV. Its spontaneous closure usually begins immediately after birth and becomes definitive during first week of life in most full-term neonates. Yet it may take much longer to close in preterms or in presence of congenital heart defects [3], as well as it may remain patent, leading to development of PSS.

**Extrahepatic shunts** are characterized by porto-mesenteric vasculature bypassing the liver through a congenital conduit. Most common drainage vessel is the IVC (porto-caval shunt), followed by renal, iliac, azygos vein and right atrium [4]. Extrahepatic shunts are classified by Morgan and Superina [5], according to Abernethy’s description [6], into two types (Fig. 2).

- **In type 1**, there is a complete portal blood diversion into systemic circulation (end-to-side shunt), with absence of intrahepatic portal branches. They are further classified into two subtypes in which splenic (SV) and superior mesenteric vein (SMV) separately drain into systemic outflow (type 1a), or join together forming a common trunk (type 1b). Pathogenesis has been attributed to excessive involution of primitive periduodenal vitelline venous loop or total failure of vitelline veins to establish critical anastomoses with hepatic sinusoids or umbilical veins [7, 8]. Resulting shunt may be due to persistence of right vitelline (draining into retrohepatic IVC) or left vitelline vein (right atrium or IVC above HV confluence) [3].

- **In type 2**, intrahepatic PV is intact, but part of portal flow is diverted into a systemic vein through a side-to-side communication. Persistence of anastomotic channels between vitelline and subcardinal veins has been proposed as a cause [9]. Other authors suggest some type 2 shunts probably arise from persistence of right vitelline vein [10].
**Fig. 2:** Different types of congenital extrahepatic PSS: partially (type 2) or completely bypassing the liver, with SV and SMV separately draining (1a) or joining together to form a common trunk (1b).


Strictly speaking, complete absence of venules within portal triads (definitively confirmed by liver biopsy) is defined as portal system agenesis. Similarly, macroscopic absence of PV, associated with the presence of intrahepatic portal venules, can be referred to as PV atresia.

Congenital PSS are often associated with hepatopaties and, in nearly one half of cases, liver tumors, due to different hepatic biohumoral responses to decreased portal blood supply. Pathogenesis could be found in inadequate delivery of growth factors and hormones to the liver, associated with compensatory increase in hepatic arterial flow [11]. Benign liver focal lesions are frequent and range from focal nodular hyperplasia (FNH) [12], to nodular regenerative hyperplasia (NRH) [13], hepatocellular adenoma [14], and hemangioma. Most focal lesions described in the literature as FNH or adenomas probably are regenerative nodules, frequently occurring in systemic disorders with liver vascular
derangement. Biopsy of these nodules is not usually necessary for confirmation of diagnosis because of benign imaging appearance. Nevertheless, potential evolution into hepatoblastoma or HCC has been reported [15]. Thus, long-term clinical and radiological follow-up are recommended.

Congenital PSS are frequently associated with other anomalies, including pre-duodenal PV, azygos continuation of IVC, situs ambiguous, malrotation, duodenal atresia, polysplenia, annular pancreas, skeletal and genitourinary anomalies [4]. Congenital heart defects are particularly related to PSS, suggesting either a prenatal insult during simultaneous development of the heart and abdominal venous system, or an adaptive response to hyperdynamic effect of the shunt. Atrial or ventricular septal defects, patent foramen ovale or ductus arteriosus, and tetralogy of Fallot are often observed [16].

**Treatment** of congenital shunts is primarily determined by the type of venous diversion (Fig. 3). Liver transplantation is the only effective treatment for symptomatic type 1 extrahepatic shunts. Surgical resection or liver transplantation are also necessary in case of malignancy, liver dysfunction or porto-systemic encephalopathy unresponsive to medical therapy, independently of the type of shunt. For type 2 extrahepatic and some cases of intrahepatic PSS, including persistent patent DV, potential therapeutic options include surgical ligation or percutaneous embolization of the shunt. This is essential if hepatic encephalopathy or benign liver tumors are associated. Anyway, sudden occlusion of the shunt may cause portal hypertension in case of hypoplastic intrahepatic portal system. It is important to assess, through color Doppler analysis of blood flow velocities, if PV and its intrahepatic branches can safely accommodate increased hepatopetal venous inflow resulting from shunt occlusion. When indicated, a staged procedure with preliminary narrowing of the shunt before final occlusion could be performed. Otherwise, transjugular intrahepatic porto-systemic shunt (TIPS) can be created to control symptomatic complications of portal hypertension, such as variceal bleeding or refractory ascites, in patients awaiting for liver transplantation.
Fig. 3: Algorithm for treatment of congenital extrahepatic PSS.

Images for this section:

Fig. 1

**Fig. 2:** Different types of congenital extrahepatic PSS: partially (type 2) or completely bypassing the liver, with SV and SMV separately draining (1a) or joining together to form a common trunk (1b).

**Fig. 3:** Algorithm for treatment of congenital extrahepatic PSS.

Imaging findings OR Procedure details

Congenital PSS are usually discovered in symptomatic infants with nonspecific liver disease or incidentally seen also in older patients undergoing radiologic evaluation for other reasons. Ultra-sonography (US) is the initial imaging modality, since easily available, non invasive, requiring no sedation and not exposing children to ionizing radiation. In most cases it can demonstrate absence of PV, Doppler analysis proving useful to determine flow direction and type of anomalous vessels identified. Yet US may fail to accurately define associated extrahepatic shunts [16].

CE-MDCT and MRI are helpful in confirming diagnosis of congenital PSS. Both angiographic techniques can clearly depict the course of anomalous porto-systemic vessels. Postprocessing techniques, such as maximum intensity projections (MIP) and multiplanar reconstructions (MPR), provide additional information. Since development of multidetector machines, CT angiography (CTA) spatial resolution has proven superior than MRI. Thus CTA can accurately define the type of shunt, identifying any residual portal branch, in a single breath hold [17].

CE-MRI is either a reliable and noninvasive technique for the evaluation of hepatic vascular anatomy, not exposing patients to ionizing radiation. Yet it results more effective in detecting eventually associated liver parenchima derangement, focal lesions, or other types of malformation, usually occuring in case of congenital PSS.

Currently, interventional angiography plays a role almost exclusively in selective embolization of type 2 congenital extrahepatic and some varieties of intrahepatic PSS, as a possible therapy. Furthermore, percutaneous transhepatic or transjugular portography allows measurement of pressure gradients, as well as transvenous liver biopsy at the same time [16, 18]. Histological sampling of liver parenchima results useful in patients suspected of having type 1 congenital extrahepatic shunts, since it may reveal residual portal venules within portal triads. In such cases, a type 2 shunt, previously misdiagnosed as type 1 basing on imaging findings alone, can be identified [19].

Our population consists of two type 1b extrahepatic, one type 1 intrahepatic congenital PSS and some representative samples of acquired shunts. Patients with congenital PSS presented benign liver focal lesions on imaging.

US color Doppler protocol

- Aplio XG scanner (Toshiba America Medical Systems, California, USA)
- Convex transducer
- Range of frequencies: 3-7 MHz
• Abdominal subcostal scans

**MDCT scanning protocol**

- LightSpeed VCT XTe MDCT 64 slices scanner (GE Medical Systems, Milwaukee, USA)
- Scan volume from dome of diaphragm to femoral diaphyses
- Helical scan; Slice thickness: 2.5 mm; Pitch: 1
- 20 G intravenous cannula in a distal arm vein
- 2 ml/kg iodinated contrast medium (Iopamiro 370 mgI/ml, Bracco Imaging Italia, Milan) + 60 ml saline chase @ 3.0 ml/s
- Dynamic scanning involving hepatic arterial and portal phases, obtained at 30 and 70 s from start of infusion respectively

**MRI scanning protocol**

- Signa Advantage 1.5 T scanner (GE Medical Systems, Milwaukee, USA)
- Intensity of magnetic field gradient: 23 mT/m
- Scan volume from dome of diaphragm to iliac crests
- Abdominal 8 channel phased-array coil
- Multiplanar gradient and fast spin echo T1- and T2-weighted images, even with fat suppression
- Axial diffusion-weighted images (b values: 0 and 800 s/mm\(^2\))
- 0.5 ml/kg gadolinium-BOPTA (Multihance, Bracco pharmaceuticals, Milan) + 30 ml saline chase @ 2.0 ml/s
- Dynamic scanning involving hepatic arterial, portal and hepatocellular excretion phase (60’ after contrast administration)

**Congenital PSS**

The first case of our population was previously reported by De Gaetano et al. [11]. It was a 28 year old woman with past history of appendectomy and subsequent laparotomy for bowel adhesions, admitted at our institution because of abdominal pain. US showed several hyperechoic focal lesions in both hepatic lobes, in particular two in segments VI and VIII (3.5 and 2 cm in size, respectively) and one in segment II (4 cm). Color Doppler analysis of hepatic vascularization showed SV and SMV joining together to form a dilated short common vein, directly draining into suprarenal IVC. No intrahepatic portal branches were identified, while hepatic artery looked markedly hypertrophied. Abdominal CTA clearly defined alterations observed as congenital extrahepatic type 1b shunt, also showing multiple hypodense liver nodules (Fig. 4). CE-MRI confirmed presence of three liver focal lesions, appearing slightly hyperintense on T1-, isointense on T2-weighted images, and hypovascular with respect to adjacent parenchyma after paramagnetic contrast medium administration. Biopsy specimens suggested diagnosis of FNH. Angiography showed porto-systemic communication described to have high flow.
rates, pressure values being 6 mmHg in the anomalous conduit and 4 mmHg in right atrium. Portal blood never reached liver parenchyma.

Fig. 4: Axial CTA (a) and T1-weighted MR images (b), showing dilated portal trunk (straight arrow) directly draining into suprarenal IVC (curved arrow), completely bypassing the liver.


The second case was a 27 year old man with arachnodactyly, facial skeleton malformations, cognitive impairment and past history of bacterial pneumonia. This patient underwent radiological examination due to abdominal pain and an alteration of liver function tests. CTA showed complete absence of portal trunk and its intrahepatic branches, normal patency of SV and SMV joining together to form a large anomalous vessel running within gastro-hepatic ligament, forming a loop around esophago-gastric junction (partially reducing it in caliber), and draining into left renal vein. This finding was appreciable on CE-MRI as well (Fig. 5). Hypertrophy of hepatic arterial circulation and accessory right hepatic artery originating from superior mesenteric artery, were also
noticed. Multiple hypodense lesions were found in both hepatic lobes, in particular two in segment VI and IV, measuring 3.5 and 1.7 cm in size respectively. Mild splenomegaly was present. Further diagnostic imaging through CE-MRI confirmed presence of liver focal lesions, hyperintense on T1- and isointense on T2- and diffusion-weighted images. After administration of paramagnetic contrast medium, they appeared hypointense in the arterial phase, isointense in following acquisitions of dynamic study, and hyperintense in hepatobiliary excretion phase, resulting referable to nodules of NRH.

**Fig. 5:** Reconstructed sagittal oblique CTA (a) and coronal MIP CE-MRI images (b) showing a large anomalous vessel originating from confluence of SV and SMV, running within gastro-hepatic ligament, forming a loop around esophago-gastric junction and draining into left renal vein.

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT

**The third case** consisted of a 26 year old man undergone abdominal US for pain in right hypocondrium. Several liver focal lesions were found, the greatest of them (13 cm in size) presenting mixed echostructure and almost completely occupying left hepatic lobe (Fig. 6). These nodules, first referred to liver metastases from unknown
origin, appeared worthy of further radiological assessment. A wide venous conduit putting into direct communication right portal branch and IVC, was put in evidence by color Doppler analysis. Sampling of bloodstream velocities showed high flow rate in the anomalous venous drainage, and analysis of waveforms demonstrated retrograde transmission of cardiac pulsatility till the level of PV (Fig. 7). CTA accurately described presence of congenital intrahepatic type 1 shunt, in which portal circulation was almost completely diverted into a new outflow path (Fig. 8). Compensatory arterial hypertrophy and accessory left hepatic artery originating from left gastric branch were also noticed. An inhomogeneous hypodense expansive lesion was documented by MDCT in left hepatic lobe, showing some arterial vessels inside after iodinated contrast medium administration (Fig. 9). CE-MRI, apart from illustrating intrahepatic shunt described (Fig. 10), resulted necessary to accurately characterize liver focal lesions. In particular, that enlarging left hepatic lobe was found to result from the confluence of multiple nodules, hyperintense on both T1- and T2-weighted images, and presenting areas of internal colliquation (Fig. 11). Inhomogeneity on hepatocellular excretion phase suggested the need for histological typing. Percutaneous liver biopsy was carried out and confirmed diagnosis of NRH. Several other lesions, detected in both hepatic lobes, were clearly referable to nodules of regenerative hyperplasia basing on imaging findings alone.
**Fig. 6:** Abdominal US showing inhomogeneous hyperechoic nodules in left (a) and right (b) hepatic lobes.

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT

**Fig. 7:** Liver color Doppler analysis showing high flow anomalous venous conduit connecting right portal branch and IVC (a), with retrograde transmission of cardiac pulsatility till PV (b).

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
Fig. 8: Axial (a) and reconstructed coronal (b) CTA images showing a large venous conduit constituting congenital intrahepatic type 1 shunt. Residual portal circulation was present.

References: Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
Fig. 9: Reconstructed coronal MDCT images before (a) and after (b) iodinated contrast medium administration, showing a huge inhomogeneous hypodense lesion in left hepatic lobe (a) presenting some arterial vessel inside (b).

References: Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
Fig. 10: Axial (a) and reconstructed coronal (b) CE-MR images showing congenital intrahepatic type 1 shunt described. Liver focal lesions (a) and mild splenomegaly (b) were also evident.

References: Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
**Fig. 11:** Axial T2-weighted (a) and CE-MR images (b) showing an inhomogenous lesion enlarging left hepatic lobe, presenting some hyperintense areas of internal colliquation (a) and uptake of paramagnetic contrast medium in the arterial phase (b).

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT

**Acquired PSS**

Some exemplificative images of acquired PSS, mainly due to portal hypertension, taken from our series, are given below (Figg. 12-15):
Fig. 12: Axial (a) and reconstructed sagittal (b) CTA images showing hepatofugal collateral circles through recanalized umbilical and inferior epigastric veins in a case of HCC with portal hypertension. Esophageal varices are also visible.

References: Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
**Fig. 13:** Reconstructed coronal (a) and oblique (b) CTA images showing gastric-left adrenal-ovaric collateral circles in case of focal PV thrombosis.

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
**Fig. 14:** CTA 3D reconstructed images showing hepatofugal collateral circles through superior mesenteric vein and right ovarian vein. It can be seen that ovarian vein appears filiform right before and dilated after the anastomosis with collateral circulation.

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
**Fig. 15:** Axial (a) and reconstructed coronal (b) CTA images showing gross spleno-renal collateral circles in case of liver cirrhosis with right lobe atrophy.

**References:** Dipartimento di Bioimmagini e Scienze Radiologiche, Policlinico Universitario A. Gemelli - Rome/IT
Fig. 4: Axial CTA (a) and T1-weighted MR images (b), showing dilated portal trunk (straight arrow) directly draining into suprarenal IVC (curved arrow), completely bypassing the liver.

Fig. 5: Reconstructed sagittal oblique CTA (a) and coronal MIP CE-MRI images (b) showing a large anomalous vessel originating from confluence of SV and SMV, running within gastro-hepatic ligament, forming a loop around esophago-gastric junction and draining into left renal vein.

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Conclusion

Correct identification and classification of vascular anomalies by means of CTA, as well as characterization of eventually associated liver focal lesions through CE-MRI, are crucial in defining a prognostic stratification of patients with congenital PSS.

All PSS conditions, either acquired or congenital, have a common physiopathological event that is reduction of portal perfusion. This is the main determining factor for development of nodular regeneration into the liver, despite compensatory increase in hepatic arterial flow. In congenital PSS, where almost complete portal diversion occurs, this pathological aspect is particularly conspicuous and a constant feature in these patients. In acquired forms, possibility of nodular regeneration mainly depends on the grade of portal hypertension and on the stage of liver function impairment.

A bias we noticed in our experience was that anyway performed radiological assessment could never document presence of venules in portal triads in case of PV atresia. This evaluation can effectively be carried out only through liver biopsy and histological sampling.

Radiologists should be aware of the importance of correct description of PSS and related diseases, such as hepatopaties, right heart impairment, secondary pulmonary hypertension and congenital alterations, either skeletal or cardiac. This in order to implement a correct therapeutic approach in patients with congenital PSS.
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