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BENIGN LIVER TUMORS ASSOCIATED WITH VASCULAR LIVER DISEASES

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Liver vascular disorders often have important physiologic consequences, both inside and outside of the liver. They may be categorized into abnormalities of outflow, abnormalities of inflow, and aberrant arteriovenous connections. These disorders often have distinct appearances on imaging examinations that can mimic other pathologies. The main common pathogenic feature is an imbalance between hepatic arterial and portal venous blood flow leading to an increased hepatic arterial inflow.

Hepatocellular nodules can develop on the background of chronic hepatic vascular disorders. The nodules can range from benign lesions such as regenerative nodules, focal nodular hyperplasia (FNH), and hepatocellular adenoma (HCA) to malignant neoplasms such as hepatocellular carcinoma (HCC). The most frequent are the FNH lesions. The preferred terminology is FNH-like, hemangioma-like, adenoma-like, as their radiological features are influenced by the background vascular abnormality of the liver. For example, in the congenital portosystemic shunts (CPSS) there is an increased arterial supply due to portal deprivation. On this background, in the arterial phase of the contrast enhanced MRI or contrast enhanced ultrasound only a mild hyperenhancement is depicted in the FNH-like nodules. MRI with hepatobiliary contrast agents is essential, as lesion signal intensity on hepatobiliary sequences is the key to characterizing liver nodules in CPSS. Biopsy is required in nodules that do not meet classical FNH criteria, nodules that increase in size or display heterogeneity or nodules that show hypo-intensity on delayed hepatobiliary contrast-enhanced MRI.

The accurate diagnosis of these nodules might be very difficult based on imaging or even histological features, but it is crucial for further management. Benign nodules such as regenerative nodules and FNH do not usually need resection, while neoplastic nodules such as HCA or HCC may warrant resection.

Keywords: vascular diseases of the liver – focal nodular hiperplasia like, adenoma-like, hepatocellular carcinoma – Budd Chiari syndrome – peliosis – congenital porto-systemic shunts.

INTRODUCTION

The major vascular disorders of the liver are represented by portal vein thrombosis (PVT), hepatic artery diseases (aneurysm, thrombosis), sinusoidal obstruction syndrome, radiation-induced liver disease, peliosis hepatis and sinusoidal dilatation, Budd-Chiari syndrome (BCS), congenital vascular malformations[1].

Portal vein thrombosis in patients with liver cirrhosis is the most common, followed by the PVT in malignant circumstances. Non-cirrhotic non-malignant PVT is an uncommon condition characterized by thrombosis of the portal vein, with or without extension into other mesenteric veins, in the absence of cirrhosis or intra-abdominal malignancy[2]. The other vascular liver disorders are even rarer, but it is important to recognize them, based on non-specific imaging features, to be able to differentiate these disorders from other conditions, as they might mimic other pathologies and also to be able to correctly evaluate their complications.

The most important pathological outcome is an increased hepatic arterial inflow. From a clinical point of view this hemodynamic imbalance has two important consequences: 1) a distinct imaging appearance of contrast-enhanced procedures, with a rapid arterial uptake of the contrast agent, and 2) the possibility to develop distinct nodular vascular entities, nodular.

Heterogeneous liver nodules have been described on the background of chronic hepatic vascular

disorders. Most of these nodules are benign lesions such as regenerative nodules, FNH, and hepatocellular adenoma (HCA), but in some circumstances malignant neoplasms such as hepatocellular carcinomas (HCC) have also been reported.

For benign lesions the preferred terminology is FNH-like, hemangioma-like, adenoma-like, as their radiological features are influenced by the background of vascular abnormality of the liver[3].

The most frequent are the FNH lesions. Wanless *et al*.[4] suggested many years ago, based on histological examinations, that FNH was a hyperplastic response of the liver parenchyma to an increase or a disturbance of hepatic blood flow due to a pre-existing vascular malformation, for example a preexisting arterial spider-like malformation[4]. Based on the frequent coexistence of FNH with other vascular anomalies they concluded that FNH are developmental in origin. The basic requirement for development of hepatic hyperplasia may be greater blood flow to a region compared to the adjacent parenchyma, also detected in nodular regenerative hyperplasia and partial nodular transformation[4]. Macroscopically, FNH is a circular, lobulated, well-circumscribed lesion, free of fibrous capsule and characterized by a central stellate fibrous scar containing large vessels. In microscopy, it consists of a hyperplasia of the liver parenchyma with benign-appearing hepatocytes, organized in nodules separated by fibrous septa, surrounding large vessels in the central scar often accompanied by bile ductular proliferation and inflammatory infiltration.

Other authors suggested a different mechanism for FNH formation, also based on histologic assessment. Kumagai *et al*.[5] considered that thrombosis of the hepatic artery and/or portal vein was the cause of hepatic necrosis and that reperfusion following hepatic arterial recanalization resulted in the nodule formation.

The clinical importance of liver nodules developed on a modified vascular background of the liver lies in their correct diagnosis and management.

In this review, the vascular disorders of the liver will be discussed, the potential risk of nodule development, their diagnosis and management.

PORTAL VEIN THROMBOSIS

As PVT in the cirrhotic patients and in those with malignancy has a different natural history, these are not discussed in this review.

Non-cirrhotic non-malignant PVT is an uncommon condition. Population prevalence estimates for portal vein thrombosis range from 0.05%–1% in autopsy studies[6], hospital series report standardized incidence rates between 0.7–3.79 per 100,000[7].

The etiology of non-cirrhotic non-malignant PVT is represented by one or more factors that affect the Virchow's triad: reduced portal blood flow, hypercoagulability, and vascular endothelial injury. The obstruction of the portal vein lumen leads to increased resistance to the flow creating back pressure within the portal system and development of portal hypertension. Two compensatory mechanisms have been postulated to maintain hepatic perfusion in the setting of portal vein obstruction. Firstly, a reflex increase in the hepatic arterial flow and secondly a rapid formation of porto-portal collaterals leading to cavernous transformation of the portal vein[2].

Bureau C *et al*.[8] described two cases of FNH in young patients with non-cirrhotic, non-tumoral PVT, highlighting the etiological connections between the increased arterial outflowed caused by portal vein thrombosis and the development of HNF. More recently, in a cohort study of 58 patients with cavernous transformation of the portal vein, FNH-like lesions were detected in 21% of patients[9]. FNH-like lesions were found with other benign liver lesions (1 hemangioma, 1 adenoma) in two patients. Only three (25%) patients with FNH-like lesions showed a progressive clinical course[9].

In case of a cavernous transformation of the portal vein, there is insufficient portal venous inflow far from the hilum, and consecutively it is an increase in the hepatic arterial flow especially in the peripheral liver segments. These hemodynamic liver changes cause central liver hypertrophy and peripheral liver atrophy[3], easily detected on imaging.

Most FNH-like lesions are typical on contrast-enhanced CT or MRI; however, a central scar often lacked due to their small size[9]. The MRI findings of HNF-like are: isointensity on T2-weighted images (82%), intense and homogeneous enhancement during the arterial phase, and lack of washout during the hepatic venous and interstitial phases (100%).

The diagnosis of FNH-like lesions is based on MRI, the most reliable sign being the detection of liver specific contrast agent on hepatobiliary phase (iso- or hyperintense lesions on hepatobiliary phase), corresponding to the bile ductular proliferation[3,9]. The size and number of the FNH-

like lesions may increase over time[9]. Adenoma and hepatocellular carcinoma are rare in these cases[10].

BUDD-CHIARI SYNDROME

The primary form of BCS occurs from the obstruction of hepatic venous outflow, usually because of hepatic vein thrombosis. Other possible causes are intrinsic venous narrowing or more rarely, webs obstructing the intrahepatic inferior vena cava or the hepatic veins. The secondary form can occur from extrinsic compression or direct venous invasion by the tumor[11]. The hemodynamic consequences of outflow obstruction are hepatic congestion and increased sinusoidal pressure, with development of portal hypertension and parenchymal fibrosis[11]. Secondary, intra- and extrahepatic venous collaterals develop, there is an increased hepatic arterial flow. Hypertrophy in areas of preserved hepatic venous outflow predispose patients to the formation of FNH-like lesions[12].

FNH-like lesions are reported in more than one-third of patients with chronic BCS and are not related with specific etiological factors of BCS[11,13]. Their size might increase or, more rarely, decrease over time. They are homogeneous and hypervascular and the largest lesions often have a central scar[14].

Certain imaging features differ from those of FNH such as hyperintensity on T1-weighted and variable signal T2weighted MR images (either iso-, hypo-, or hyperintense)[13]. An important aspect is that these FNH-like lesions might present wash-out of the contrast agent on CT or MRI during portal venous and/or delayed phase, because of increased signal intensity of the surrounding liver due to increased arterial inflow[13]. Biopsies in these cases might be required, as some of the nodules also might increase their size. Sometimes even the histological examination might be inconclusive as there are no specific immunohistochemistry markers[13].

Hepatocellular adenoma-like also might develop on the background of BCS[15]; they have different immunohistochemistry expression for liver fatty acid binding protein, glutamine synthetase, nuclear b-catenin, C reactive protein. Their malignant potential is considered higher than adenomas developed on normal vascular liver background[15].

Hepatocellular carcinoma (HCC) might develop in patients with BCS[16]. Serum alpha-fetoprotein (AFP) is helpful in monitoring patients with BCS and suspected nodules of HCC. Patients with long-term inferior vena obstruction are at a higher risk of developing HCC than those with pure hepatic vein involvement[16,17]. HCC usually appears hypervascular, heterogeneous, and solitary by imaging. The histological diagnosis might be difficult because frequently HCCs are highly differentiated tumors[16].

Monitoring patients with BCS and increasing in size vascular nodules might be challenging, both on imaging and histology assessment. Contrast-enhanced ultrasound or specific liver contrast agent MRI combined with alpha-fetoprotein monitoring might be helpful. Contrast-enhanced ultrasound and/or MRI are recommended as they depict with high accuracy mild hyperenhancement. On contrast-enhanced ultrasound, FNH-like lesions displayed center-to-periphery uptake of contrast-agent and the lesions remain hyperechoic on portal venous and delayed phases, enhancement of most HCCs was heterogenous on arterial phase and hypoechoic on portal and delayed phases[18].

CONGENITAL PORTO-SYSTEMIC SHUNTS

Congenital portosystemic shunt(s) (CPSS) are rare vascular malformations of embryonic origin through which intestinal blood flow bypasses the liver partially or completely, and thereby reaches the systemic circulation unfiltered[19].

CPSS are often associated with systemic complications (pulmonary vascular complications, liver nodules, neurological, endocrine, metabolic, hematological complications). Liver nodules of various histological types are highly prevalent in this population and may be malignant. Porto-pulmonary hypertension is the most life-threatening complication in patients with CPSS and requires regular monitoring both before and after shunt closure[19].

The complications associated with CPSS can be challenging to manage. At the time of diagnosis, a careful assessment of all possible complications might be done. If porto-pulmonary hypertension is present, a multidisciplinary approach and prompt treatment should be done. Understanding shunt anatomy and portal vasculature, quantifying portal pressure and portosystemic gradient with an occlusion test and evaluating nodule histology and size will determine the closure approach (endovascular, surgery or even liver transplantation). A risk stratification should be formulated, longitudinal follow-up is required for all patients prior to shunt closure, as well as post closure

for any patient with a liver nodule or pulmonary hypertension.

Different type of liver nodular lesions might be present in patients with CPSSs being related to major portal deprivation[20]. The most frequent clinical scenario is incidentally detections of multiple, heterogeneous liver nodules in a young patient. These nodules might not have typical vascular pattern and after a better assessment of the liver vessels on imaging, CPPSs are discovered (Fig. 1). Most patients have multiple liver nodules and FNH-like lesions, hepatocellular adenoma (HCA)-like or regenerative nodular hyperplasia, but HCC might also develop[3,19].

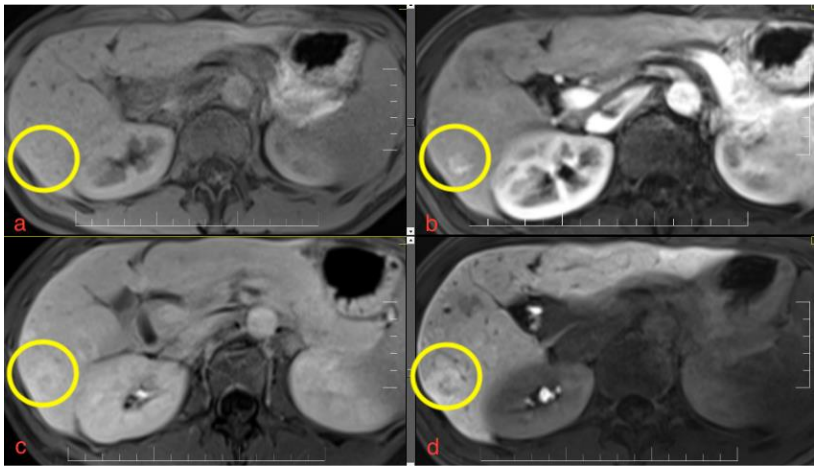

Fig 1. Focal nodular hyperplasia-like tumour. MRI with Primovist appearance.a) T1: iso-hypointense; b) T1 with contrast: early arterial enhancement) enhancement persists into delayed phases c).

The anatomic form of CPSS might influence the tumor type, owing to the degree of portal flow and compensatory arterial supply. Intrahepatic CPSS are typically associated with benign liver masses, while both benign and malignant tumors have been reported in extrahepatic CPSS[3,19].

FNH-like lesions are more often atypical on imaging than in patients with normal livers and may enlarge over time[3,19]. These atypical lesions may be explained by complete and chronic portal blood deprivation. Contrast-enhanced MRI and contrast enhanced ultrasound have high accuracy for depiction of mild hyperenhancement. MRI with hepatobiliary contrast agents has high accuracy for liver nodules characterization.

FNH lesions show no central scar, and only weak hyperenhancement on hepatic arterial phase, because high non-nodular liver enhancement results in diminished nodule-to-liver contrast ratio[19]. Hepatocellular adenoma subtyping on imaging is also very difficult in the setting of CPSS.

Before shunt closure, imaging is recommended every 6 months. In case of a “suspicious nodule”, imaging should be repeated every 3 months, or a biopsy should be obtained for histopathological diagnosis and immunohistochemical assessment (glutamine synthetase, b-catenin, serum amyloid A, C-reactive protein, CD34 glypican-3LFABP (liver-fatty acid binding protein)). If there is evidence of b-catenin activation, imaging monitoring at every 3 months should be done, considering the risk of HCC[22].

HEREDITARY HEMORRHAGIC TELANGIECTASIA

Hereditary hemorrhagic telangiectasia (HHT), also known as Rendu–Osler–Weber syndrome, is an autosomal dominant disorder that interferes with angiogenesis and its control mechanisms[3]. Microscopic telangiectasia and direct arteriovenous, arterioportal, and porto-venous shunts might be present, resulting in an increase in hepatic arterial flow. Liver involvement is present in 67–84% of patients, being associated with mutations in the ALK1 gene mutations (type 2)[23].

The spectrum of vascular liver lesions in patients with HHT is represented by FNH-like, intrahepatic shunts, nodular regenerative hyperplasia, sinusoidal dilatation and ischemic cholangiopathy that may coexist simultaneously in the same patient[24,25]. Buscarini *et al*.[26] estimated the prevalence of FNH-like lesions in HHT as 2.9%, significantly higher than in the general population. Most patients with FNH-like lesions had severe hepatic shunts and

were women[26]. FNH-like lesions might increase in size overtime. The contrast-enhanced MRI with hepatobiliary contrast agents may differentiate FNH-like lesions from large telangiectasia. Liver biopsy is normally contraindicated in HHT because of a higher risk of bleeding.

Hepatocellular adenomas and hepatocellular carcinoma were rarely reported[27].

PELIOSIS HEPATIS

Peliosis hepatis is a rare benign vascular condition characterized by sinusoidal dilatation and the presence of blood-filled cystic cavities within the liver parenchyma[28]. The cysts are lined by endothelial cells in the phlebectatic subtype while they lack an endothelial lining in the parenchymal subtype, and the cystic spaces can vary in size from 1 mm to several centimeters[29,30]. The same process may occur in the spleen, bone marrow, lymph nodes, and more rarely in the lungs, stomach, intestine, parathyroid, pancreas, pituitary gland, and kidneys. Peliosis hepatis pathogenesis remains uncertain, being associated with drugs (anabolic steroids, corticosteroids, immunosuppressants, selective estrogen receptor modulators, and androgen), hematological neoplasia and chronic infections (tuberculosis, human immunodeficiency virus)[30,31]. Event most of the patients are asymptomatic and discovered incidentally, some severe cases with liver rupture, portal hypertension and liver insufficiency were reported[30]. PH lesions usually appear as single or multiple areas of varying-sized cysts with hypoattenuation to liver parenchyma on unenhanced CT, hyperintense to liver parenchyma or heterogeneous signal intensity on T2-weighted imaging, and hypointense to liver parenchyma or heterogeneous signal intensity on T1-weighted imaging[32] (Fig. 2).

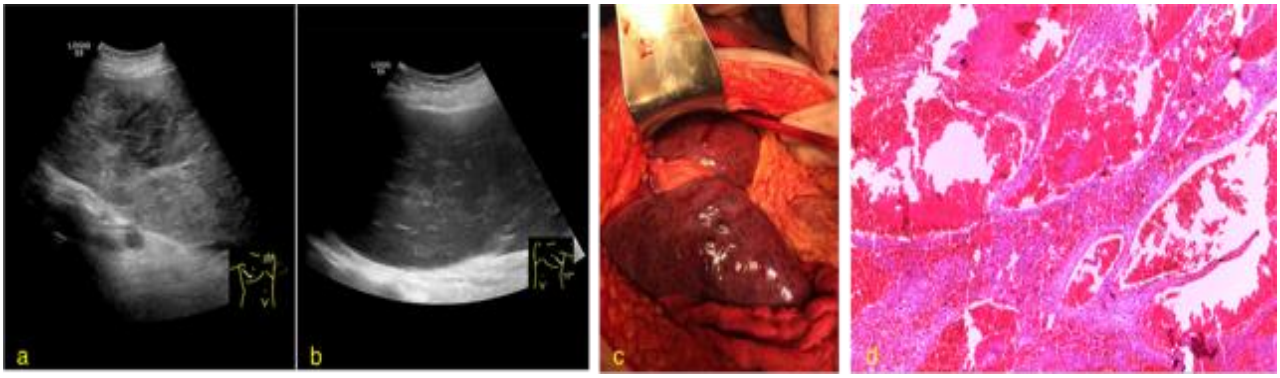

Fig 2. Peliosis hepatis. a) multiple hypoechoic masses in an enlarged liver; b) multiple hypoechoic masses in an enlarged spleen; c) liver: multiple nodules on the surface; d) histology assessment: multiple cavities (corresponding to dilatation of the sinusoids) filled with blood.

SINUSOIDAL OBSTRUCTION SYNDROME

Sinusoidal obstruction syndrome (SOS) is characterized by the damage to small hepatic vessels affecting particularly sinusoidal endothelium. Damaged sinusoids can be associated with a partial or complete occlusion of small hepatic veins, hence the previous denomination of hepatic veno-occlusive disease (VOD). Exposure to certain exogenous toxins, especially oxaliplatin, appears to be specific to this condition and is frequently included in its definition[33].

This disorder is characterized by a spectrum of pathologic changes, from sinusoidal dilation, peri-sinusoidal haemorrhage, peliosis and nodular regenerative hyperplasia[34,35]. Very rarely liver nodules mimicking liver metastases have been described[35,36], especially in colorectal cancer patients treated with oxaliplatin.

FONTAN SURGICAL PROCEDURE

Liver nodules have also been described in patients after the Fontan surgical procedure (anastomosis between the vena cava or right atrium and the pulmonary arteries and causing chronic passive liver congestion). Liver pathology varies from passive venous congestion to hepatic ischemia and chronic congestive hepatopathy (previously known as cardiac cirrhosis)[37]. FNH-like lesions were reported in patients with high Fontan pressures[38]. Also, HCC cases might be present after longer period[39]. Differential diagnosis between FNH-like lesions and HCC might be difficult, especially in the presence of cirrhosis.

CONCLUSIONS

Even if rare, vascular disorders of the liver should be considered in the clinical scenario of a

young person with multiple benign nodules. Many vascular disorders of the liver are associated with benign hepatocellular tumours, secondary to an increase in the arterial flow. FNH-like lesions are the most common, as this nodule is a hyperplastic response of the liver parenchyma to an increase or a disturbance of hepatic blood flow. Hepatocellular adenoma-like or even hepatocellular carcinoma were described, requiring multiple serological, imaging and even biopsies assessments. Imaging, and especially contrast enhanced ultrasound and MRI with specific contrast agents are depicting with high accuracy a mild hyperenhancement of these nodules on the vascular background of the liver characterized by an increase arterial outflow.

REFERENCES

1. DeLeve LD, Valla DC, Garcia-Tsao G; American Association for the Study Liver Diseases. Vascular disorders of the liver. Hepatology. 2009 May;49(5):1729-64. doi: 10.1002/hep.22772. PMID: 19399912; PMCID: PMC6697263.
2. Willington AJ, Tripathi D. Current concepts in the management of non-cirrhotic non-malignant portal vein thrombosis. World J Hepatol. 2024 May 27;16(5):751-765. doi: 10.4254/wjh.v16.i5.751. PMID: 38818283; PMCID: PMC11135268.
3. Vilgrain V, Paradis V, Van Wettere M, Valla D, Ronot M, Rautou PE. Benign and malignant hepatocellular lesions in patients with vascular liver diseases. Abdom Radiol (NY). 2018 Aug;43(8):1968-1977. doi: 10.1007/s00261-018-1502-7. PMID: 29460040.
4. Wanless IR, Mawdsley C, Adams R. On the pathogenesis of focal nodular hyperplasia of the liver. Hepatology. 1985 Nov-Dec;5(6):1194-200. doi: 10.1002/hep.1840050622. PMID: 4065824.
5. Kumagai H, Masuda T, Oikawa H, Endo K, Endo M, Takano T. Focal nodular hyperplasia of the liver: direct evidence of circulatory disturbances. J Gastroenterol Hepatol. 2000 Nov;15(11):1344-7. PMID: 11129233.
6. Ogren M, Bergqvist D, Björck M, Acosta S, Eriksson H, Sternby NH. Portal vein thrombosis: prevalence, patient characteristics and lifetime risk: a population study based on 23,796 consecutive autopsies. World J Gastroenterol. 2006;12:2115–2119. doi: 10.3748/wjg.v12.i13.2115. [DOI] [PMC free article] [PubMed] [Google Scholar]
7. 7.Ageno W, Dentali F, Pomero F, Fenoglio L, Squizzato A, Pagani G, Re R, Bonzini M. Incidence rates and case fatality rates of portal vein thrombosis and Budd-Chiari Syndrome. Thromb Haemost. 2017;117:794–800. doi: 10.1160/TH16-10-0781. [DOI] [PubMed] [Google Scholar]
8. Bureau C, Péron JM, Sirach E, Selves J, Otal P, Vinel JP. Liver nodules resembling focal nodular hyperplasia after portal vein thrombosis. J Hepatol. 2004 Sep;41(3):499-500. doi: 10.1016/j.jhep.2004.04.025. PMID: 15336457.
9. Marin D, Galluzzo A, Plessier A, Brancatelli G, Valla D, Vilgrain V. Focal nodular hyperplasia-like lesions in patients with cavernous transformation of the portal vein: prevalence, MR findings and natural history. Eur Radiol. 2011 Oct;21(10):2074-82. doi: 10.1007/s00330-011-2161-z. Epub 2011 Jun 28. PMID: 21710271.
10. Amarapurkar P, Bhatt N, Patel N, Amarapurkar D. Primary extrahepatic portal vein obstruction in adults: a single center experience. Indian J Gastroenterol. 2014 Jan;33(1):19-22. doi: 10.1007/s12664-013-0368-4. Epub 2013 Sep 6. PMID: 24222368.
11. LeGout JD, Bolan CW, Bowman AW, Caserta MP, Chen FK, Cox KL, Sanyal R, Toskich BB, Lewis JT, Alexander LF. Focal Nodular Hyperplasia and Focal Nodular Hyperplasia-like Lesions. Radiographics. 2022 Jul-Aug;42(4):1043–1061. doi: 10.1148/rg.210156. Epub 2022 Jun 10. PMID: 35687520.
12. Brancatelli G, Vilgrain V, Federle MP, Hakime A, Lagalla R, Iannaccone R, Valla D. Budd-Chiari syndrome: spectrum of imaging findings. AJR Am J Roentgenol. 2007 Feb;188(2):W168-76. doi: 10.2214/AJR.05.0168. PMID: 17242224.
13. Vilgrain V, Lewin M, Vons C, Denys A, Valla D, Flejou JF, Belghiti J, Menu Y. Hepatic nodules in Budd-Chiari syndrome: imaging features. Radiology. 1999 Feb;210(2):443-50. doi: 10.1148/radiology.210.2.r99fe13443. PMID: 10207428.
14. Maetani Y, Itoh K, Egawa H, et al. (2002) Benign hepatic nodules in Budd-Chiari syndrome: radiologic-pathologic correlation with emphasis on the central scar. AJR Am J Roentgenol 178:869–875
15. Sempoux C, Paradis V, Komuta M, Wee A, Calderaro J, Balabaud C, Quaglia A, Bioulac-Sage P. Hepatocellular nodules expressing markers of hepatocellular adenomas in Budd-Chiari syndrome and other rare hepatic vascular disorders. J Hepatol. 2015 Nov;63(5):1173-80. doi: 10.1016/j.jhep.2015.06.017. Epub 2015 Jun 25. PMID: 26119687.
16. Moucari R, Rautou PE, Cazals-Hatem D, Geara A, Bureau C, Consigny Y, Francoz C, Denninger MH, Vilgrain V, Belghiti J, Durand F, Valla D, Plessier A. Hepatocellular carcinoma in Budd-Chiari syndrome: characteristics and risk factors. Gut. 2008 Jun;57(6):828-35. doi: 10.1136/gut.2007.139477. Epub 2008 Jan 24. PMID: 18218675.
17. Paul SB, Shalimar, Sreenivas V, Gamanagatti SR, Sharma H, Dhamija E, Acharya SK. Incidence and risk factors of hepatocellular carcinoma in patients with hepatic venous outflow tract obstruction. Aliment Pharmacol Ther. 2015 May;41(10):961-71. doi: 10.1111/apt.13173. Epub 2015 Mar 23. PMID: 25809735.
18. Zhang R, Qin S, Zhou Y, Song Y, Sun L. Comparison of imaging characteristics between hepatic benign regenerative nodules and hepatocellular carcinomas associated with Budd-Chiari syndrome by contrast enhanced ultrasound. Eur J Radiol. 2012 Nov;81(11):2984-9. doi: 10.1016/j.ejrad.2012.01.023. Epub 2012 Feb 15. PMID: 22341409.
19. McLin VA, Franchi-Abella S, Brütsch T, Bahadori A, Casotti V, de Ville de Goyet J, Dumery G, Gonzales E, Guérin F, Hascoet S, Heaton N, Kuhlmann B, Lador F, Lambert V, Marra P, Plessier A, Quaglia A, Rougemont AL, Savale L, Sarma MS, Sitbon O, Superina RA, Uchida H, van Albada M, van der Doef HPJ, Vilgrain V, Wacker J, Zwaveling N, Debray D, Wildhaber BE. Expert management of congenital portosystemic shunts and their complications. JHEP Rep. 2023 Oct 20;6(1):100933. doi: 10.1016/j.jhepr.2023.100933. Erratum in: JHEP Rep. 2024

Jan 30;6(3):101024. doi: 10.1016/j.jhepr.2024.101024. PMID: 38234409; PMCID: PMC10792643.
20. Pupulim LF, Vullierme MP, Paradis V, Valla D, Terraz S, Vilgrain V. Congenital portosystemic shunts associated with liver tumours. Clin Radiol. 2013 Jul;68(7):e362-9. doi: 10.1016/j.crad.2013.01.024. Epub 2013 Mar 26. PMID: 23537576.
21. Franchi-Abella S, Branchereau S, Lambert V, Fabre M, Steimberg C, Losay J, Riou JY, Pariente D, Gauthier F, Jacquemin E, Bernard O. Complications of congenital portosystemic shunts in children: therapeutic options and outcomes. J Pediatr Gastroenterol Nutr. 2010 Sep;51(3):322-30. doi: 10.1097/MPG.0b013e3181d9cb92. PMID: 20601902.
22. Lisovsky M, Konstas AA, Misdraji J. Congenital extrahepatic portosystemic shunts (Abernethy malformation): a histopathologic evaluation. Am J Surg Pathol. 2011 Sep;35(9):1381-90. doi: 10.1097/PAS.0b013e3182230ce4. PMID: 21836489.
23. Sabbà C, Pompili M. Review article: the hepatic manifestations of hereditary haemorrhagic telangiectasia. Aliment Pharmacol Ther. 2008 Sep 1;28(5):523-33. doi: 10.1111/j.1365-2036.2008.03775.x. Epub 2008 Jun 20. PMID: 18573106.
24. Wanless IR, Gryfe A. Nodular transformation of the liver in hereditary hemorrhagic telangiectasia. Arch Pathol Lab Med. 1986 Apr;110(4):331-5. PMID: 3754122.
25. Brenard R, Chapaux X, Deltenre P, Henrion J, De Maeght S, Horsmans Y, Borbath I, Leenaerts A, Van Cauter J, Francque S, Sersté T, Moreno C, Orlent H, Mengeot P, Lerut J, Sempoux C. Large spectrum of liver vascular lesions including high prevalence of focal nodular hyperplasia in patients with hereditary haemorrhagic telangiectasia: the Belgian Registry based on 30 patients. Eur J Gastroenterol Hepatol. 2010 Oct;22(10):1253-9. doi: 10.1097/MEG.0b013e32833ac4a1. PMID: 20631626.
26. Buscarini E, Danesino C, Plauchu H, de Fazio C, Olivieri C, Brambilla G, Menozzi F, Reduzzi L, Blotta P, Gazzaniga P, Pagella F, Grosso M, Pongiglione G, Cappiello J, Zambelli A. High prevalence of hepatic focal nodular hyperplasia in subjects with hereditary hemorrhagic telangiectasia. Ultrasound Med Biol. 2004 Sep;30(9):1089-97. doi: 10.1016/j.ultrasmedbio.2004.08.004. PMID: 15550313.
27. Lee JH, Lee YS, Kim PN, Lee BH, Kim GW, Yoo HW, Heo NY, Lim YS, Lee HC, Chung YH, Suh DJ. Osler-Weber-Rendu disease presenting with hepatocellular carcinoma: radiologic and genetic findings. Korean J Hepatol. 2011 Dec;17(4):313-8. doi: 10.3350/kjhep.2011.17.4.313. PMID: 22310796; PMCID: PMC3304659.
28. Yamada Y, Kurokawa R, Kurokawa M, Tsujimoto R, Shimura A, Maki H, Kondo A, Abe O. Longitudinal CT, MRI, and 18F-FDG PET/CT Imaging Findings of Peliosis Hepatis: A Case Report. Cureus. 2024 Jun 23;16(6):e62997. doi: 10.7759/cureus.62997. PMID: 39050295; PMCID: PMC11266828.
29. Calistri L, Nardi C, Rastrelli V, Maraghelli D, Grazioli L, Messerini L, Colagrande S. MRI of Peliosis Hepatis: A Case Series Presentation With a 2022 Systematic Literature Update. J Magn Reson Imaging. 2023 Nov;58(5):1386-1405. doi: 10.1002/jmri.28673. Epub 2023 Mar 29. PMID: 36988385.
30. Crocetti D, Palmieri A, Pedullà G, Pasta V, D'Orazi V, Grazi GL. Peliosis hepatis: Personal experience and literature review. World J Gastroenterol. 2015 Dec 14;21(46):13188-94. doi: 10.3748/wjg.v21.i46.13188. PMID: 26675327; PMCID: PMC4674738.
31. Yu CY, Chang LC, Chen LW, Lee TS, Chien RN, Hsieh MF, Chiang KC. Peliosis hepatis complicated by portal hypertension following renal transplantation. World J Gastroenterol. 2014 Mar 7;20(9):2420-5. doi: 10.3748/wjg.v20.i9.2420. PMID: 24605041; PMCID: PMC3942847.
32. Iannaccone R, Federle MP, Brancatelli G, Matsui O, Fishman EK, Narra VR, Grazioli L, McCarthy SM, Piacentini F, Maruzzelli L, Passariello R, Vilgrain V. Peliosis hepatis: spectrum of imaging findings. AJR Am J Roentgenol. 2006 Jul;187(1):W43-52. doi: 10.2214/AJR.05.0167. PMID: 16794138.
33. Valla DC, Cazals-Hatem D. Sinusoidal obstruction syndrome. Clin Res Hepatol Gastroenterol. 2016 Sep;40(4):378-85. doi: 10.1016/j.clinre.2016.01.006. Epub 2016 Mar 30. PMID: 27038846.
34. Morris-Stiff G, White AD, Gomez D, Cameron IC, Farid S, Toogood GJ, Lodge JP, Prasad KR. Nodular regenerative hyperplasia (NRH) complicating oxaliplatin chemotherapy in patients undergoing resection of colorectal liver metastases. Eur J Surg Oncol. 2014 Aug;40(8):1016-20. doi: 10.1016/j.ejso.2013.09.015. Epub 2013 Nov 1. PMID: 24370284.
35. Alexandrino H, Oliveira D, Cipriano MA, Ferreira L, Tralhão JG, Castro E Sousa F. Oxaliplatin toxicity presenting as a liver nodule – case report. BMC Cancer. 2015 Apr 10;15:247. doi: 10.1186/s12885-015-1247-4. PMID: 25886004; PMCID: PMC4393578.
36. Gupta V, Chopde A, Patkar S, Deodhar K, Goel M. Oxaliplatin-Induced Sinusoidal Obstruction Syndrome Masquerading as Colorectal Liver Metastasis: A Case Report. J Gastrointest Cancer. 2023 Jun;54(2):682-686. doi: 10.1007/s12029-022-00835-x. Epub 2022 Jun 6. PMID: 35666356.
37. Hilscher MB, Johnson JN, Cetta F, et al. Surveillance for liver complications after the Fontan procedure. Congenit Heart Dis 2017;12:124–132
38. Bryant T, Ahmad Z, Millward-Sadler H, et al. Arterialised hepatic nodules in the Fontan circulation: hepatico-cardiac interactions. Int J Cardiol 2011;151:268–272
39. Josephus Jitta D, Wagenaar LJ, Mulder BJ, et al. Three cases of hepatocellular carcinoma in Fontan patients: Review of the literature and suggestions for hepatic screening. Int J Cardiol 2016;206:21–26