PORTAL CHOLANGIOPATHY:

A PICTORIAL REVIEW

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LEARNING OBJETIVES

- To review the pathophysiology, imagiologic findings, main differential diagnosis and management of portal cholangiopathy (PC).

BACKGROUND

- Portal cholangopathy is a rare disorder that may complicate extra-portal venous obstruction and portal cavernoma formation, the enlargement of biliary veins leading to **compression** and **deformation of large bile ducts**.

- Biliary abnormalities occur as a consequence of **extrinsic compression** by **collateral vessels** and **ischemic injury** due to venous thrombosis.

- Portal cholangiopathy, symptomatic or not, as been described in **70-100%** of patients with extrahepatic obstruction of the portal vein, and is much less common in cirrhotic patients.

BACKGROUND

- The **portal cavernoma** is composed by two venous systems:
  - paracholedochal plexus of Petren (parallel to the ductal wall)
  - epicholedochal venous plexus of Saint (surface of the bile duct)

- In some patients, the cavernous transformation may turn into a "mass-like" solid structure with multiple indistinguishable collateral veins with consequent encasement and compromise of the biliary ducts.

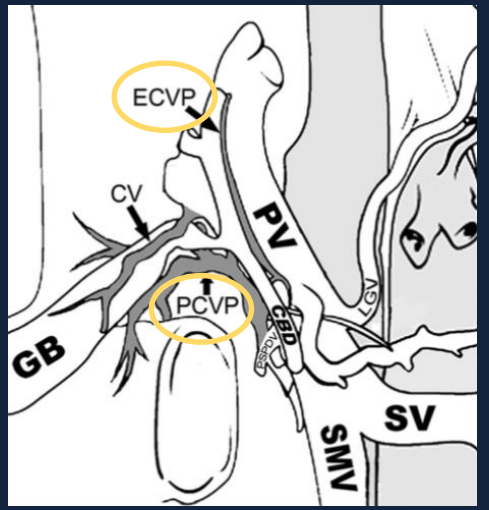

Fig.1- Diagram showing the venous systems of portal cavernoma: ECVP- epicholedochal venous plexus; PCVP- paracholedochal venous plexus; PV- portal vein; SV- splenic vein; SMV- superior mesenteric vein; GB- gallbladder; CV- cystic vein; CBD- common bile duct; LGV- left gastric vein; PSPDV- posterior superior pancreaticoduodenal vein.
(References: Shin SM, Kim S, Lee JW, et al. Biliary abnormalities associated with portal biliopathy: evaluation on MR cholangiography. AJR Am J Roentgenol 2007;188:W341-7)

BACKGROUND

- Most of the patients are asymptomatic and biliary changes are frequently an incidental imaging finding.

- Patients who have long standing disease usually have cholangiography abnormalities with episodes of biliary pain, cholangitis and cholestasis.

- Cholelithiasis and hepatolithiasis are found in **5-20%** of the patients with PC and are probably secondary to biliary stasis.

- Abnormal laboratory tests, with total bilirubin and alkaline phosphatase levels elevated are detected in **40-80%** of the cases.

BACKGROUND

- PC may mimic other diagnosis as **cholangiocarcinoma, sclerosing cholangitis**, **lymphoplasmocytic cholangiopathy, choledocholithiasis** or **extrinsic compression of the bile duct by metastatic adenopathies.**

- Although ultrasound imaging with colour Doppler may show biliary and gallbladder abnormalities associated with extra-portal venous obstruction, cholangiography images remain essential to **confirm the diagnosis of PC** in the majority of the cases.

- The use of appropriated imaging modalities as magnetic resonance imaging (MRI) may **avoid misdiagnosis** and can be used for **differential diagnosis**.

BACKGROUND

Although endoscopic retrograde cholangiopancreatography (ERCP) is considered the gold standard modality to define biliary changes

Invasive and prone to complications!

PC is a slowly progressive pathology …

Increasingly being **replaced** by MR cholangiography

Reserving ERCP for advanced stages as **treatment** or diagnosis of **misleading cases**.

IMAGING FINDINGS OR PROCEDURE DETAILS

- We retrospectively selected 7 patients with portal cholangiopathy of a total of 21 patients with portal cavernoma from our institution, between the years of 2011 to 2016.
- The purpose of this review was to illustrate the imaging findings and complications of portal cavernoma and PC on US imaging with color Doppler, computed tomography (CT) and MR cholangiography.

US DOPPLER

- First imaging modality in the diagnose of porto-portal collaterals vessels secondary to extra-hepatic portal vein obstruction.

- A decrease in portal vein diameter, increased echogenicity of the tissue in the hilum, and multiple anechoic tubular structures with Doppler flow, are direct indicators of cavernomatous degeneration.

- Usually, common biliary duct indentation are seen secondary to extrinsic compression by the enlarged paracholedochal venous plexus.

COMPUTED TOMOGRAPHY (CT)

- The signs of collateral circulation and the compressive effect of cavernous transformation of portal vein around the bile duct are well documented in CT images and its post-processing tools.

- The contrast-enhanced CT in portal phase is useful to evaluate the extension of **portal cavernoma**, the presence of **choledocal** and **intrahepatic varices**, as well the presence of **gallbladder varices**.

- CT images can also show **secondary biliary duct dilatation** caused by the cavernoma, excluding other causes of obstruction such as **biliary lithiasis**, **cholangiocarcinoma** or **extrinsic compression by masses**.

MR CHOLANGIOGRAPHY

- The noninvasive imaging modality of choice to **correct diagnose** portal cholangiopathy.

- **Adequate characterization** of the intra and extrahepatic bile duct, with a comparable capacity to endoscopic retrograde cholangiopancreatography (ERCP) to evaluate **morphological biliary changes**.

- Gadolinium-enhanced magnetic resonance imaging illustrate the **vascular abnormality** in the upper abdomen (or a mass-like solid tumour when collateral vessels are not individualized), as the **biliary enlargement** consequent to portal cavernoma.

MR CHOLANGIOGRAPHY

- Two big advantage of MR cholangiography are the the lack of ionizing radiation and the capability to identify all the morphologic changes of the biliary tree.

- The most common abnormalities of PC include irregular strictures of the extra and intrahepatic bile ducts, upstream dilatation, angulation and bile duct parietal irregularities.

- **Biliary stasis** adjacent to the dominant strictures can predispose to lithiasis.

MR CHOLANGIOGRAPHY

- MR imaging is extremely helpful in recognizing the cause of bile duct obstruction, **excluding malignant diagnosis** as c**holangiocarcinoma**.

- Determining the exact site of stenosis helps to guide **therapeutic interventions**.

- Noninvasive follow-up of these group of patients.

PRINCIPAL IMAGING FEATURES	
US	• Serpentine anechoic vessels with hepatopetal flow • Luminal narrowing of the common biliary duct • Biliary duct dilatation proximal to the focal area of stenosis • Intra-hepatic dilatation • Gallbladder varices
CT	• Intra and extra-hepatic portions of the parabiliary and peribiliary plexuses • Gallbladder varices • Mass-like tumour vs. serpiginous • Dilatation of the intra and extrahepatic bile ducts
MR COLANGIOGRAPHY	• Intra and extra-hepatic portions of the parabiliary and peribiliary plexuses • Gallbladder varices • Mass-like tumour vs. serpiginous • Irregular strictures of the extra and intrahepatic bile ducts • Angulations and a wavy appearance of the bile ducts • Dominant stenosis segments

Table 1- Principal imaging features associated with portal cholangiopathy.

Portal cholangiopathy: Morphological classification

Type	Findings
I	involvement of extrahepatic ducts (a)
II	involvement of intrahepatic bile ducts only (b)
IIIa	extrahepatic bile duct and unilateral intrahepatic bile duct involvement (c)
IIIb	extrahepatic bile duct and bilateral intrahepatic bile duct involvement (d)

Table 2- Classification system for portal cholangiopathy associated with the location of biliary abnormalities.

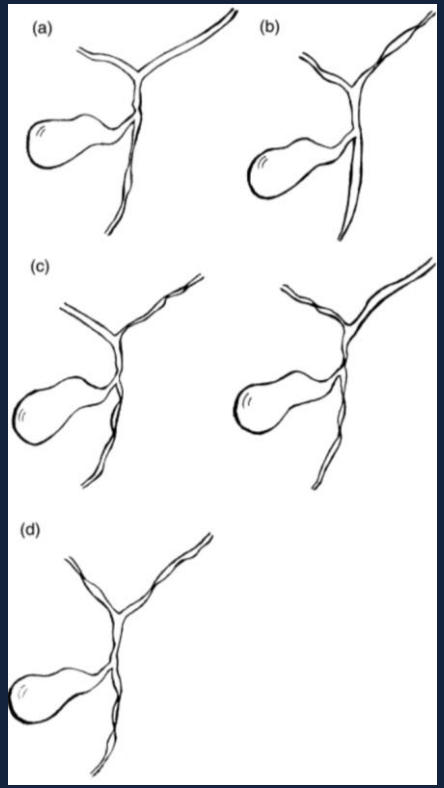

Fig.2- Graphic representation of changes in portal cholangiopathy based on the location and extent of cholangiographic abnormalities.

(References: **Chandra R, Kapoor D, Tharakan A et al. Portal biliopathy. J Gastroenterol Hepatol 2001;16:1086-92.)**

CASE 1

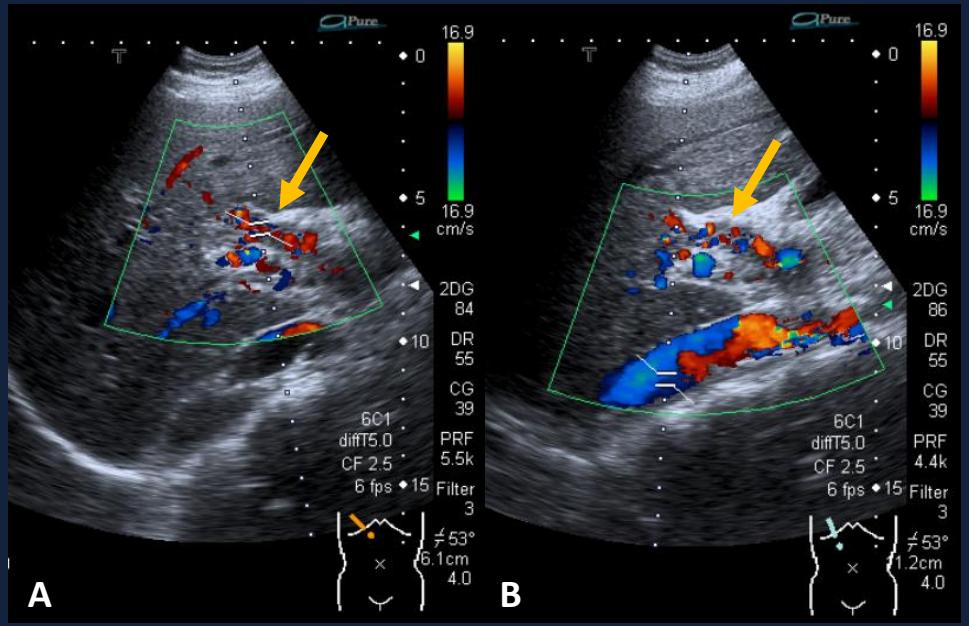

Fig.3- 36-year old female with a known hypercoagulability condition (JAK2+) and portal cavernoma. (A) and (B) Doppler-US images showing porto-portal collaterals corresponding to cavernous transformation of the portal vein (arrow).

CASE 1

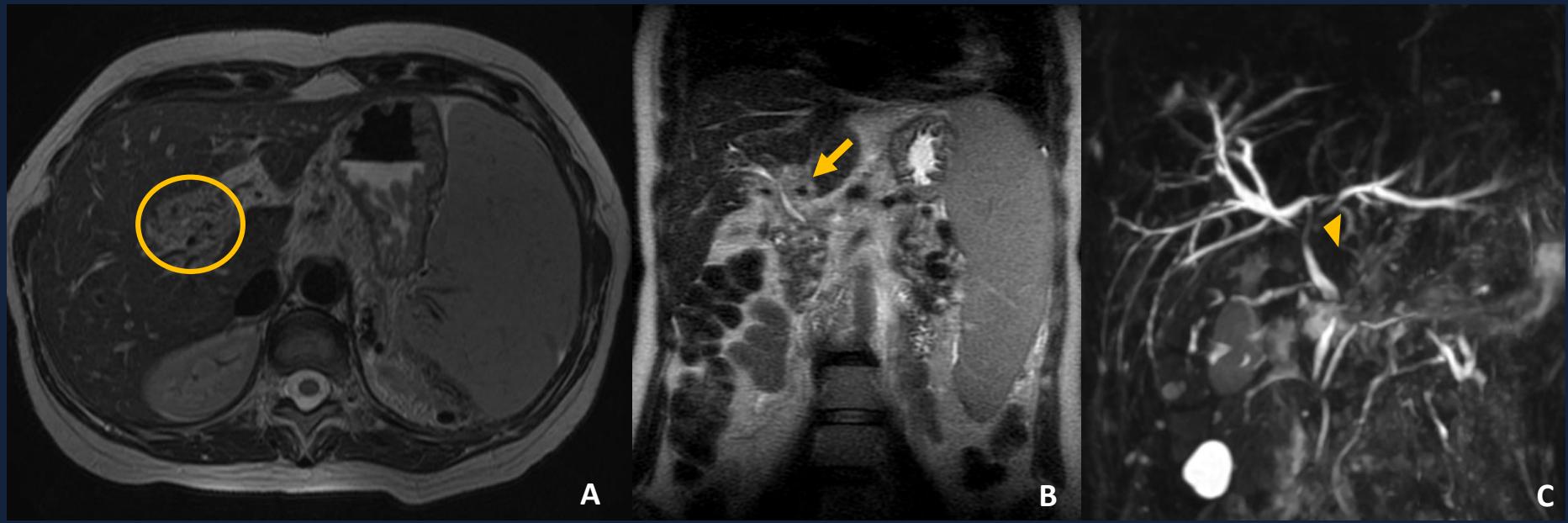

Fig.4- 36-year old female with a known hypercoagulability condition (JAK2+) and portal cavernoma. T2-weighted axial (A) and coronal (B) MR images, show a serpiginous cavernoma (circle) with consequent extrinsic compression of common biliary duct (arrow). MRCP (C) images demonstrate stenosis of the main biliary duct at the hilar level (arrowhead) with consequent right and left biliary branches dilatation.

CASE 2

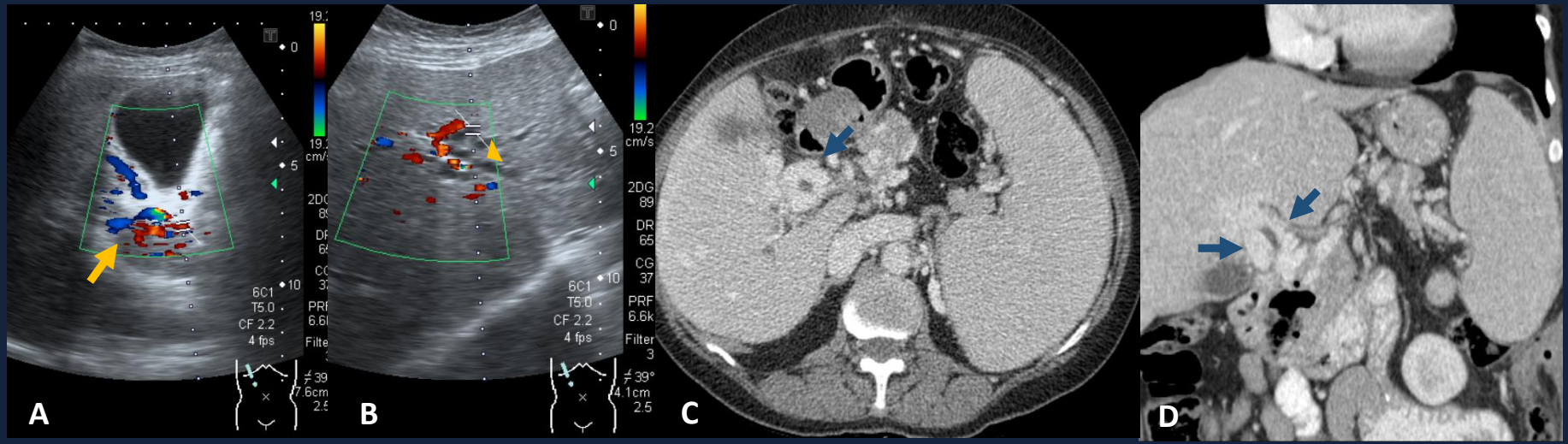

Fig.5- 63-year old female patient with portal cavernoma. (A) and (B) Doppler-US images revealing portal cavernoma (arrow) with associated dilatation of the intra-hepatic bile duct branches (arrowhead). (C) Axial and (D) coronal contrast-enhanced CT images demonstrating a tumor-like solid cavernoma (blue arrows) involving the common billiary duct.

CASE 2

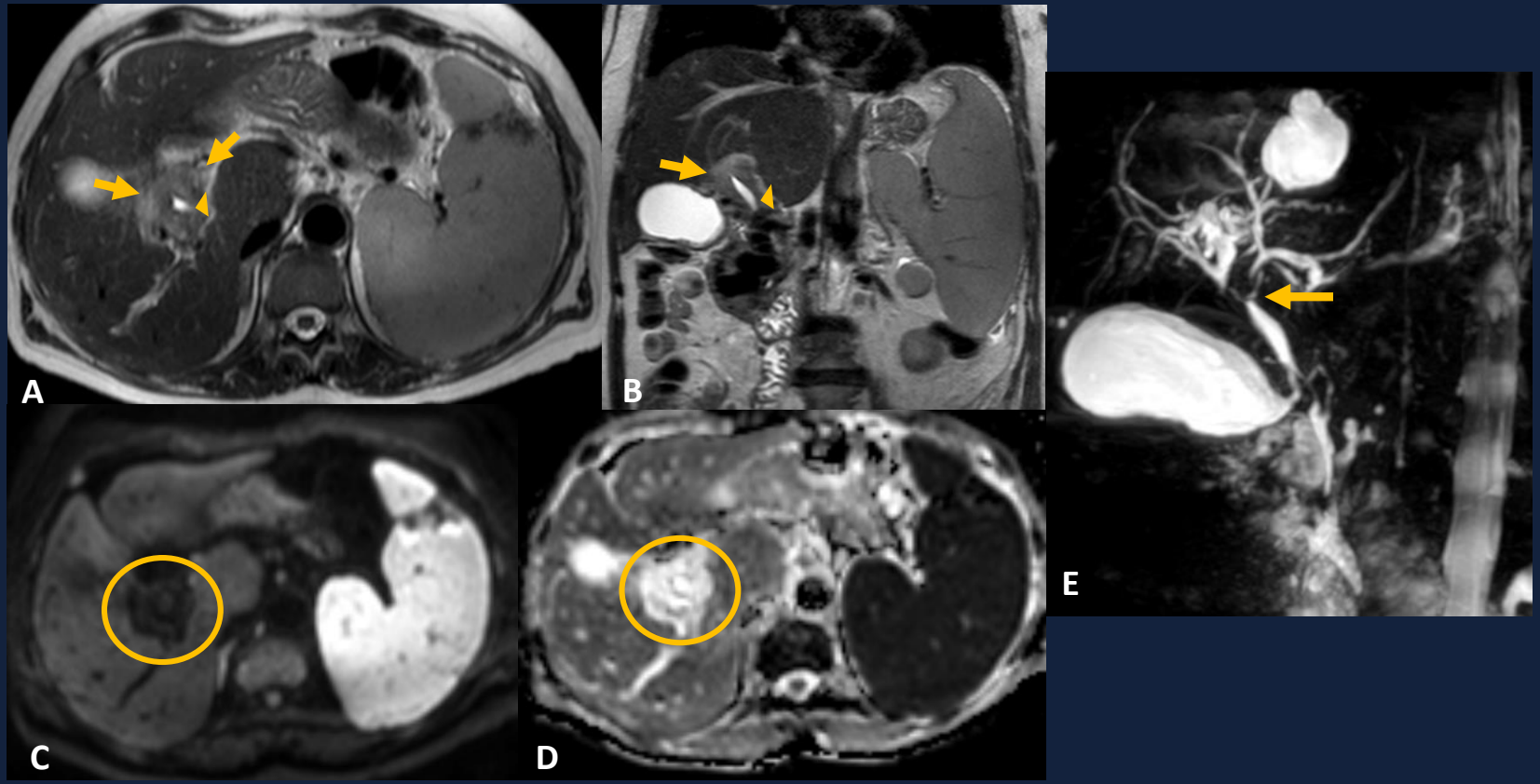

Fig.6 – 63-year old female patient with portal cavernoma. (A) Axial and (B) Coronal T2-weighted images showing a hyperintense mass-like cavernoma (arrows) with encasement of the common billiary duct (arrowhead). (C) DWI b1000 images reveal a hypointense tumor-like cavernoma (circle) that shows no restriction on ADC map (D) (circle). (E) MRCP demonstrate stenosis of de main billiary duct and hepatic duct bifurcation (arrow) due to extrinsic compression from enlarged vein with upstream dilatation.

CASE 3

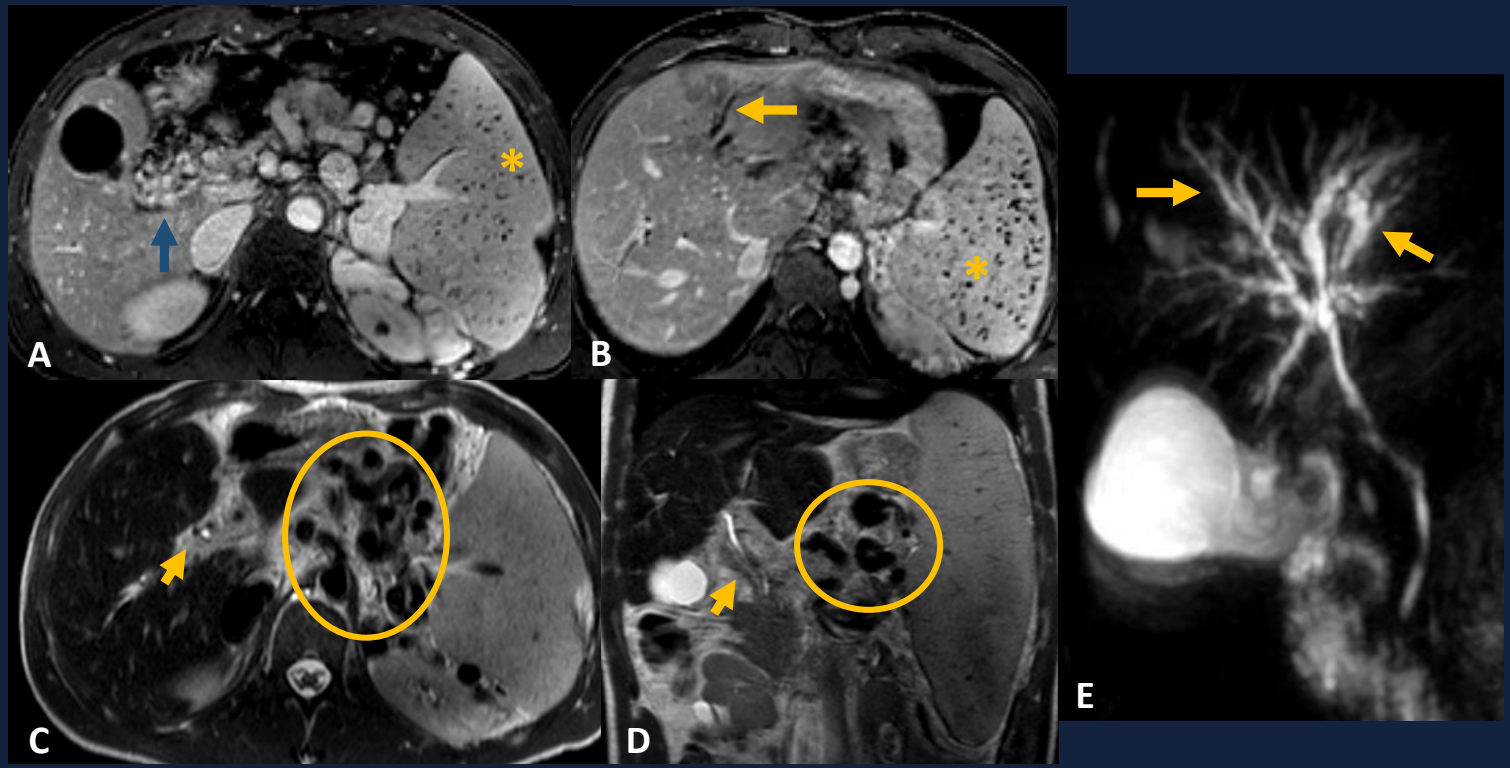

Fig.7 – 29-year old male patient with portal cavernoma. (A) and (B) Axial contrast-enhanced T1-weighted MR images demonstrating serpiginous portal cavernoma (blue arrow) with consequent intra-hepatic bile ducts dilatation (yellow arrow). Additionally, splenomegaly and siderotic foci (*) are seen within the spleen (Gamna-Gandy bodies) in association with portal hipertension. (C) Axial and (D) coronal T2-weighted MR images demonstrate the portal cavernoma compressing the main bile duct (arrow). Splenic varices (circle) are also visible due to elevated pressure in the portal venous system. (E) MRCP image demonstrate multiple irregularities of intra-hepatic bile ducts (arrows).

CASE 4

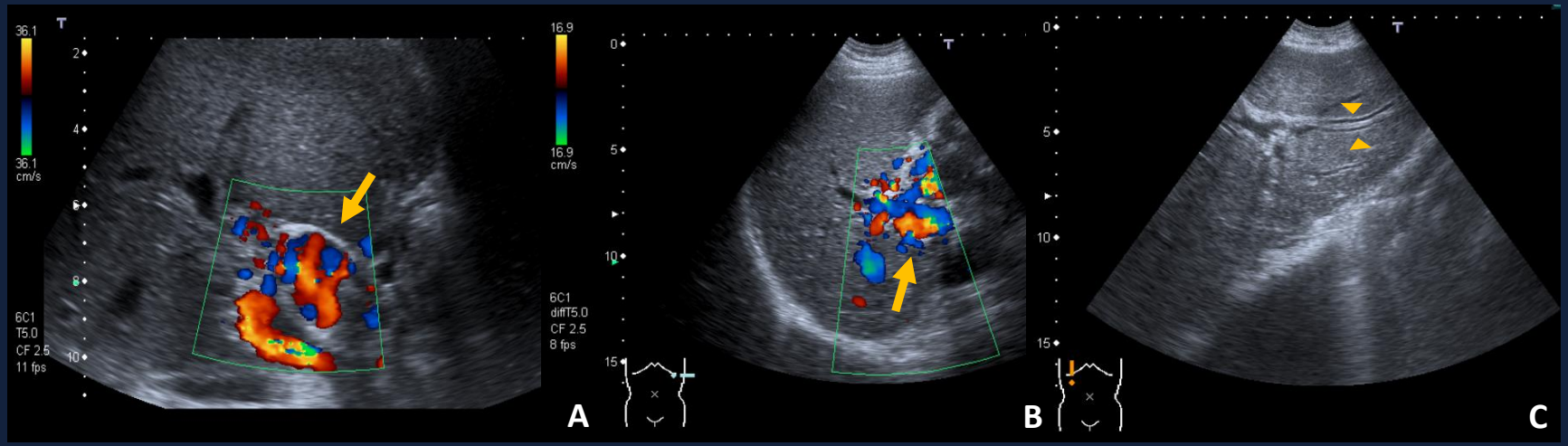

Fig.8- 62-year old male patient with portal cavernoma. (A) and (B) Doppler-US images demonstrating portal cavernoma transformation (arrow) due to extra-hepatic portal vein obstruction. (C) US image show intra-hepatic bile duct dilatation (arrowhead) due to extrinsic compression from paracholedocal veins of the portal cavernoma.

CASE 4

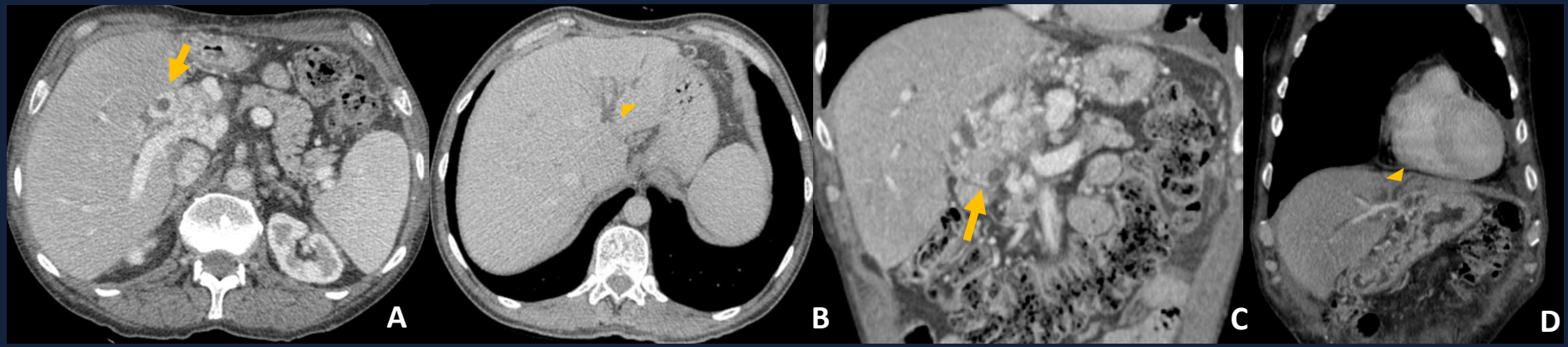

Fig.9- 62-year old male patient with portal cavernoma. (A) and (B) Axial contrast-enhanced CT images showing portal cavernoma (arrow) surrounding the common bile duct with consequente intra-hepatic bile ducts dillatation, especially the branches of the left hepatic lobe (arrowhead). (C) and (D) Coronal contrast-enhanced CT images confirming the existence of portal cavernomatous transformation (arrow) with upstream dilatation (arrowhead).

CASE 5

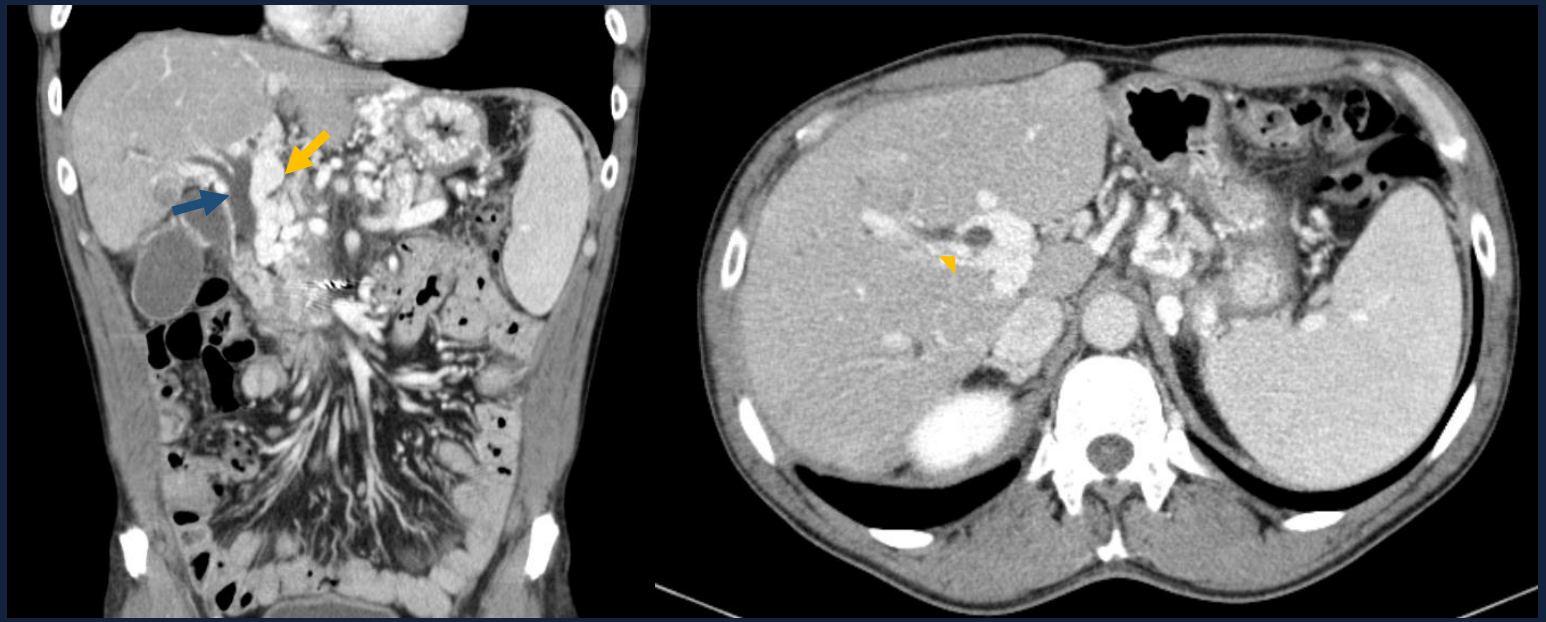

Fig. 10- 56-year male patient with alcoholic and HCV-related cirrhosis, with a known portal cavernoma. (A) Coronal and (B) axial contrast-enhanced CT images show marked dilatation of the common bile duct and hilar branches (blue arrows) secondary to a serpiginous cavernoma. Enlarged paracholedocal (yellow arrow) and epicholedocal veins (arrowhead) are demonstrated as the two venous plexus composing the cavernoma.

CASE 5

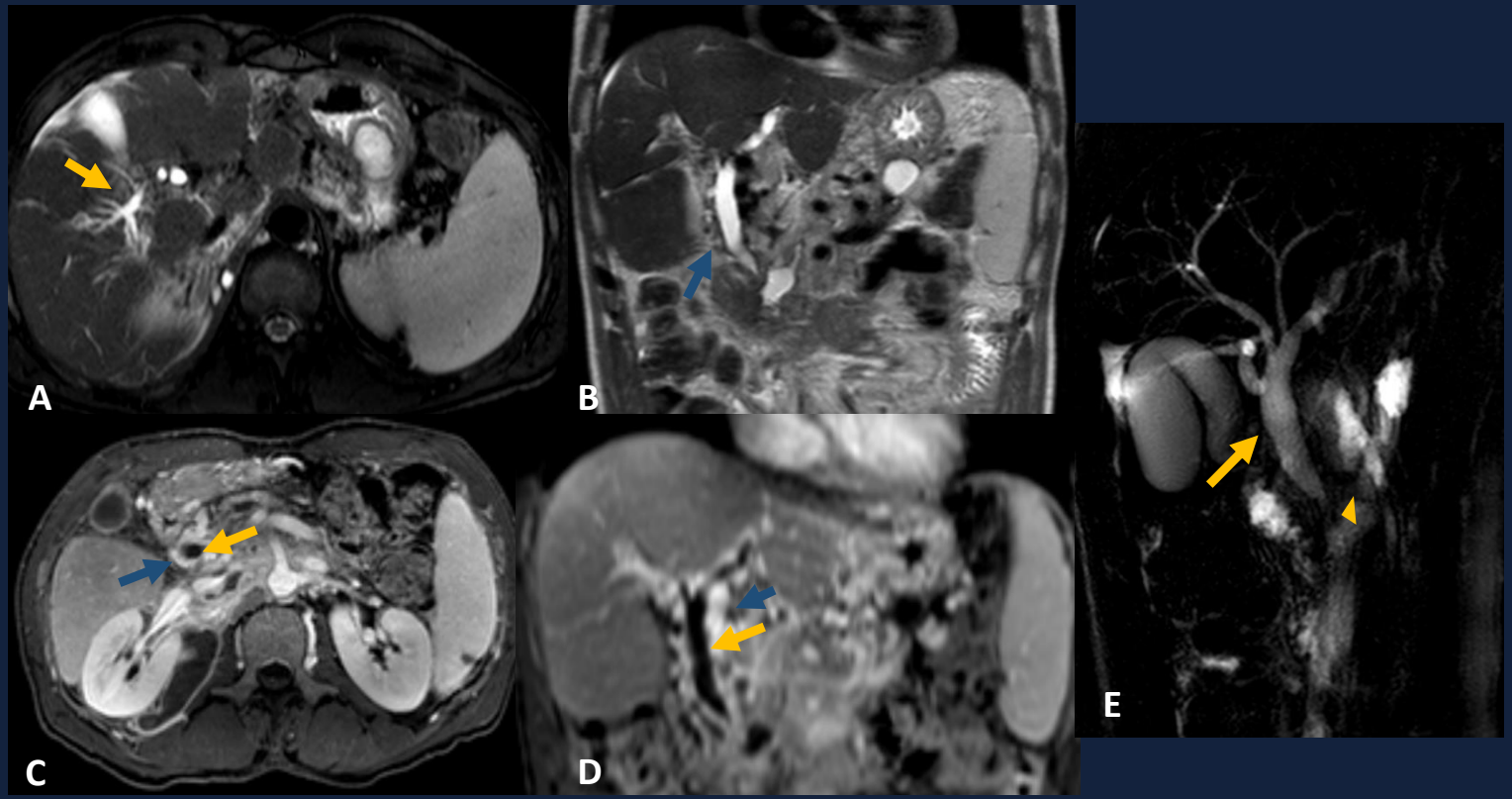

Fig. 11- 56-year male patient with alcoholic and HCV-related cirrhosis, with a known portal cavernoma. (A) T2-weighted FS axial (B) and T2-weighted coronal MR images, demonstrating intra-hepatic biliary branches (yellow arrow) and common bile duct dilatation (blue arrow). (C) Axial and (D) coronal contrast-enhanced T1-weighted MR images showing a dilated hypointense common bile duct (yellow arrow) surrounded by an enhanced serpiginous portal cavernoma (blue arrow). (E) MRCP image displaying a hole common bile duct dilatation (arrow) and a narrowing at its lower end (arrowhead).

CASE 6

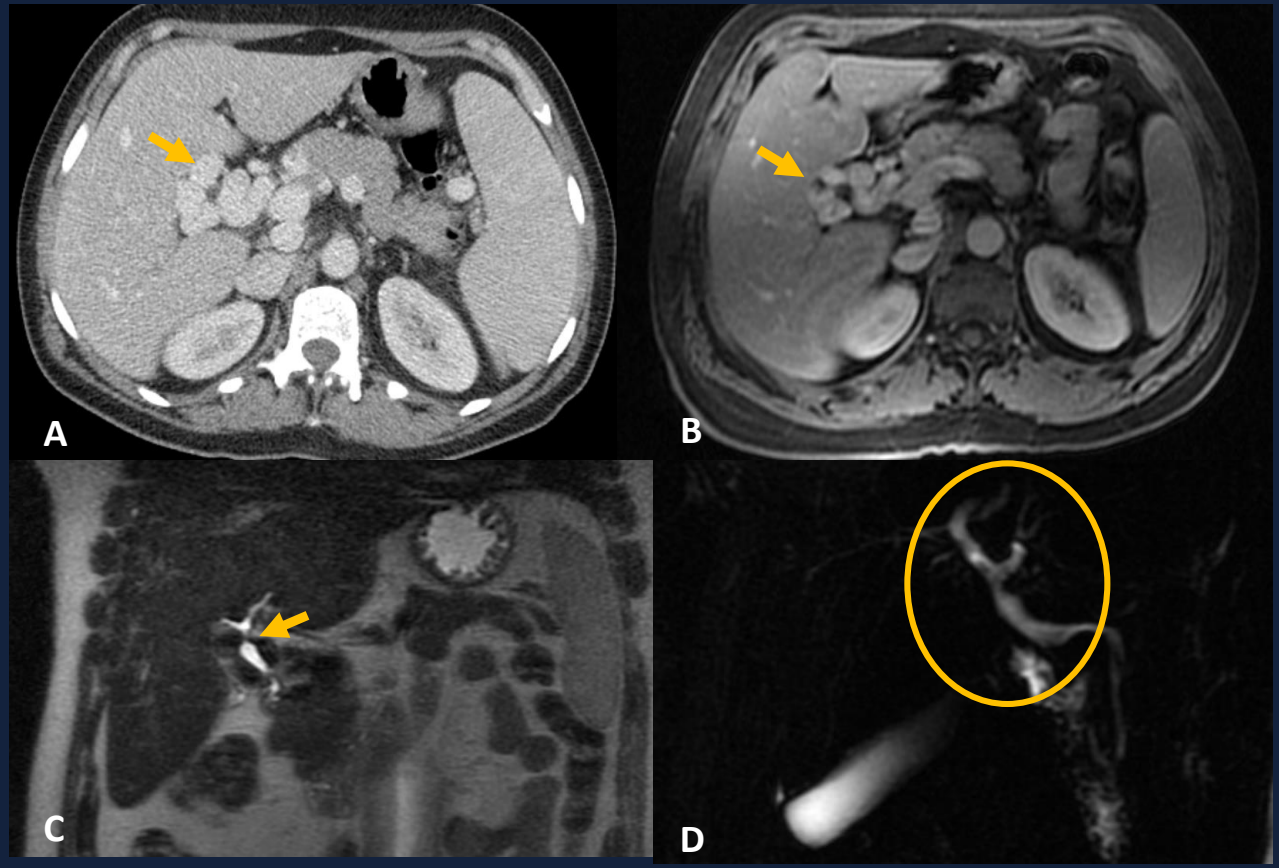

Fig. 12- 49-year female patient with Sneddon’s syndrome and cavernous transformation of the portal vein . (A) Axial contrast-enhanced CT and (B) axial contrast-enhanced T1-weighted MR images show a notable serpiginous cavernoma with enhancement of the venous plexos formed in the late fase (arrows). (C) T2-weighted coronal MR image with stenosis of the common biliary duct (arrow), with irregular caliber and upstream dilatation seen in MRCP (D) image (circle).

CONCLUSION

- MR imaging cholangiography should be performed to patients with portal cavernoma who have persisting biliary tract abnormalities or cholestasis.

- An early diagnosis is important to manage symptomatic patients and to prevent complications.

- MR is the modality of choice as it provides non-invasive evaluation of biliary and vascular abnormalities.

REFERENCES

1. Khuroo MS, Yattoo GN, Zargar SA et al. Biliary abnormalities associated with extrahepatic portal veinous obstruction. Hepatology 1993;17:807–13.
2. Dilawari JB and Chawla YK. Pseudosclerosing cholangitis in extra- hepatic portal venous obstruction. Gut 1992;33:272–6.
3. Bayraktar Y, Balkanci F, Kayhan B et al. Bile duct varices or ‘‘pseudo-cholangiocarcinoma sign’’ in portal hypertension due to cavernous transformation of the portal vein. Am J Gastroenterol 1992;87:1801–6.
4. Chandra R, Kapoor D, Tharakan A et al. Portal biliopathy. J Gastroenterol Hepatol 2001;16:1086-92
5. Shin SM, Kim S, Lee JW, et al. Biliary abnormalities associated with portal biliopathy: evaluation on MR cholangiography. AJR Am J Roentgenol 2007;188:W341-7.
6. Besa C, Cruz JP, Huete A., et al. Portal biliopathy: a multitechnique imaging approach. Abdom Imaging 2012; 37:83-90.
7. Akaki S, Kobayashi H, Sasai N, et al. Bile duct stenosis due to portal cavernomas: MR portography and MR cholangiopancreatography demonstration. Abdom Imaging 2002;27:58-60.

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