Cholangiopathy associated with portal hypertension: up date

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A Brief History

Fraser and Brown first reported symptomatic biliary obstruction due to collateral vessels in a patient with EHPVO in 1944 .

Choledochal varices were first demonstrated on cholangiography by **Williams** in 1982 .

That collateral decompression by **portosystemic shunt** (PSS) surgery can lead to reversal of biliary obstruction was first reported in 1988.

the era of **endoscopic management** in this condition began in 1993 with the first report of endoscopic **biliary stenting** in this condition .

The nomenclature

- **Dilawari** ⇒ "pseudosclerosing cholangitis"
- **Bayraktar** ⇒ "pseudocholangiocarcinoma sign"
- **Malkan** ⇒ "cholangiopathy associated with portal hypertension".
- **Sarinet** ⇒ "portal biliopathy"
- **Dhiman** ⇒ "portal hypertensive biliopathy".

Other terminology that have been used include:
"vascularbiliopathy""ischemic cholangiopathy", "portal ductopathy"
"portal cholangiopathy"

- The term **"Portal cavernoma cholangiopathy (PCC)"** was agreed upon as the consensus nomenclature **in 2013 by the Working Party of the Indian National Association for Study of the Liver.**

- since it implied the **presence of a portal cavernoma** resulting in abnormalities of the biliary tree including extra- and intra-hepatic bile ductular system, gallbladder and cystic duct

Definition

- **uncommon** clinical condition in western countries, but more prevalent in the Indian subcontinent due to increased prevalence of neonatal umbilical sepsis and dehydration in children.

- defined as the **presence of abnormalities in the biliary tree** (including biliary tree and gallbladder) in patients with **non-cirrhotic/non-neoplastic** extrahepatic portal vein obstruction **(EHPVO) and portal cavernoma.**

- **EHPVO** is a primary vascular disorder of the portal vein in children and adults manifested by longstanding thrombosis of the main portal Vein and accounts for 40 % of cases of portal hypertension worldwide.

Natural history

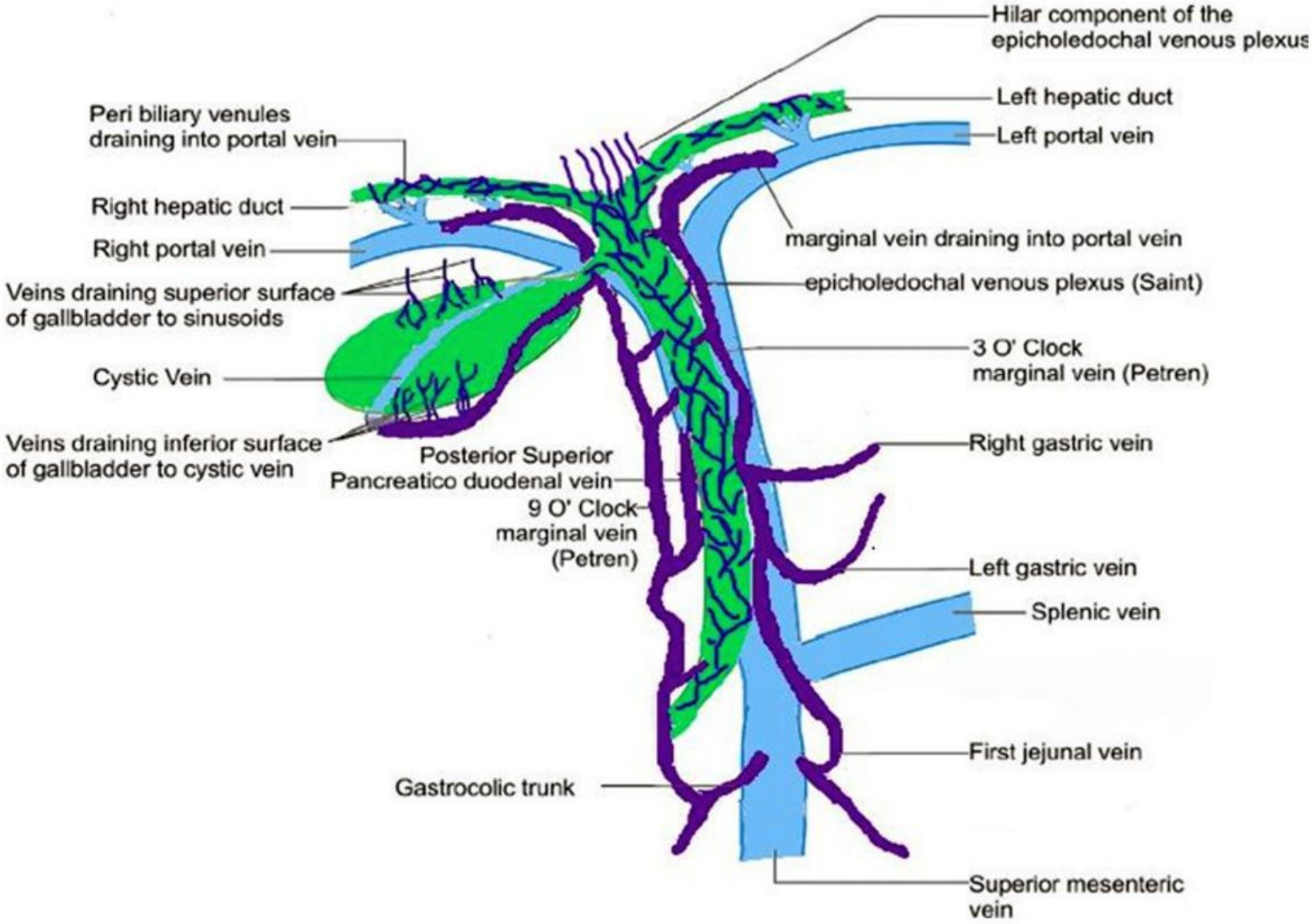
- Nearly all patients with EHPVO 70 -100 % have radiologic manifestations of portal cavernoma cholangiopathy, but the majority are asymptomatic initially.

- Biliary changes described in patients with cirrhosis is primarily intrahepatic and likely be due to parenchymal disease; absence of portal cavernoma also explains the absence of biliary changes in extrahepatic ducts in patients with cirrhosis.

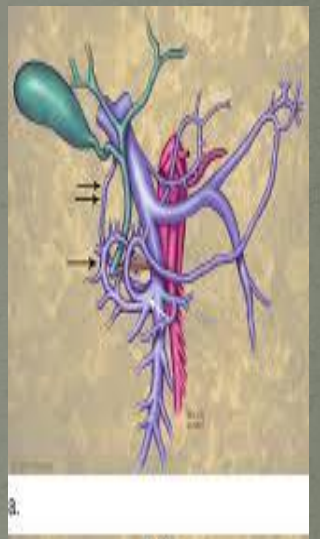
- However, progressive PCC may become symptomatic in 5–38 % of patients , typically 8–10 years after diagnosis , and lead to severe complications, including secondary biliary cirrhosis(2-4%).

- No evidence of malignant potential on long term follow-up exists in the literature

Portal cavernoma


Hilar component of the epicholedochal venous plexus
Left hepatic duct
Left portal vein
Peri biliary venules draining into portal vein
Right hepatic duct
Right portal vein
marginal vein draining into portal vein
Veins draining superior surface of gallbladder to sinusoids
epicholedochal venous plexus (Saint)
3 O' Clock marginal vein (Petren)
Cystic Vein
Veins draining inferior surface of gallbladder to cystic vein
Right gastric vein
Posterior Superior Pancreatico duodenal vein
9 O' Clock marginal vein (Petren)
Left gastric vein
Splenic vein
First jejunal vein
Gastrocolic trunk
Superior mesenteric vein

Conditions associated with cavernous transformation of the portal vein thrombosis
Myeloproliferative disorders
Hypercoagulable states
Pancreatitis
Pylephlebitis
Behçet syndrome
Cirrhosis (rare)
Idiopathic (30%)



PATHOGENESIS

rsible component of PCC

ogensis

ical extrinsec compression causing biliary irregularities

component of PCC

c ischemia of the bile duct wall leads to inflammation and fibrosis with high grade stenosis.

ment by a fibrous "tumor-like-cavernoma": Cavernous transformation result in fibrous hilar mass multiple tiny collater

classification

Sarin identified four PB types according with the biliary abnormalities localization	
involvement of **extrahepatic** bile duct only .	**type I**
involvement of **intrahepatic** bile duct only .	type II
involvement of **extrahepatic bile duct and unilateral** intrahepatic bile duct (left or right).	type IIIa
involvement of **extrahepatic bile duct and bilateral** intrahepatic ducts	type IIIb

Llop classified PB into different degrees of severity:	
biliary tree **irregularities** or **angulations**	**grade I**
indentatins or **strictures** without dilation	grade II
strictures with dilation (defined as intrahepatic duct ≥ 4 mm or extrahepatic duct ≥ 7 mm).	grade III

Prevalence and clinical characteristics

- symptoms are present in only 5–35 % of patients.
- Symptoms could be in the form of **long standing jaundice due to chronic cholestasis, or biliary pain with or without cholangitis due to biliary stones**
- **Jaundice** is invariably present in patients with symptomatic PCC, and was present at diagnosis in about 2/3rd of patients
- **Jaundice** is usually mild; mean serum bilirubin level was 2-3 mg/dL .
- Half to 2/3rd of patients have experienced **cholangitis.**
- **Abdominal pain** is seen in about half of symptomatic PCC.
- Diagnosis of EHPVO antedates symptomatic PCC by 8–10 years.

Risk factors associated with the development of symptoms:

Older age•
•History, of gastroesophageal variceal b
•biliary stricture with dilatation.
• presence of gallstones, and common b
abnormal liver function tests•
longer duration of disease•

Diagnosis

There is a consensus that a diagnosis of portal cavernoma related cholangiopathy should be established on three simultaneous arguments

- presence of a portal cavernoma
- typical cholangiograph changes
- absence of other causes of these

:Different diagnosis

Differential Diagnosis of Portal Cavernoma Cholangiopathy.
Primary sclerosing cholangitis Bile duct
Bile duct neoplasms
Biliary tract surgery
Biliary parasitosis
Choledocholithiasis
Congenital abnormalities of the biliary tract
Ischemic bile duct stricture
Toxic bile duct strictures
Strictures due to autoimmune and chronic pancreatitis

US

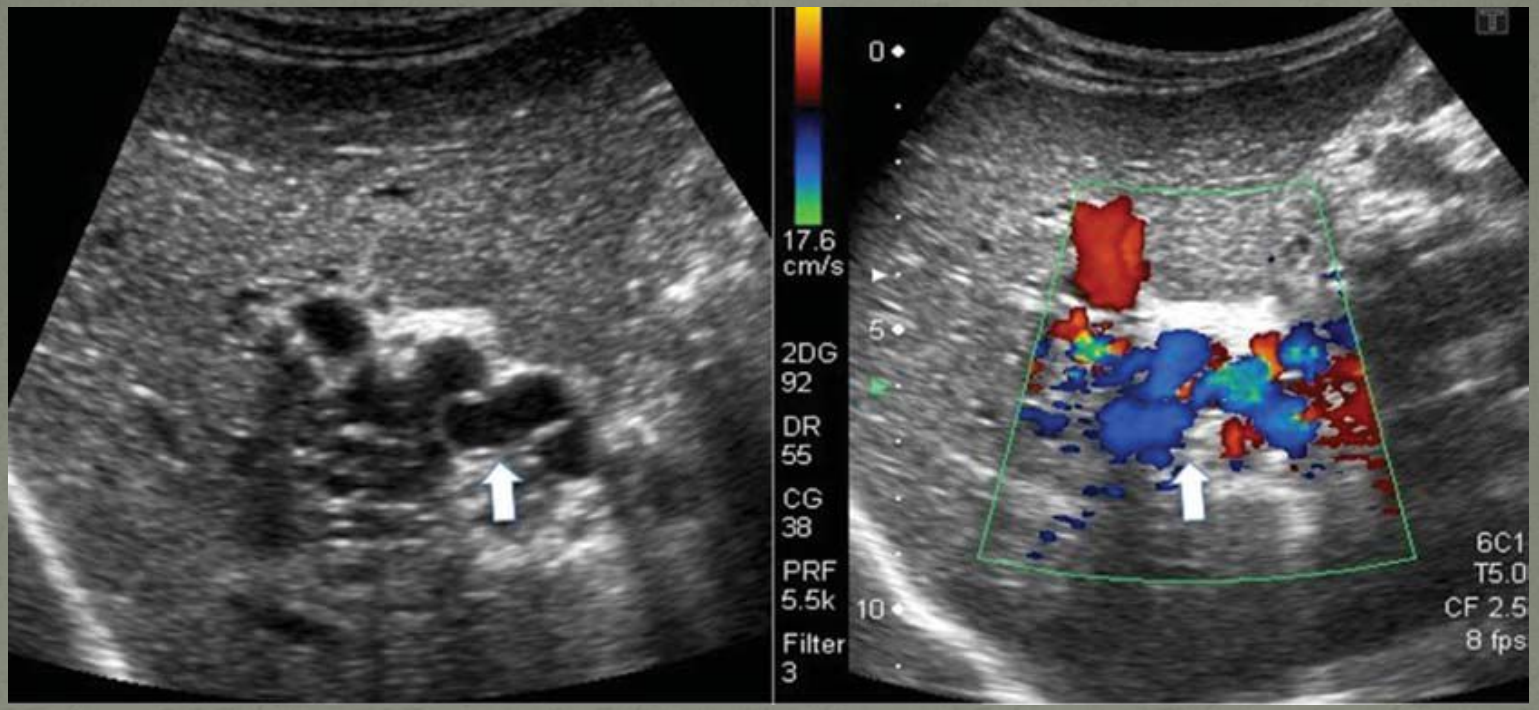
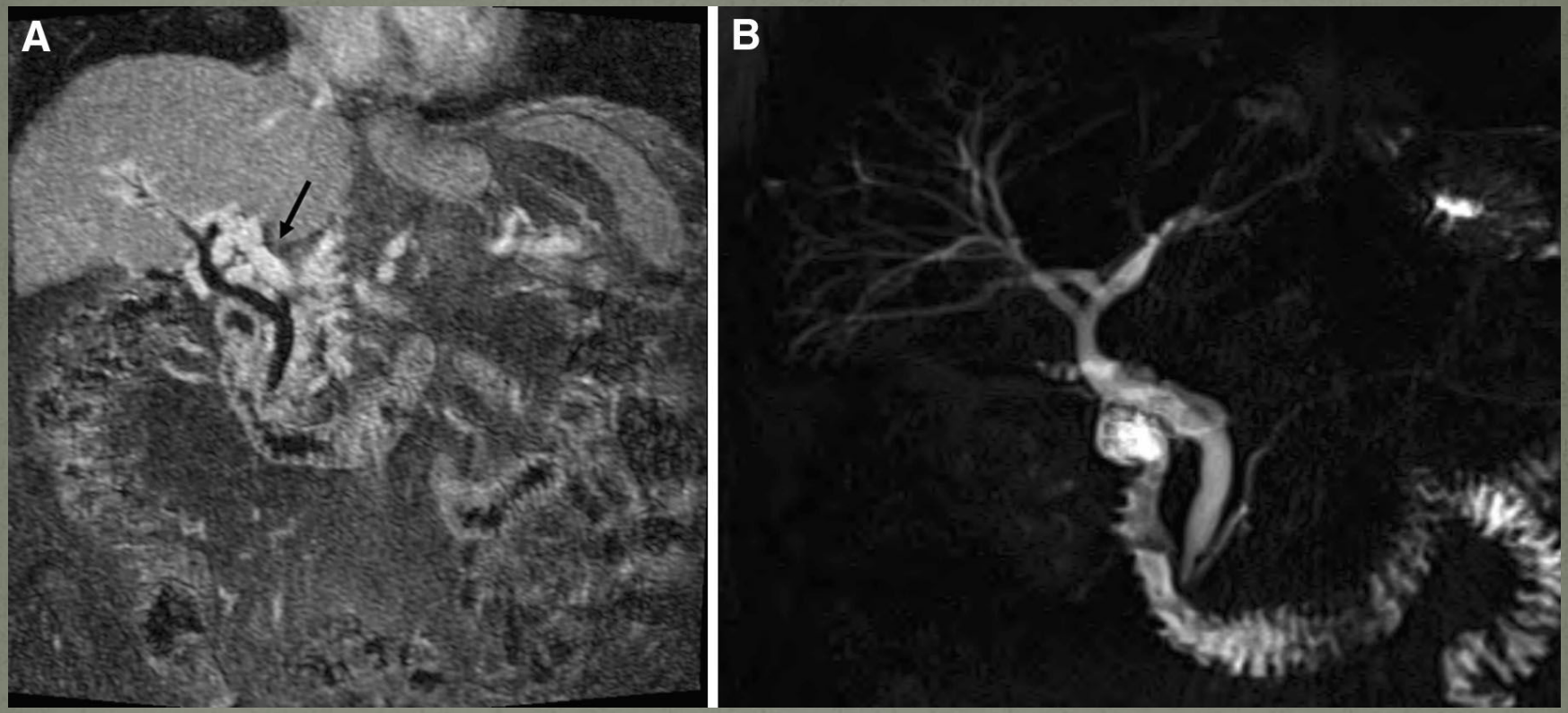


Figure 2. Ultrasonography (A) and color Doppler mapping (B) identifying multiple tortuous periportal veins in the hepatic hilum (arrows) corresponding to cavernous transformation of the portal vein.

CT

Cavernous transformation of the portal vein, CT appearance. A, B Axial and coronal contrast-enhanced CT images show multiple collateral vessels (black arrows) at the porta hepatis consistent with a portal cavernoma in the setting of portal vein thrombosis. Note mild central intrahepatic biliary dilatation. There is also splenomegaly

MRCP



Cavernous transformation of the portal vein, portal cavernoma cholangiopathy, MR appearance. A Coronal contrast-enhanced T1-weighted image shows multiple collateral vessels at the porta hepatis (black arrow). B Coronal MIP MRCP image from the same patient shows mild intrahepatic biliary dilatation, left greater than right.

A standardized nomenclature has been proposed at the INASL working party, which is as follows

1-Extrinsic Impressions/Indentations Smooth thumb-like impressions on the bile duct, with a nodular contour.

2- Shallow Impressions/Indentations Smooth non-contiguous impressions on the bile duct, less than one-quarter of ductal Diameter.

3-Irregular Ductal Contour Fine-wavy, irregular contour of the bile duct walls due to contiguous shallow indentations, less than one-quarter of the ductal diameter.

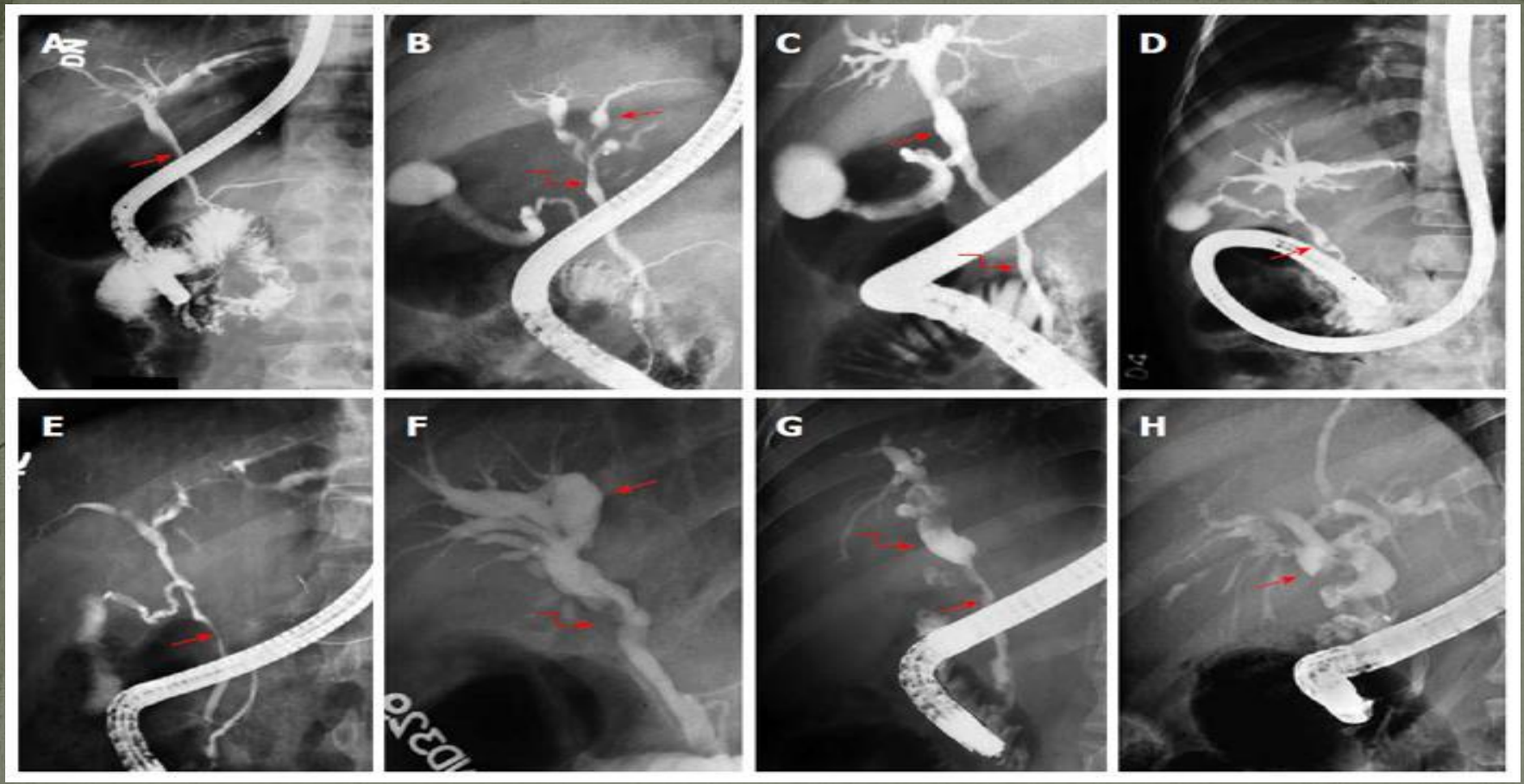
4-Filling defects: round, oval, or elongated defects ,can represent stones, prolapsing intra-luminal varices, or clots.

5--Stenosis: might be associated with upstream dilatation and may be due to extrinsic compression by collaterals or intrinsic narrowing or stricturing due to mural fibrosis.

6-Bile Duct Angulation It is proposed that an angle of < 145between lower and upper CBD be considered as significant.

7-Ectasia It is the dilated segment of biliary tree without any evident downstream obstruction

8-Upstream Dilatation Proximal dilatation can be similarly classified as “mild tomoderate” or “severe,”

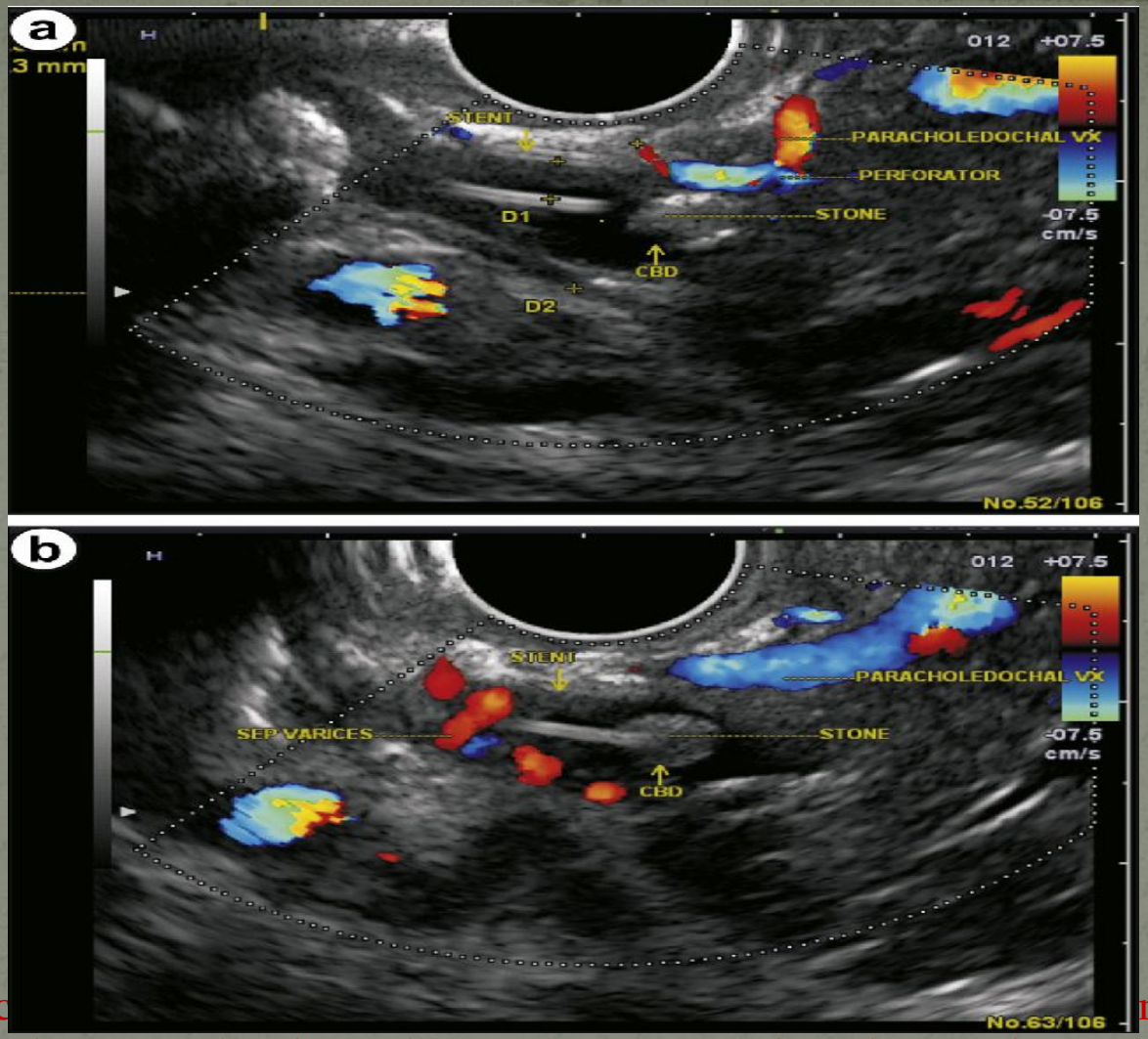


(curved arrow);
D: Angulation (arrow) of common bile duct;
E: Large smooth impression (arrow) on common bile duct
F: Angulation common hepatic duct (curved arrow) and gross ectasia of intrahepatic ducts (arrow);
G: Long smooth stricture of common bile duct (arrow) with upstream dilatation (curved arrow);
.[. H: dilated bile ducts with multiple filling defects (arrow)

Endoscopic Ultrasound

- Role of EUS in the diagnosis and management of PCC is evolving.
- EUS with Doppler can accurately differentiate paracholedochal, epicholedochal,intracholedochal and subepithelial varices.

- **This distinction is especially relevant if a subsequent ERCP is planned**, since the presence of intracholedochal and subepithelial varices may increase risk of bleeding during stone extraction and stricture dilatation respectively.



a & b On endosc rge, serpiginous, anechoic vascular channels in and/or surrounding the extrahepatic biliary tracts. In this image the presence of stent near the stone allows detection of varices inside

- the CBD (a and b)

cholangioscopy

- Normal bile duct mucosa is observed as a flat surface, witth fine networks of thin microvessels
- Benign inflammatory mucosal lesion may be observed as a slightly homogeneous papillogranular surface.
- Cholangioscopic findings suggesting malignant lesion show thick irregular tortuous vessels ("tumor vessels"), irregular papillogranular.
- Cholangioscopy has also allowed for a direct view at the subepithelial endocholedochal varises in PCC.

Management

- Asymptomatic patients with PCC do not require any treatment
- it should be focused on the management of portal hypertension and relief of biliary obstruction.
- Patients with symptomatic PCC, are candidates for the intervention ,whether surgical or endoscopic.

Endoscopic treatment

- Initially ERCP in PCC were used to tackle emergency conditions like cholangitis due to strictures or stones using plastic stents pending surgery.
- Subsequently endoscopic stone removal in PCC was demonstrated to be a safe procedure, with only few reports of hemobilia.
- Many patients are not candidates for surgery due to absence of a shuntable vein or liver dysfunction. These patients may require long-term endoscopic management in the form of repeated plastic stent exchanges or placement of removable covered metal stents

- Sphincterotomy has not shown to be associated with increased bleed rates in PCC and use of Dormia baskets and balloon extractors has been shown to be safe.

- These are generally preoperative procedures, **to be followed by porto- systimic shunt surgery.**

- **Repeated stent exchanges** may occasionally result in resolution of stenosis but this option is usually resorted to as fall-back therapy for those with no surgical options for portal decompression.

- Portal decompression surgery may be the first phase of therapy in patients in whom endoscopic intervention is not required such as a patient with isolated stenosis of CBD and with a shuntable vein

Hemobilia

- The risk of hemobilia has been studied as a major concern during endoscopic management of PCC.
- Intra-choledochal varices, masquerading as filling defects, may be the source of bleeding during calculi extraction.
- Moreover, manipulation of the bile duct also carries a significant risk of complications.

- Hemobilia has also been reported for other interventions such as exchange of plastic stents, pneumatic balloon dilatation of stricture, and balloon-occluding cholangiogram during endoscopic management of PCC.

- Hemobilia can be managed conservatively in most cases and is not more troublesome than in patients without PCC.
- different modalities can used to treat acut hemobilia: Endoscopic balloon compression, terlipressin injection have been described .

- FcSEM is a good treatment option when conservative treatment or other endoscopic treatments fail and is the preferred option when there is acute massive bleeding.

Plastic or metalic stent?

In the past

- Data suggest that repeated exchanges with bundles of plastic stents for 12–18 months, is an effective strategy in management biliary stricture.
.
- the place for covered, removable self-expanding metallic stents in the management of symptomatic PCC is being explored presently.

Recent finding

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Pseudocholangiocarcinoma Sign: Management of Portal Cavernoma Biliopathy with Fully-Covered Self-Expandable Metal Stent

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We present a rare case of symptomatic PCB with a common bile duct (CBD) stricture, refractory to conventional endoscopic therapy in a patient unfit for surgery. Owing to the risk of repeated plastic stent exchange, we chose a more definite endoscopic approach with temporary placement of fully-covered self-expandable metal stent (fcSEMS). In this paper, we also report our successful results following application of fcSEMS.

An 82-year-old man presented to our hospital with fever, right-upper quadrant abdominal pain, and jaundice. He had undergone cholecystectomy at another hospital for acute cholecystitis in 2002, complicated with an EHPVT. In addition, he had a history of aortic stenosis, hypertension, and congestive heart failure.

His laboratory values were indicative of leukocytosis (16.700/uL) and elevated liver enzymes, with a total bilirubin of 8.9 mg/dL.

Abdominal ultrasound revealed dilated intrahepatic bile ducts (IHBD) and CBD, as well as an EHPVT.

The patient was diagnosed with acute cholangitis and treated with antibiotics. Endoscopic retrograde cholangio-pancreatography (ERCP) showed asymmetric stenosis in the middle portion of the CBD (compatible with portal biliopathy type I2) and some small stones above it, with dilation of the proximal CBD and IHBD (Fig. 1A). After sphincterotomy and careful stone extraction with Dormia's basket, a 10-Fr plastic stent was placed. Balloon dilation was avoided due to hemobilia risk, which has been reported before [3,4].

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ORIGINAL ARTICLE

Management of portal cavernoma-associated cholangiopathy: Single-centre experience

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PORTAL HYPERTENSION (J GONZALEZ-ABRALDES AND E TSOCHATZIS, SECTION EDITORS)

Portal Cavernoma Cholangiopathy

Dominique-Charles Valla[1,2]

Abstract

Purpose of Review Portal cavernoma cholangiopathy is a rare condition, a proper recognition of which is critical. Solid data o this condition are scarce. This review aims at updating current knowledge on its definition, manifestations, diagnosis, an management.

Recent Findings A consensus has been reached to prefer the denomination portal cavernoma cholangiopathy for the bile duc irregularities associated with portal cavernoma. Such irregularities are characterized by stenosis with or without dilatation and ar mostly related to the impinging on bile duct lumen by portoportal collaterals. While bile duct irregularities are found in over 80% of patients with portal cavernoma, clinical manifestations, and complications (cholecystitis and biliary stones, but rarely chole stasis) occur in only 5–35% of them. Diagnosis can be and should be based on findings at magnetic resonance cholangiograph

Recent finding

- There are no guidelines regarding the use of SEMS in symptomatic PCB mainly due.
- When biliary access is difficult, some workers have resorted to the placement of covered removable self-expanding metallic stents.
- Plastic stents, in particular, were more likely to be blocked and required more endoscopy sessions for replacement.
- However, it is also important to remember that repeated plastic stenting carries a possible risk of complications.

- Patients who had a FcSEMS inserted had a better outcome in term of prolongation of "symptoms free" period and fewer requirements for restenting.

- FcSEMS alone achieved eradication of the choledochal varices in most of the patients .

- Nevertheless, varices recurrence was noted. This may suggest a **repeated stenting programis required to eradicate choledochal varices similar to that of oesophageal varices banding.**

In conclusion

- According to the results obtained in recent studies, we believe placement of fcSEMS might be a good option Especially when surgery is contraindicated, progressive plastic stenting fails, and no episodes of hemobilia have occurred during previous endotherapy.
- It is reasonable to think that if bleeding has not occurred during plastic stent exchanges, the risk for it to occur during removal of a metal stent is smaller.
- However, more data are needed regarding this treatment approach.

Cholangiograms obtained during endoscopic retrograde cholangio-pancreatography (ERCP). Asymmetric stenosis in the middle portion of the common bile duct (CBD) ("pseudocholangiocarcinoma sign") with small stones above it and dilation of the proximal biliary ducts. (B) Following failure of progressive plastic stenting, a 80×10-mm fully-covered self-expandable metal stents (fcSEMS) was placed. (C) Stent removal 6 months thereafter .revealed improvement in the stenotic segment of the CBD


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