

# Portal Biliopathy



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## Portal Biliopathy

Abnormalities in biliary system (extra  $\pm$  intra hepatic) from extrahepatic portal venous obstruction / portal cavernoma (PC) Fraser (1944), Gibson (1965), Hunt (1965)- CBD obstruction by PC Biliary obstruction improved after surgical decompression of PH

Choudhri G DDS 1988:33:6267

## Diagnosis Imaging

Portal cavernoma

Characteristic Cholangiographic alterations on MRC / ERC

Lack of other biliary causes, PSC, Cholangiocarcinoma

Dhiman & Chawla etal JCEH 2014:4

**Synonyms** Portal Cavernoma Cholangiopathy (INASL 2014)  
Pseudosclerosing cholangitis, Portal Hypertensive Biliopathy  
Portal Cholangiopathy, Portal Ductopathy  
Portal Pseudocholangiocarcinoma

Rare Cirrhosis/ NCPF- Intrahepatic biliary radicals- hepatic nodularity

INASL Consensus- 2014 Portal Cavernous Cholangiopathy  
Series published- limited number of patients (8-60)

**Limitations** Prospective studies lacking  
Cirrhosis with PB

## Bile Duct Vascular Anatomy

### Artery

Branches of Celiac trunk Posterosuperior pancreaticoduodenal  
Right & Left , hepatic & cystic branches perforate bile duct wall/  
intramural/ sub epithelial plexus

Supraduodenal bile duct- least vascularised - ischemia

**Vein** Epicholedochal Plexus of Saint fine reticular <1mm  
Paracholedochal Plexus of Petren Parallel to common bile duct  
Drain to Gastric, Gastrocolic, posterior superior  
pancreatoduodenal veins

Perforators- extra-mural to intra-mural veins

Portal Vein Thrombosis- jump collaterals

PV- solid tumor like Portal Cavernoma- encircling bile ducts

## Pathogenesis

Block of Splenoportomesenteric venous system (children > adults)

## Compression Collaterals around CBD

(indentations, scalloping and filling defects) by Plexus of Saint & Petren pericholedochal plexus - reversible bile shunts

## Ischemia

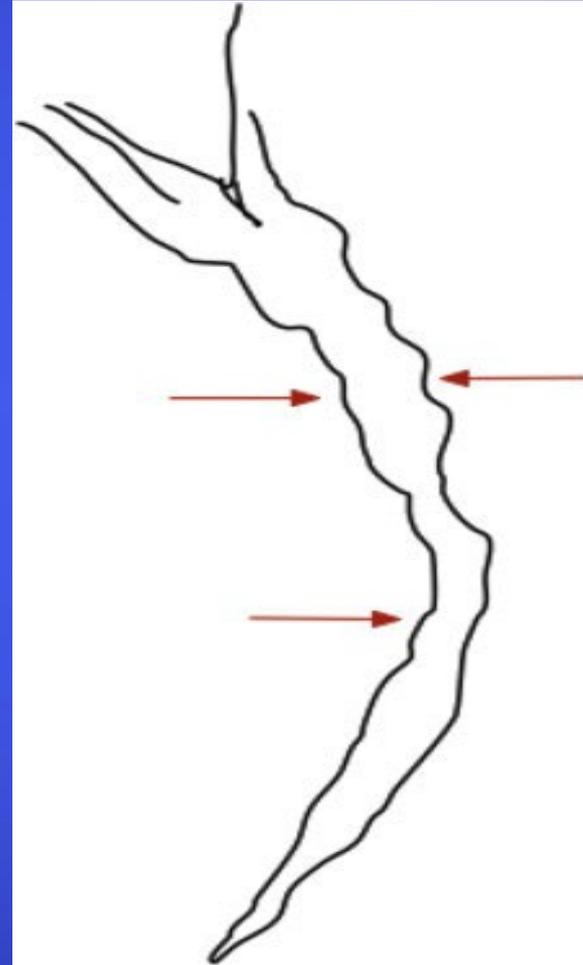
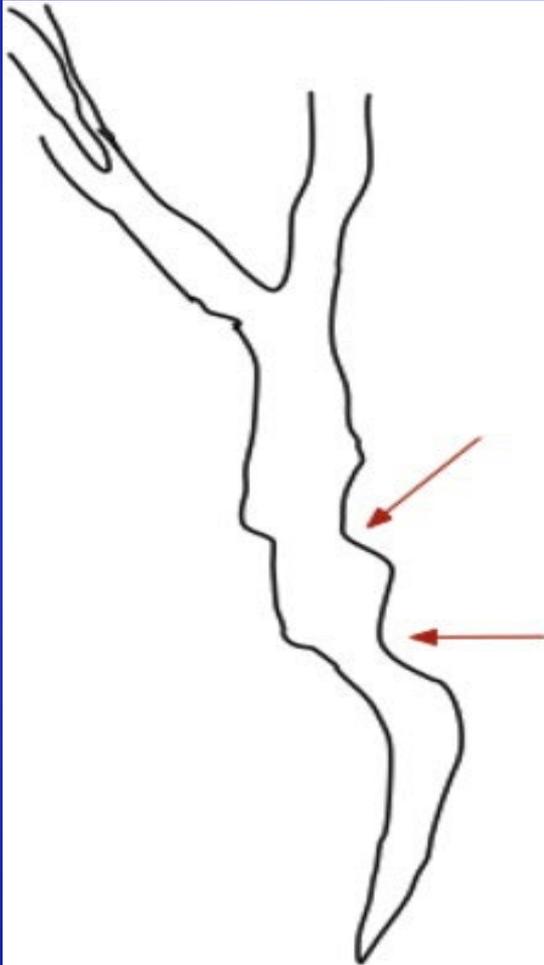
Thrombosis & Sclerosis of small venules and arterioles of bile ducts, local ischemia, prolonged local wall compression, short segment fibrotic strictures, wall thickening and portal cavernoma

## Infection

Cholangitis- inflammation, neogenesis-fibrous tissue deposition  
Some ischemic other compression

Extrahepatic biliary abnormalities 60-97% left HD more severe

## Bile Duct Abnormalities



## Natural History PB

Sparse Not well studied progressive?

Disease of young 6yrs 78-100 % asymptomatic

No clinical or laboratory parameters predict symptomatic PB

22 PVT, 45 portal cavernoma fup 4yr- non progressive

Llop etal Gut 2011:60:853

53 PB in children <13 yrs MRC all asymptomatic

Extra & intrahepatic ducts- 85%, Severe- 58.5%

Venkatesh etal JCEH 2022:12

More often PV+SV+SMV

↑ angulation of CBD increase chances of stones

Waker etal Radiol 2011:258

60% PB develop in a year after PVT

Llop etal Gut 2011:60

## Symptomatic

Prognosis poor if effective treatment not given

5-38 % - once fibrotic stricture develop

Older, longer duration of EHPVO (8-14 yr), Dilated segments of bile ducts, stones

Recurrent Jaundice 52.8%, abdominal pain 19.4%, Cholangitis 27.1%

Stones Gall bladder-35%, possibly related to biliary stasis

↑ lithogenicity, hemolysis, hypersplenism

Choledocholithiasis 40%, higher in symptomatic than asymptomatic-  
Independent of GS

Bhatia etal GE Endo 1995:42:172, Saraswat etal JCEH 2014:4, Girish etal Acad J.surg 2020:3:176

long/ multifocal strictures ± choledochal or intrahepatic calculis

Condat etal Hep 2003:37:1302, Dhiman etal JCEH 2014, Irene etal WJG 2016:7:22

## Gall Bladder Vx

34% EHPVO, 13% Cirrhosis, 24% NCPF

Dilawari & Chawla Gut 1992:33

57/ 97 symptomatic PB had non shuntable veins  
Index bleed 12 yr ago

Repeated ERCP interventions

Shukla etal JCEH 2017:7:328

## End Stage Liver Disease

2-4% extensive venous thrombosis- progressive liver  
dysfunction- recurrent cholangitis, secondary biliary cirrhosis

Condat etal Hep 2022:37

## Investigations

Liver Function Tests- Normal

**US and colour Doppler** screening modality- atretic/ recanalised PV  
Portal cavernoma, mass with serpiginous collaterals, biliary dilation,  
GB varices, GS, choledocholithiasis

Webb etal Cv Med 1979-48 Vipert etal Am Srg 2007:246

**Hyperbilirubinemia** Indication for further biliary imaging

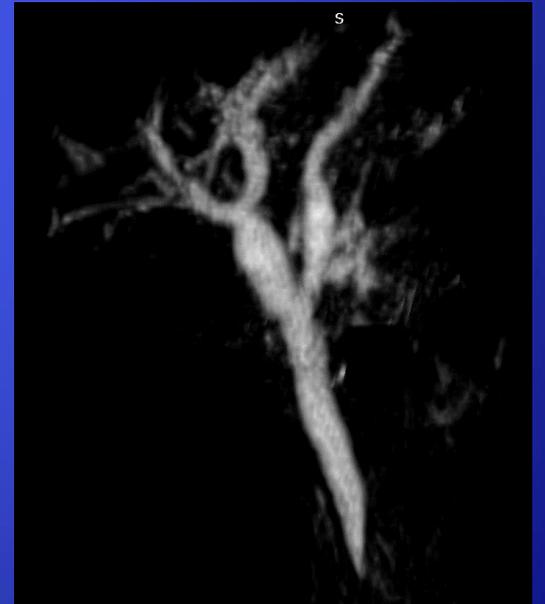
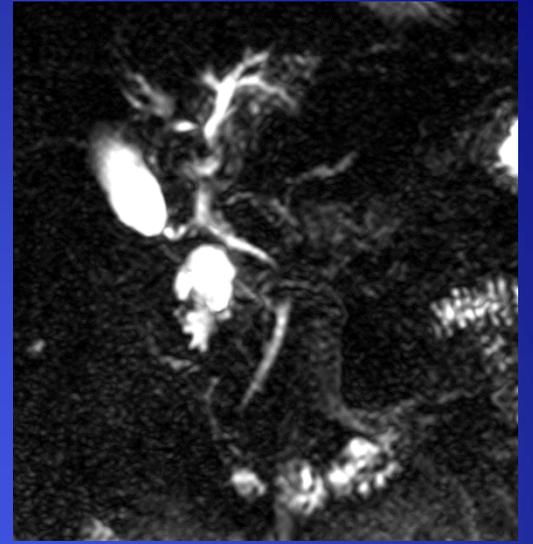
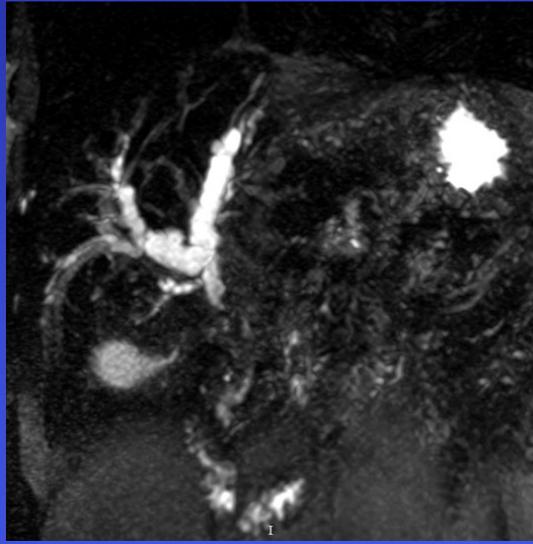
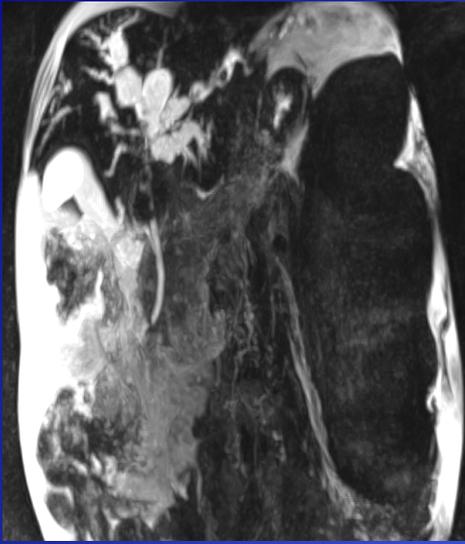
**MRC & Portography** vascular & biliary anatomy- non-invasive

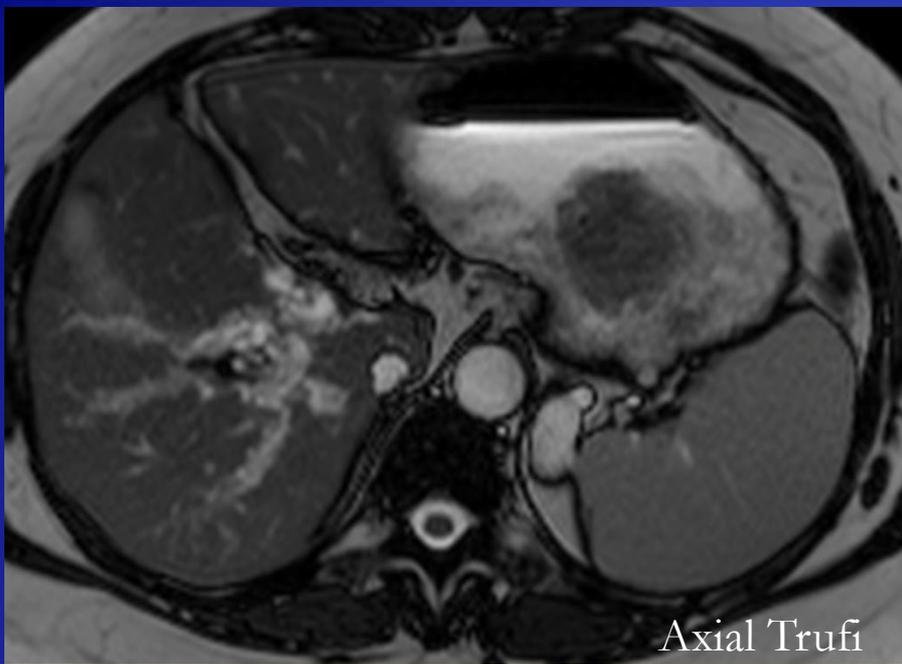
**Gold standard** detects strictures- biliary tree

CBD Undulating, narrowing and nodular extrinsic defects

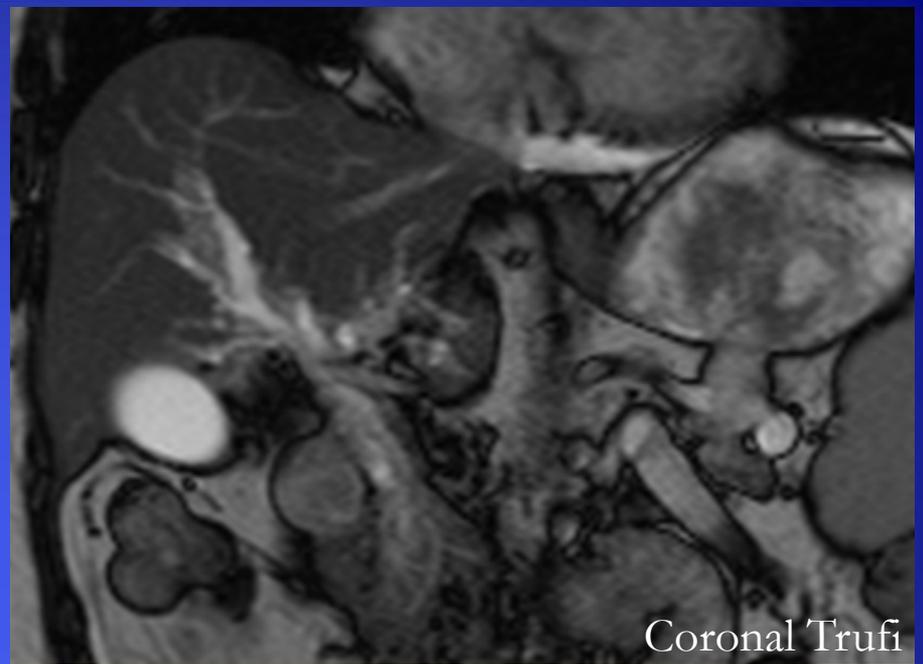
Differentiates stones from varices- filling defects

Roadmap for biliary & vascular intervention



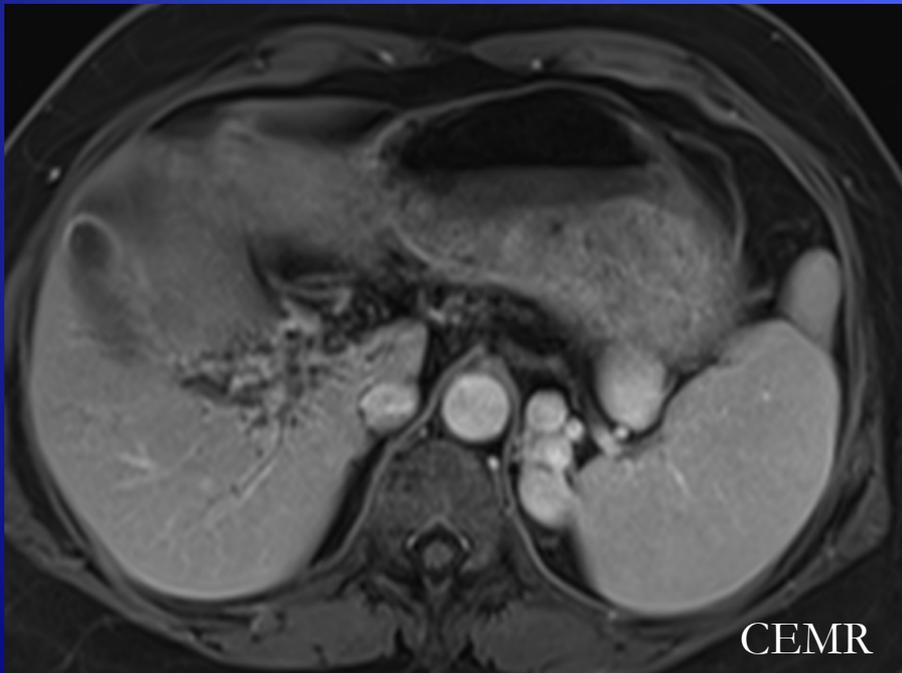


Axial Trufi

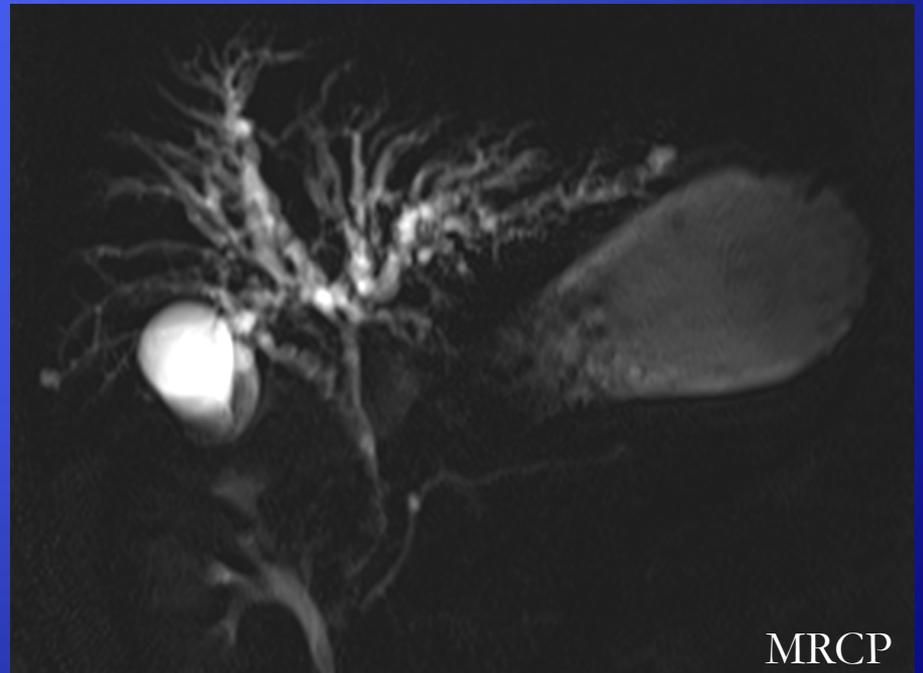


Coronal Trufi

Portal cavernoma with dilated central biliary radicles



CEMR



MRCP

## CECT

Extent of thrombosis & collaterals

Assess biliary dilation & CBD changes, less accurate than MRCP, radiation exposure

Pancreatic head enlargement (pseudo mass)- peripancreatic collaterals

*Agurre et al Clin Imaging 2012:36, Khuroo et al Hep 1993:17*

## ERCP

If treatment required- cholangitis, Stones - invasive

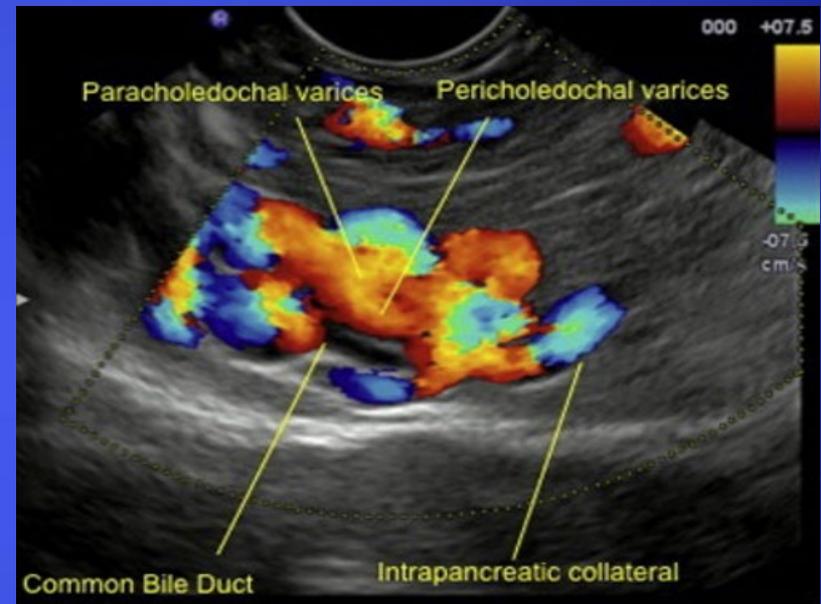
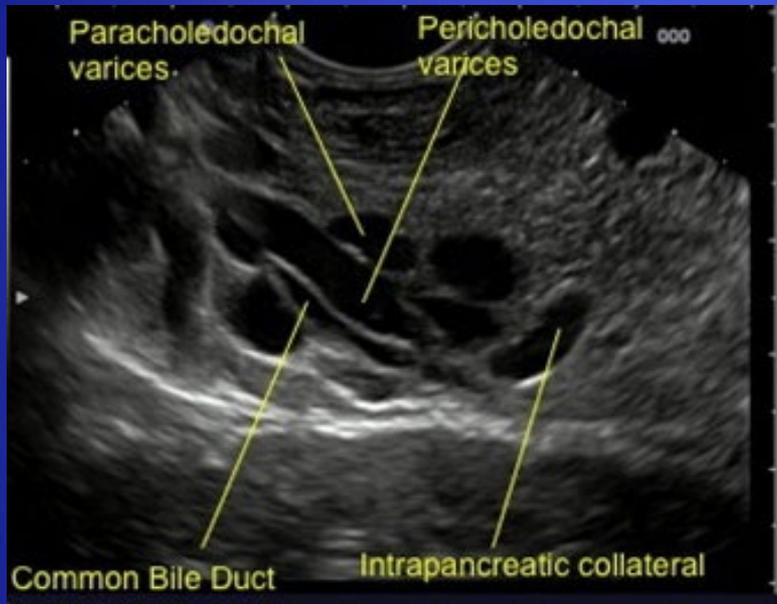


## EUS

As accurate as MRC- operator dependant, invasive  
Bile duct varices- multiple anechoic vascular channels  
within epicholedochal or outside wall paracholedochal  
differentiates from stones

Visualises relationship- Collaterals & biliary tree  
EUS doppler identifies perforators between  
paracholedochal, epi & intra choledochal varices  
Helpful in Biliary endotherapy

# Para and Peri-Choledochal Varices



## Classification of PB

- Location Type I** Extrahepatic bile duct
- II Intrahepatic bile ducts
  - III a. Extrahepatic & unilateral intrahepatic bile duct
  - b. Extrahepatic/ bilateral intrahepatic bile duct

Chandra & Sarin etal JGH 2001:16:1086

- Severity Gr I** Irregularities/ angulations of biliary tree
- II Strictures without dilation
  - III Stricture with dilations
- Extrahepatic >7 mm, Intrahepatic <4mm

Llop etal Gut 2015:60

- Types**
- Varicoid extrinsic compression
  - Fibrotic
  - Mixed

Shin etal AJR etal 2007:188

## Differentials

Primary Sclerosing Cholangitis, Cholangiocarcinoma  
HIV cholangiopathy, Biliary Ascariases  
IgG4 Cholangiopathy, Pericholedochal LN

## Histology

50 patients PB with portal cavernoma  
Dilated multiple portal venous channels (72%)  
Hepatic artery thickening (70%), Elastosis of PV (50%)  
Aberrant vascular channels around portal tract (54%)  
Bile ductular reaction 44%  
Extreme wall thickening in cavernoma

Rastogi etal Diagnostics 2023:13

Hushoff etal Diagnostics 2022:10

**Management** No intervention if asymptomatic LFT  $\pm$  N

**Intervention** Only in symptomatic

Step wise approach less to more invasive procedures

Endoscopic  $\pm$  Radiological  $\pm$  Surgical complementary

Dilemmas for Physicians and Surgeons

Experience limited

### **ERCP Biliary Interventions**

If cholestasis, choledocholithiasis & cholangitis,

Cholangiolar abscess & stricture

Sphincterotomy/ Balloon sphincteroplasty -  $\uparrow$  risk of bleed (rare)

Hemobilia, mild, settles conservatively occasionally

Terlipressin/ induced hypertension

? balloon extraction / Dormia basket

Intracholedochal Vx mimics stones

Layee et al GE Endo 2009:70

Pericholedochal collaterals & congested biliary walls surgical back up

Sharma et al GI Endo 2009:70:171

- Nasobiliary Drainage
- Stent exchanges- Plastic- cholangitis, Relapses common  
Largest series 130 ERCP in 20 symptomatic PB  
Balloon Dilation and Plastic stent exchanges every 3-4 months till LFT  
Complications hemobilia and recurrent cholangitis, sludge, blood  
compliance  
Stricture resolution with prolonged bundles of plastic stenting

Saraswat etal JCEH 2014:4

- Non covered metallic stent not used- intimal hyperplasia
- Self expanding covered metallic removable stent(FCSEMS) in  
strictured CBD non responding to repeated plastic stunts  
control haemorrhage Rescue Therapy  
Cholecystitis in 48 hours block of cystic duct  
Asymptomatic 3 years (1-10 yrs follow up)

Buyruk etal BMC Gastro:2023:23:414

114 pts- 30% single endoscopic intervention

43% multiple plastic exchanges      3.5% metallic stent

30.7% surgical treatment shunt      1.7% TIPS

0.9% LTx

Irene etal WJG 2016;22:9909:20

- Laser / Mechanical lithotripsy stones

Goenka etal GI Endo 2014:80

Long term outcomes in Non shuntable PB or failed surgery

35 PB- 37% Cholangitis, 51% Jaundice

3-29 procedures- total 363 endoscopic sessions

Hemobilia 6%, 28.5% frequent stent exchange cholangitis

Secondary biliary cirrhosis- 11.4%

Giri etal JCEH 2022

**Cholangioscopy** case reports- Biliary casts described  
clotted blood & bile

Marco etal Endo IO 2015:3

## Intervention radiology

**PTBD** if cholangitis- unstable hemodynamic altered mentation

**TIPS** Technically challenging- well selected success 87-100%

5 yr primary patency 73%

Restenosed 70-90% 28-50% not relieved

Rapid recurrence after stent

Transjugular Transhepatic Transsplenic

Transmesenteric Collateral vein stenting

6 cases TIPS- PB- No complications

Decompresses peribiliary plexus

PTBD + TIPS

## Surgical

Definitive, highly complex and risky

Portal decompression, Shunts reverse PB 60-80%- within 2 wks

39 PB patients PSR shunt relieved PB (65%) &

facilitated safe, second stage biliary decompressive procedure

Hepaticojejunostomy/ Choledochoduodenostomy

Agarwal etal HPB 2011:13:33-23

Non selective shunts preferred

Proximal SR shunt/ mesocaval 5-30%

SV non shunable- mesogonadal vein shunt

mesorenal shunt, portal Vx & Cava

Borg etal Radiol 2004:231, Camerto etal dig Surg 2010:27

Rex shunt - procedure of choice for EHPVO left branch usually not patent collaterals

## Non shuntable veins (25-30%)

Bilo enteric drainage Hepaticojejunostomy

18 patients fup 11 mo 12 yr

One operative mortality

Recurrence in 2 pts

Aggarwal etal HPB 2011:13

## Esophagogastric devascularisation with splenectomy

may help- lowers portal pressure?

Chattopadhaya etal HPB:2012:14:441-7

## Biliary Surgery

Even after shunt- bleed Roux-en y anastomosis

4 case series Roux en Y biliary Hepatico jejunostomy

Bleed due to collaterals in hepatoduodenal pedicle

Ochoa Int J Surg 2023:110

**Laparoscopic cholecystectomy** for symptomatic GS in 16 patients

successful with good surgical hand

excellent outcomes in selected groups with prior portal vein

decompression

Han et al. Ann.HBP.surgery:2023:27:366

**Liver Tx** Non-Shunable vein, ineffective biliary drainage and progressive liver damage, recurrent cholangitis, repeated stents, secondary biliary cirrhosis  
DDLT preferable- small for portal inflow or graft & other venous collaterals

Gupta et al JCEH 2014:4

## Conclusions

Portal biliopathy always present in extrahepatic portal vein obstruction

Mostly asymptomatic No treatment

Symptomatic long term prognosis poor Cholangitis, CBD stones

Management Endoscopic- stents plastic/ FCSEMS

Decompressive shunt surgery  $\pm$  Bilo enteric drainage

TIPS/ PTBD/ Liver Transplantation

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