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Endoscopy in Primary Sclerosing Cholangitis

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Goals

- Role of ERCP in PSC
  - Initial Diagnosis
  - Complications
- Managing procedure risks
- Role of MRCP

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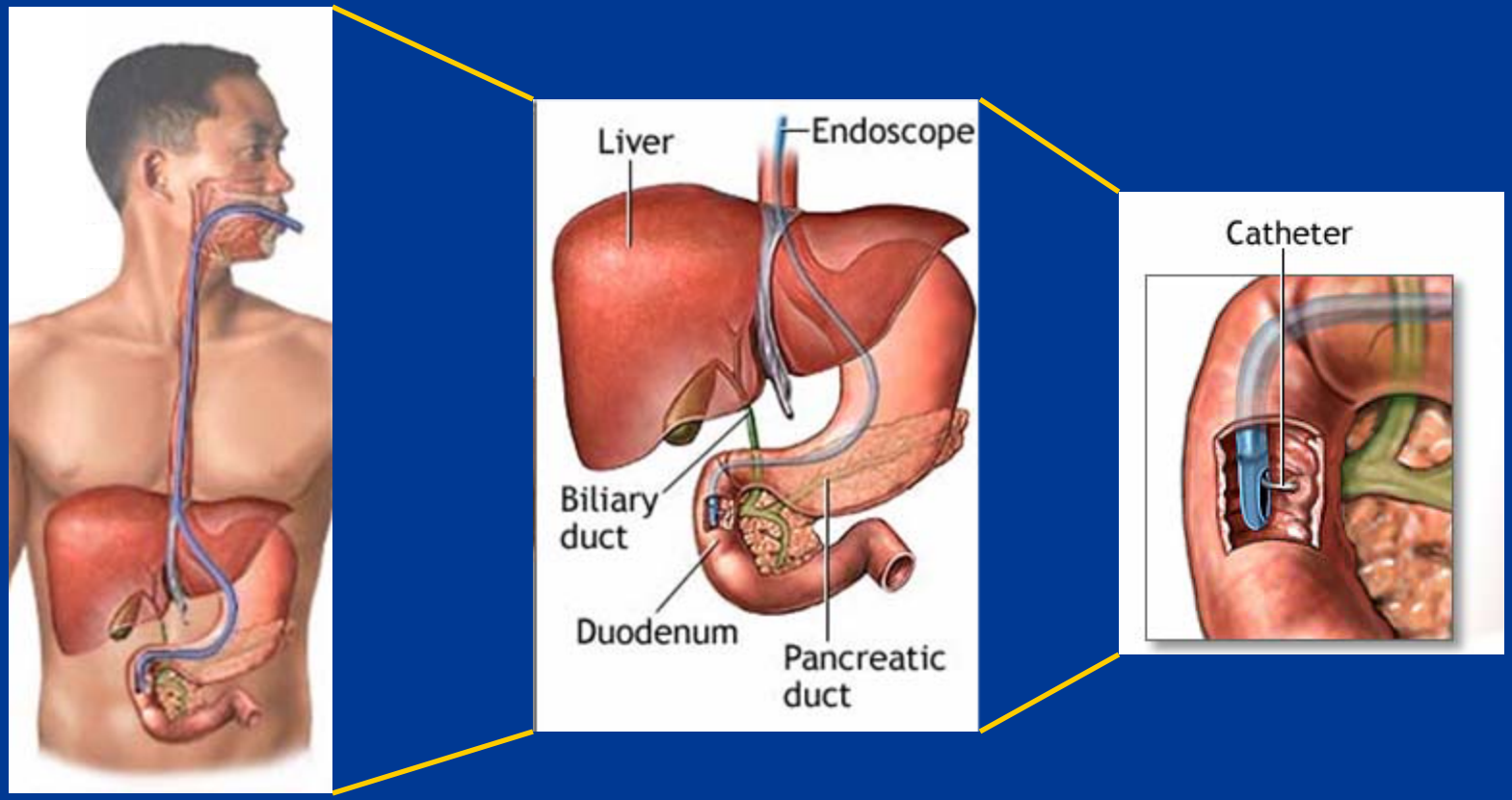
Endoscopic Retrograde CholangioPancreatography



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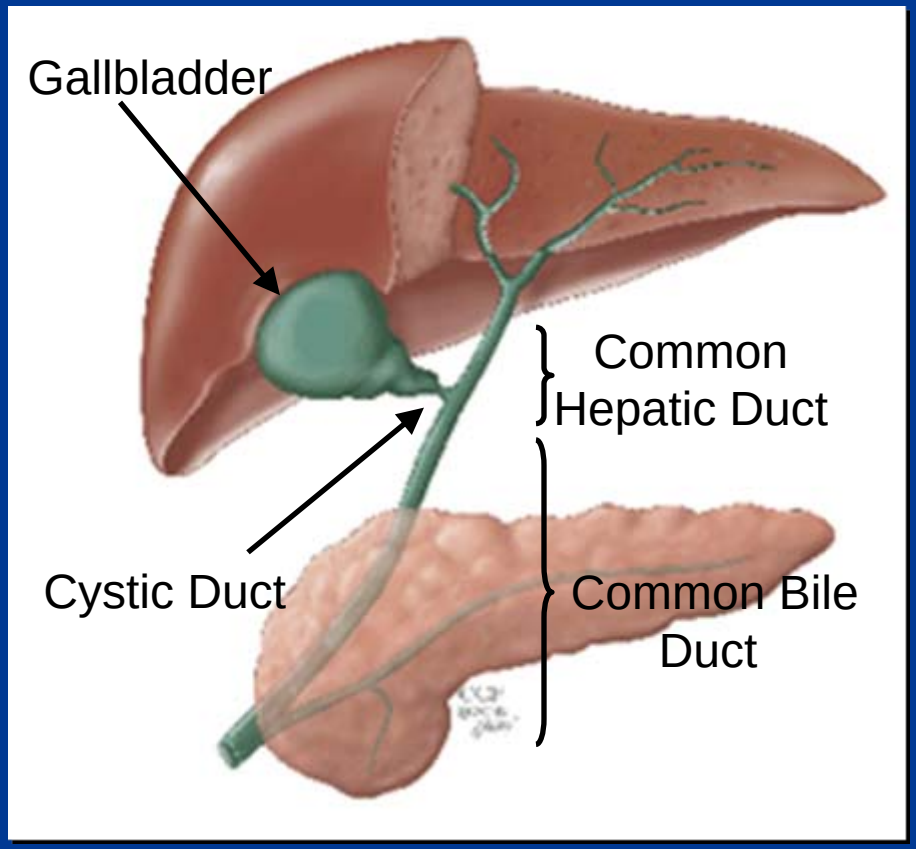
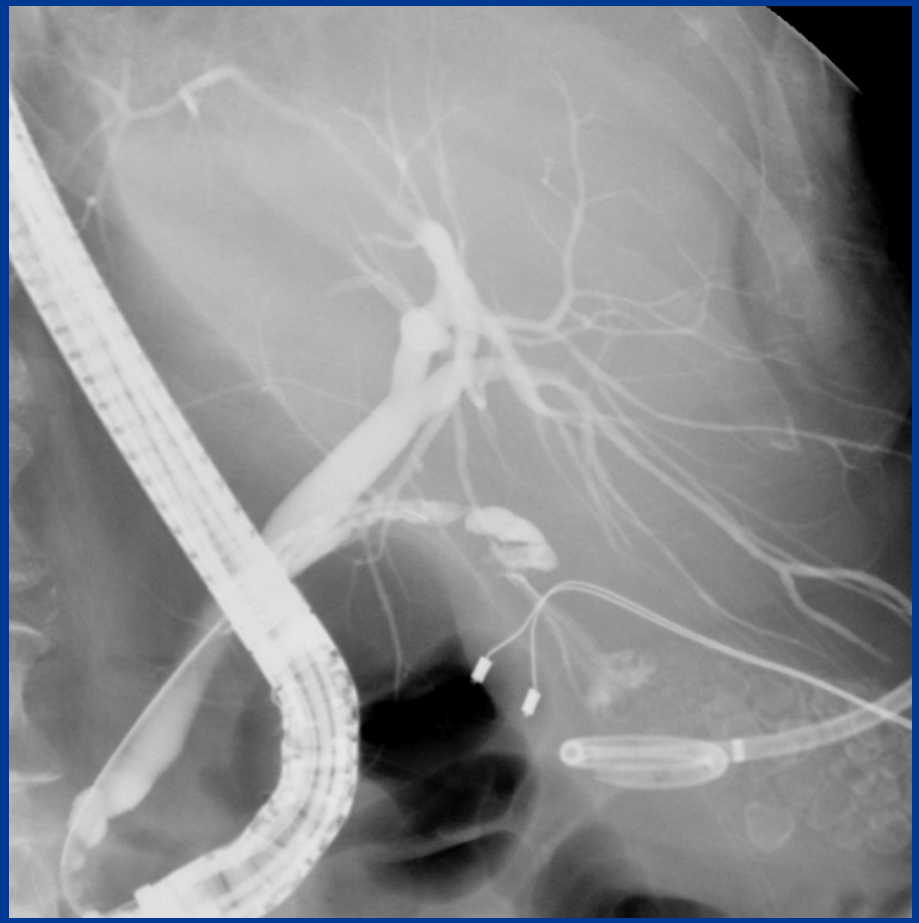
ERCP – anatomy on cholangiogram





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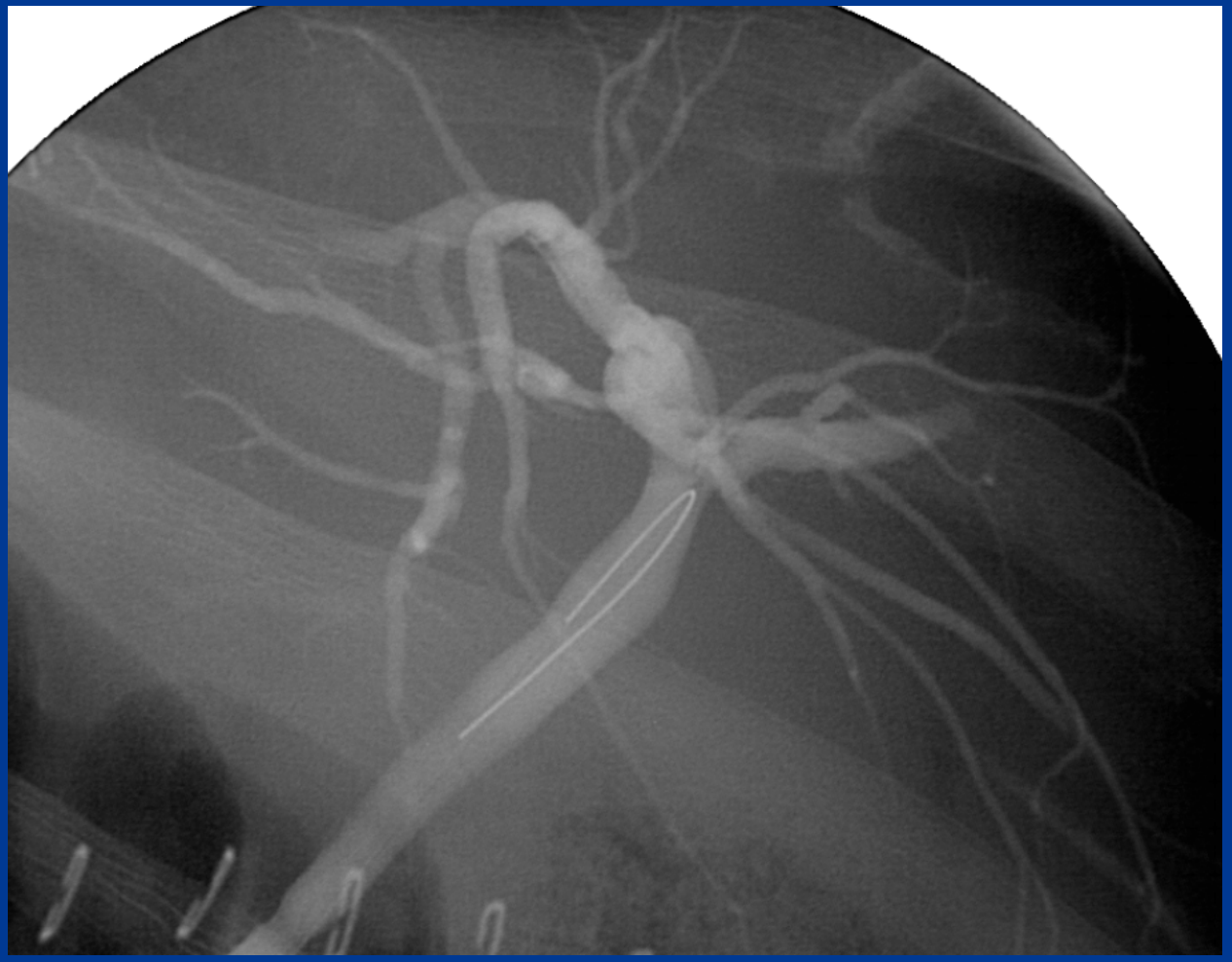
Normal Intrahepatic Cholangiogram



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PSC - pathogenesis

- **Progressive inflammatory destruction and fibrosis of intra and extra hepatic bile ducts**
- **Primary injury to medium - large sized bile ducts (>100 *u*m in diameter)**
- **Smaller ducts captured on liver biopsy demonstrate nonspecific changes of obstruction or disappear (ductopenia)**

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PSC – diagnostic criteria

- **Chronic cholestatic liver enzyme elevation**
  2-3x ↑alk. Phos. / bilirubin vs. AST/ALT > **6 mo.**
- **Compatible bile duct injury on ERCP (or MRCP)**
- **Exclusion of other causes of bile duct obstruction**
- **Liver biopsy plays limited role**

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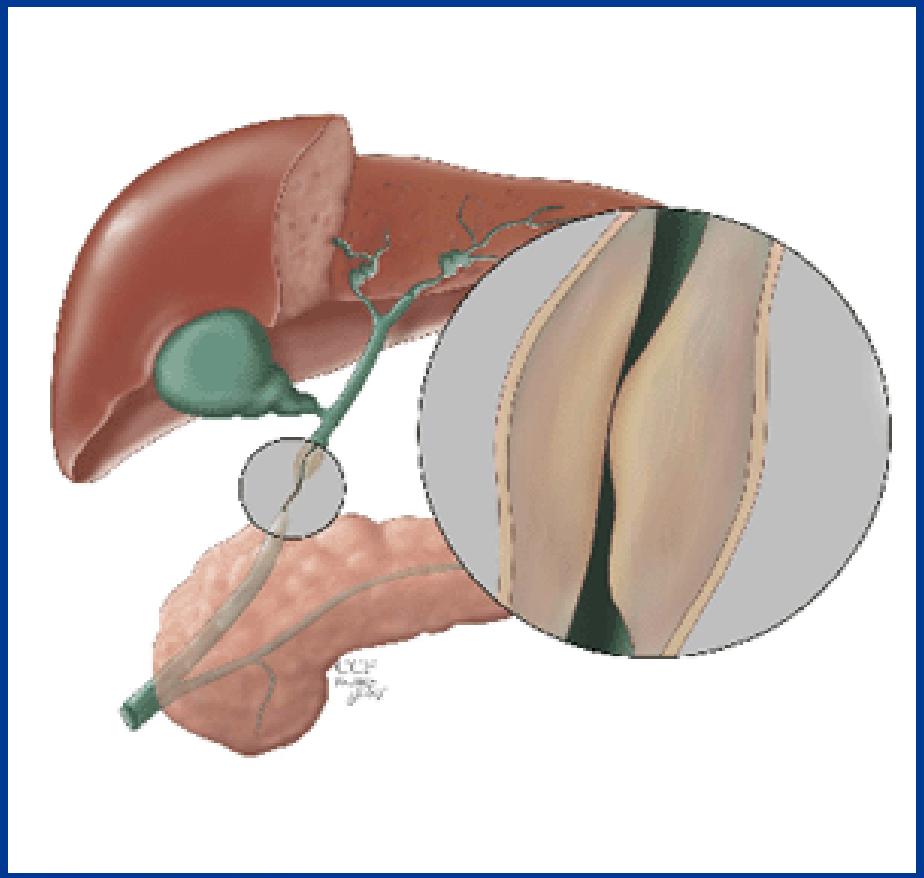
PSC – diagnostic criteria



Cholangiogram changes

- Strictures
- Saccular dilatation
- Beading
- Diverticulum

Location

- 10% intrahepatic ducts
- 15% extrahepatic ducts
- 75% combined

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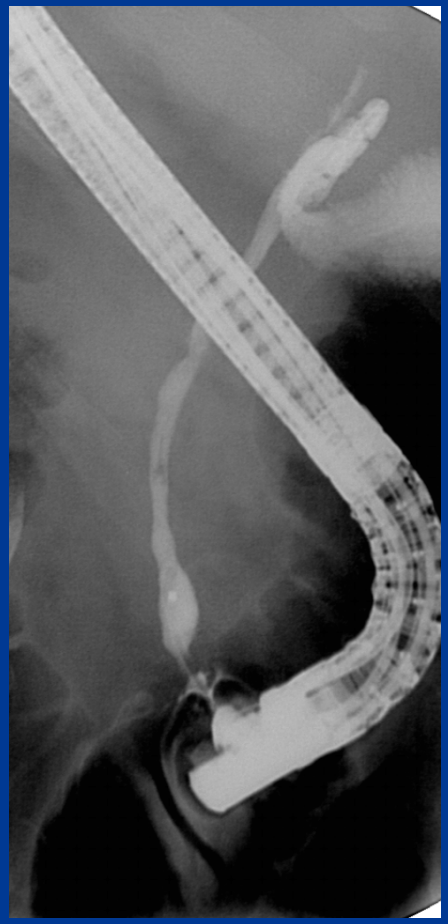
PSC – extrahepatic bile duct



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PSC – intrahepatic ducts

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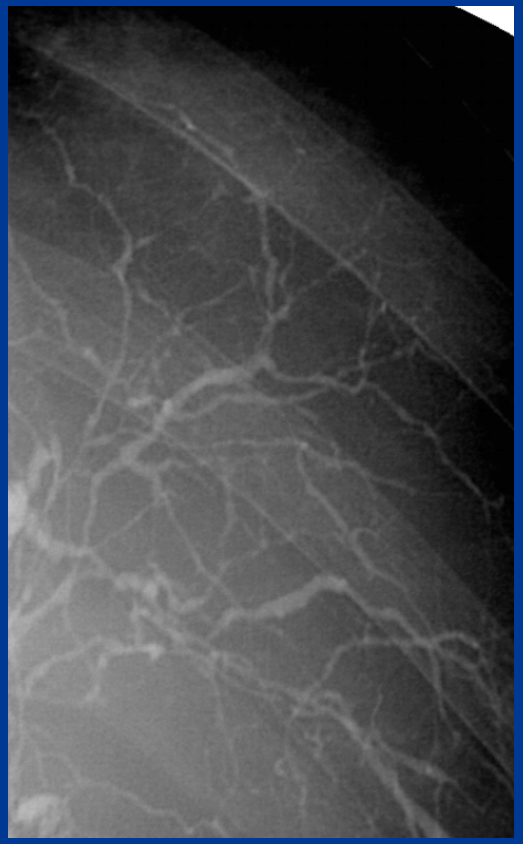
PSC – intra and extrahepatic ducts



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PSC – special subgroups

- **Small Duct PSC**
- **PSC- Autoimmune Hepatitis Overlap**

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Small duct PSC

- 5% of PSC cases
- Diagnostic criteria
  - Chronic cholestatic liver disease
  - Liver biopsy compatible with PSC
  - Normal cholangiogram
  - Other causes of liver disease excluded

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Small duct PSC

- Limited progression to large duct PSC (23%)
- Better prognosis
  - Less risk for cholangiocarcinoma
  - Longer interval to transplantation

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PSC - Autoimmune Hepatitis Overlap

- Less than 10% of PSC patients
- ? More common in younger patients
- Symptomatic presentation
- Circulating autoantibodies more common (ANA, SMA, pANCA)

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PSC - Autoimmune Hepatitis Overlap

- **Diagnostic criteria**
  - **Cholangiogram compatible with PSC**
  - **Liver biopsy – more pronounced inflammatory portal tract infiltrate and interface hepatitis vs. PSC**
- **Responds to immunosuppressive therapy – symptoms, biopsy, and cholangiogram**

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PSC – role of liver biopsy

- **Not required in all cases**
- **Confirm diagnosis and assess stage of fibrosis**
- **Required to exclude overlap with autoimmune hepatitis**
- **Required to diagnose small duct PSC**

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PSC - complications

- Stones
- Dominant stricture
- Cholangiocarcinoma
- Cholangitis

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PSC - complications

- Often identified by change in clinical status
  - Abdominal pain
  - Fever
  - Jaundice
  - Rising alk. Phos. , bilirubin
- Accelerate progression of disease

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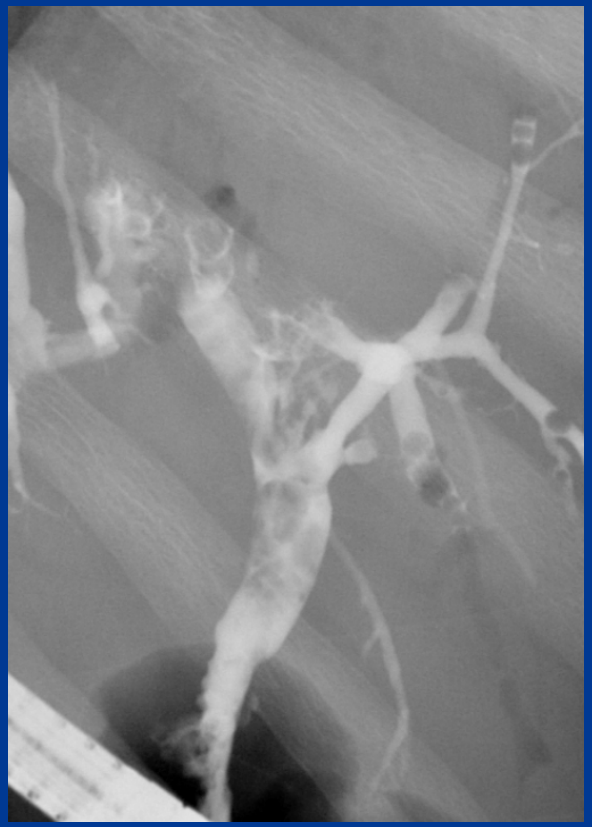
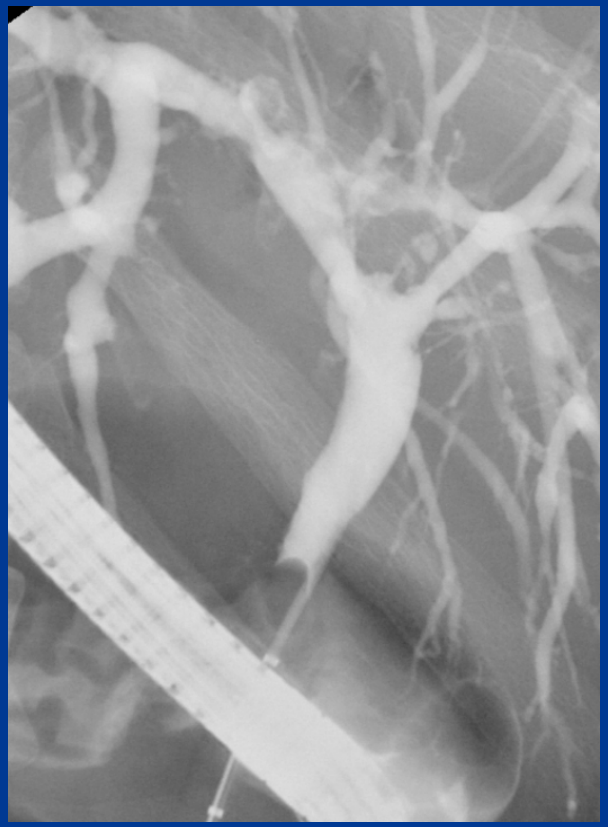
PSC + bile duct stones





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PSC + dominant stricture

- Develops in 10-15 % of patients
- Discrete narrowing in the extrahepatic bile duct
- 80% respond to endoscopic therapy
- Similar presentation for cholangiocarcinoma
- Response to endotherapy may differentiate

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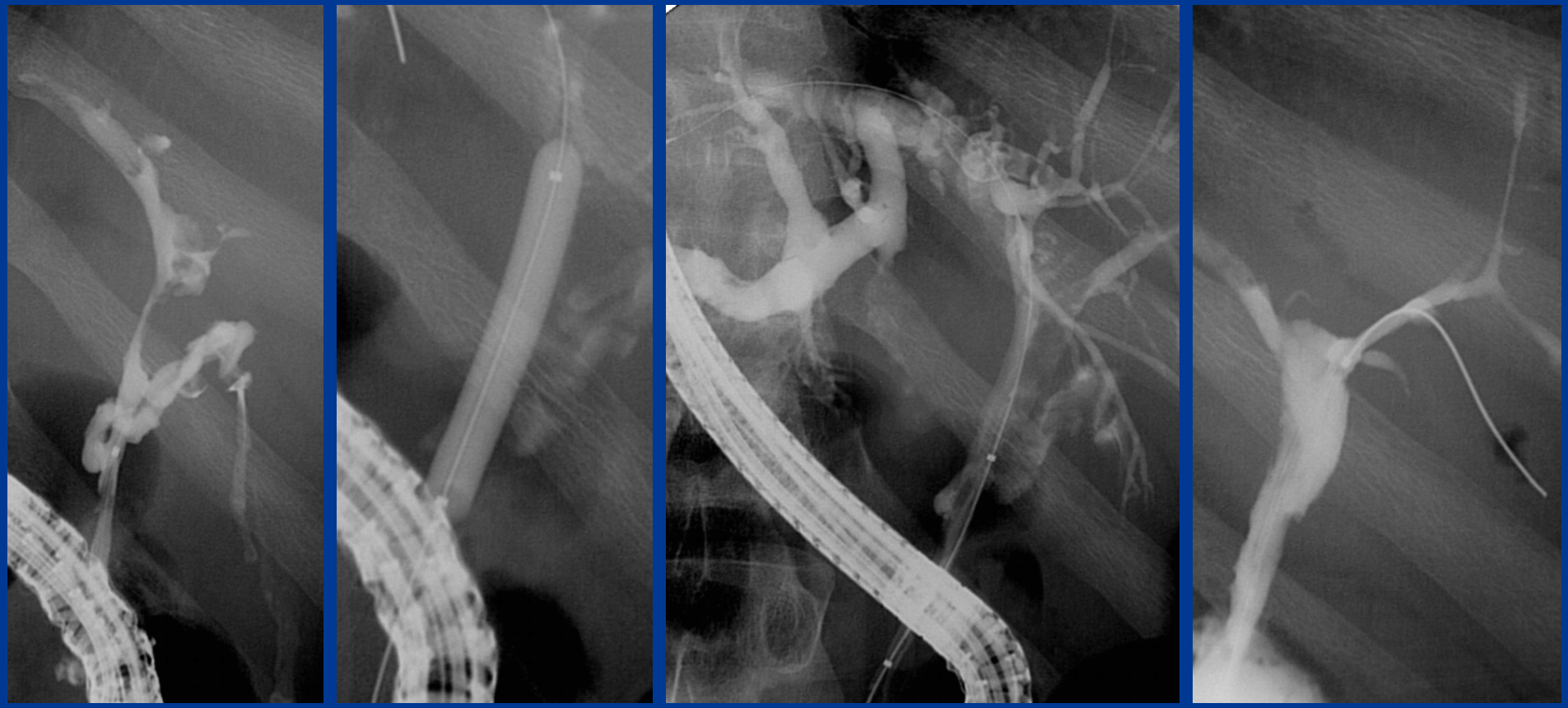
PSC + dominant stricture



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PSC + dominant stricture

- Balloon dilatation may suffice.
- Long term stenting should be avoided
  - bacterial contamination
  - stone formation
- Exchange every 4-8 weeks until resolution
- Retrospective data suggests endoscopic therapy improves survival.

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PSC + Cholangiocarcinoma

- Develops in 0.6-1.5% pts/year
- 10% lifetime risk
- No correlation with duration of disease or presence of cirrhosis
- Most commonly presents with change in status
  - pain, fever, rising liver enzymes, bilirubin, CA19-9
- Occasionally an incidental finding at ERCP or transplant

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Cholangiocarcinoma - diagnosis

- **CA19-9**
- **CA19-9/CEA index**
- **MRI / MRCP**
- **ERCP with tissue sampling**
- **Cholangioscopy with tissue sampling**
- **Positron Emission Tomography (PET scan)**

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PSC + Cholangiocarcinoma on ERCP

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Cholangiocarcinoma - tissue sampling

- Negative cytology does not exclude tumor
- Special techniques may enhance accuracy
  - Immunocytochemistry for K-*ras*, *p*53
  - Fluorescent In Situ Hybridization
  - Digital Image Analysis

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PSC + Cholagiocarcinomna - therapy

- **Stents placed across an obstructing tumor can improve the clinical status temporarily**
- **Chemotherapy, radiation, and surgery alone or in combination do not improve prognosis**
- **Chemotherapy + radiation prior to transplantation provides best opportunity for long term survival.**

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PSC + Cholangiocarcinomna - therapy

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PSC + Cholangiocarcinoma - surveillance

- **No clear evidence of benefit**
- **Serial CA 19-9 is a practical approach**
- **Serial ERCPs with tissue sampling increases risk of complications.**

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ERCP in PSC - risks

- Complications in up to 10% of procedures
- Pancreatitis, cholangitis, perforation, bleeding
- Minimize with
  - proper indication for procedure
  - pre-procedure antibiotic
  - avoid pancreatic duct injection
  - proper indication for therapy

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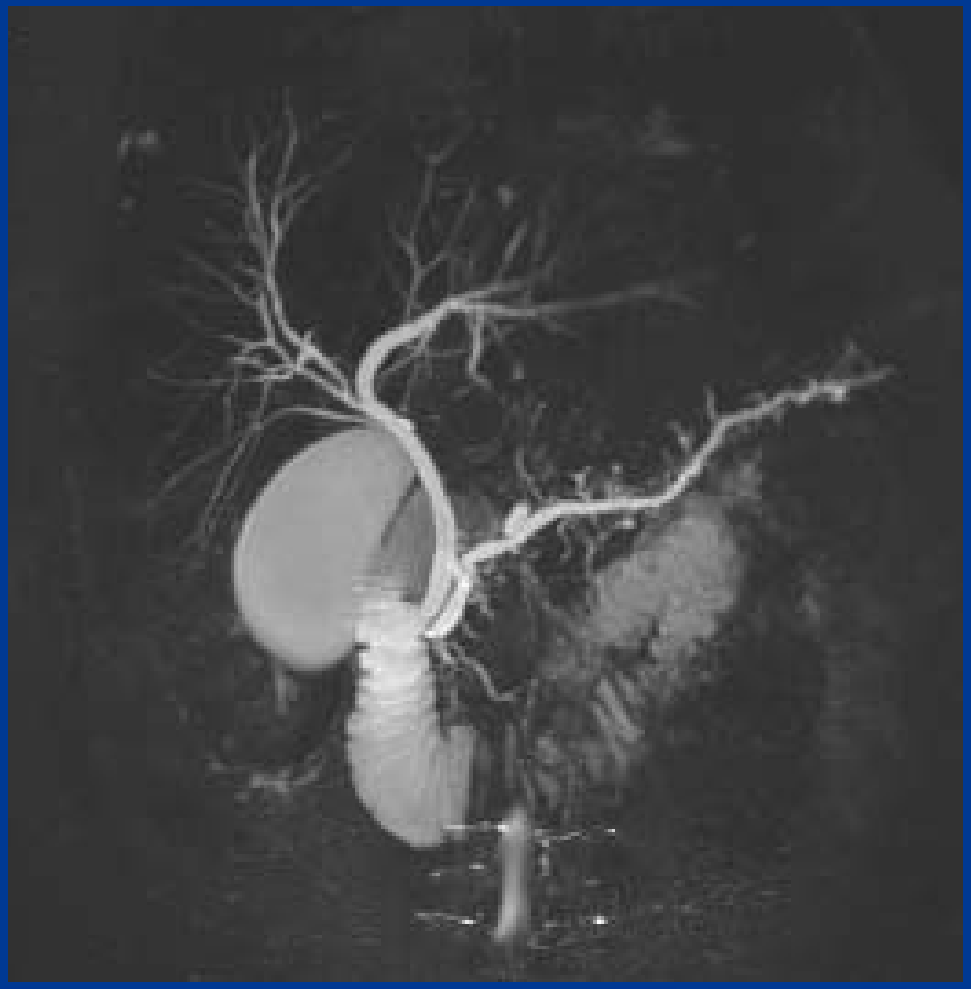
MRCP - normal



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MRCP in PSC

PRO

- No sedation, scope insertion, duct manipulation, contrast exposure, radiation
- Visualize liver parenchyma, and abdominal structures
- Applicable with altered anatomy
- Visualize ducts above obstruction

CON

- Less sensitive in defining bile duct abnormalities
- No tissue sampling
- No therapeutic options

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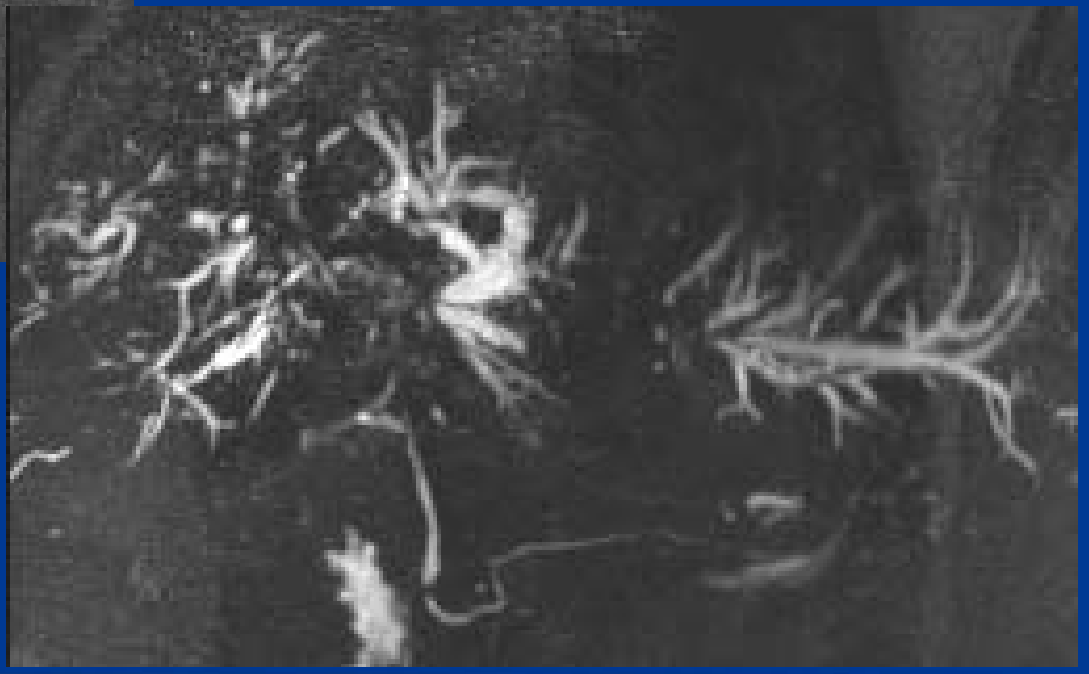
PSC on MRCP



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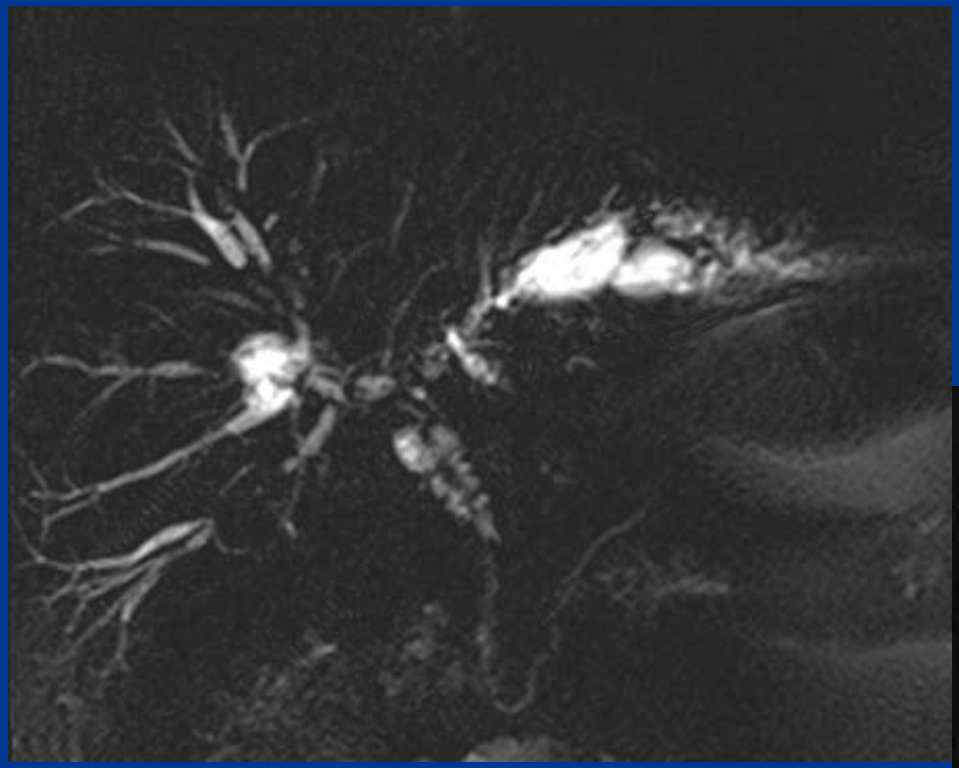
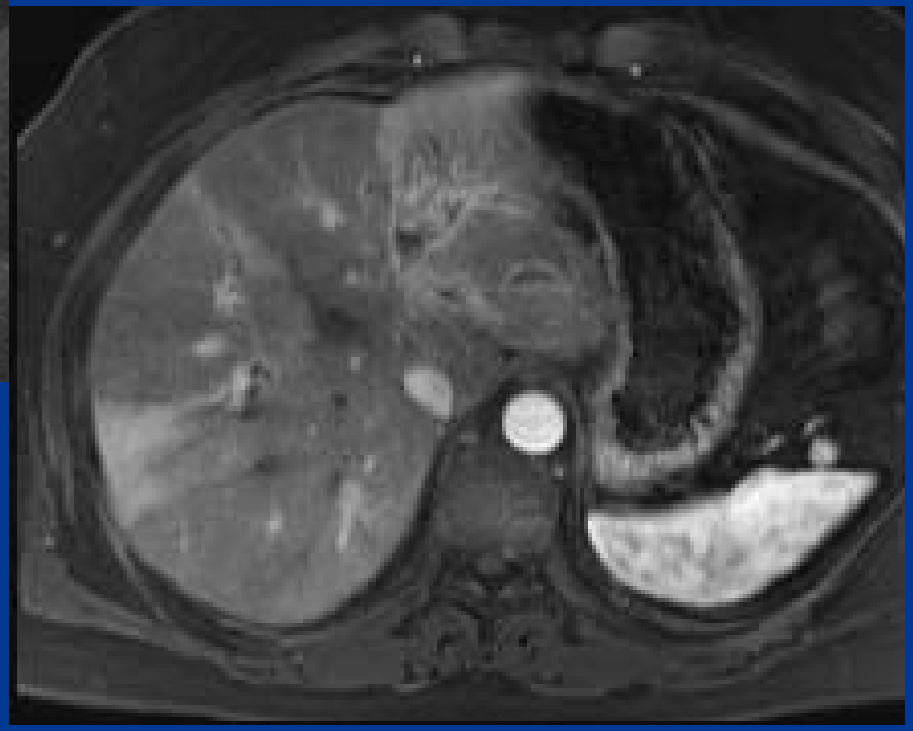


PSC on MRCP

Strictures, abnormal duct walls & peribiliary enhancement of active cholangitis



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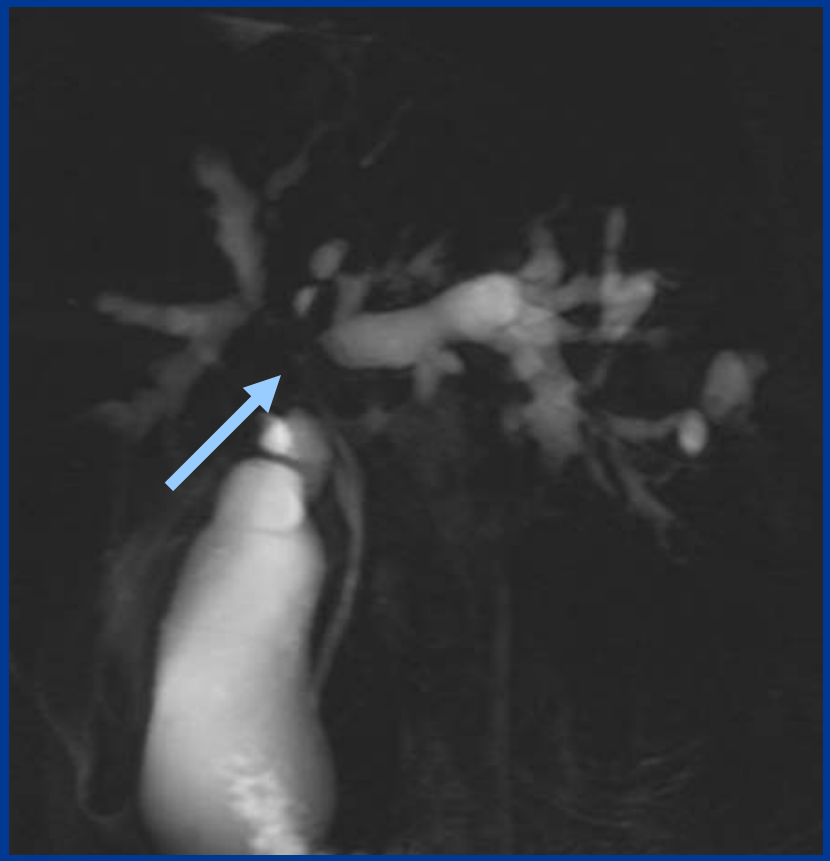
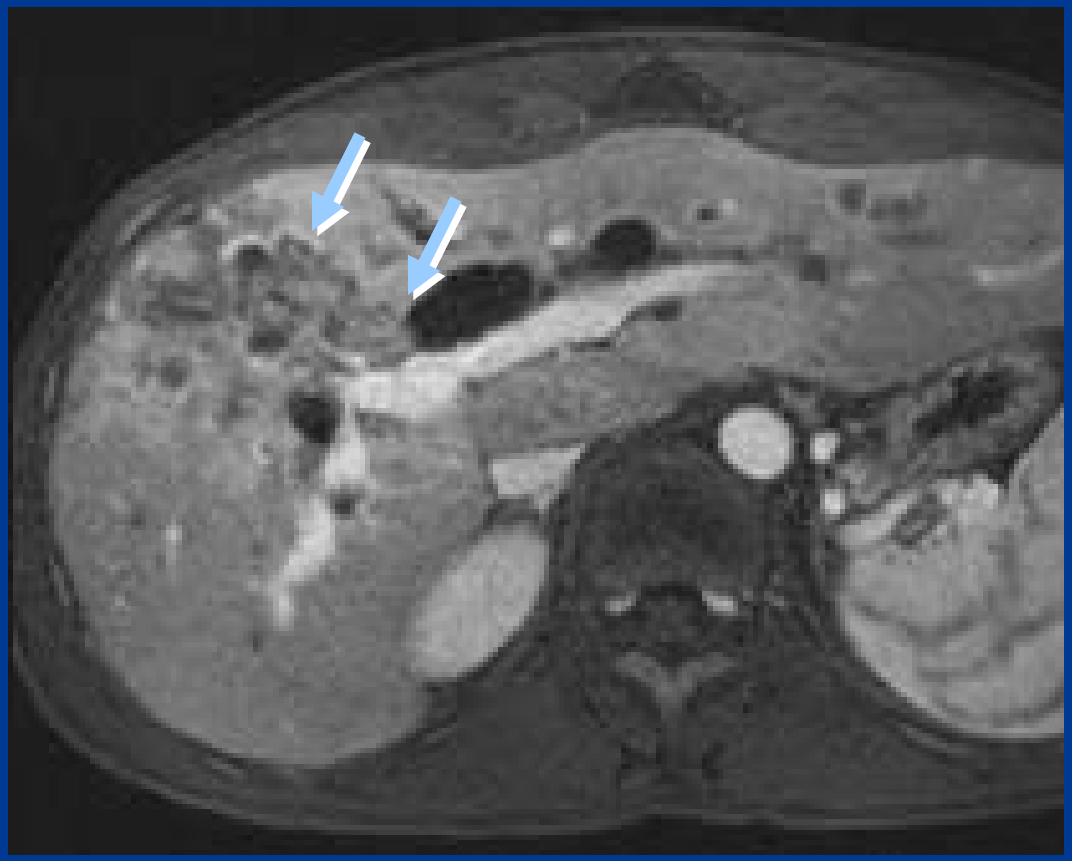
PSC with Cholangiocarcinoma





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MRCP in PSC

- **Overall best initial option for diagnosis**
- **May not avoid subsequent ERCP**
- **Compliments ERCP when evaluating dominant stricture or cholangiocarcinoma**

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PSC - Summary role of ERCP

- **Required for diagnosis but role changing with refinement and experience with MRCP**
- **Required to fully evaluate change in clinical status**
- **Provides options for tissue sampling**
- **Provides treatment options for complications**
- **Risks significant but can be limited proper case selection , preparation, and proper indications for therapeutic intervention**

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Thank You !!!