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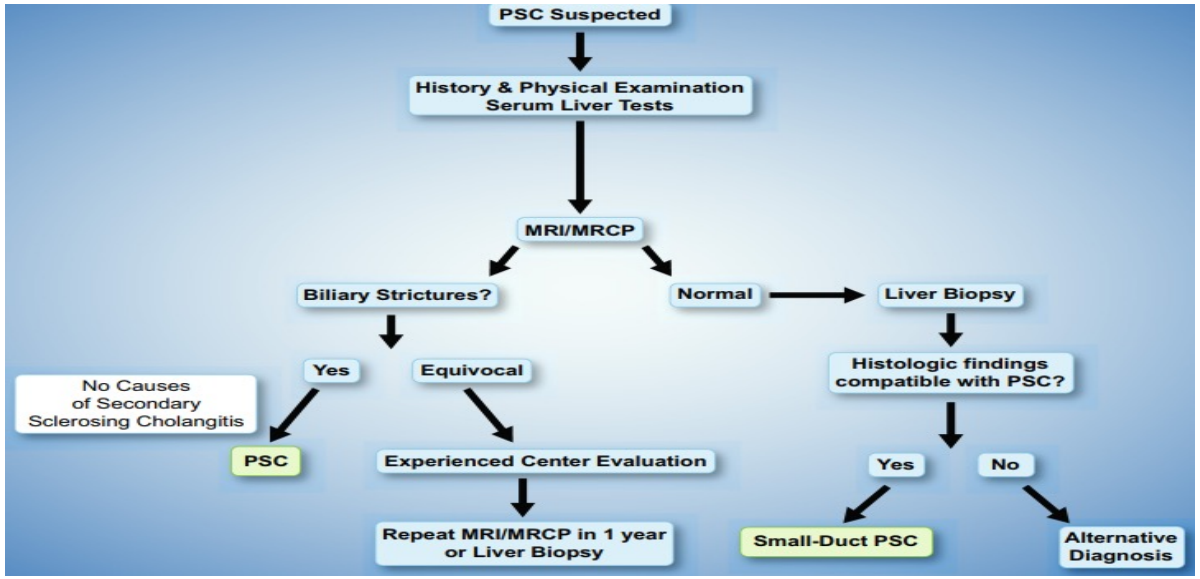
# AASLD Practice Guidance on Primary Sclerosing Cholangitis (PSC) and Cholangiocarcinoma – Part I on PSC

Infographics Creator – Cindy Ye

## Epidemiology and Symptoms

- **At least 70-80%** of patients with PSC have **IBD**.
- PSC-AIH overlap occurs in up to 5% of adults.
- Symptoms include:
  - Fatigue and **pruritus (can occur in absence of biliary obstruction).**
  - Anxiety/Depression
  - Abdominal and fever

## Diagnosis




- Consider in all patients with ↑ ALP +/- GGT.
- Exclude secondary sclerosing cholangitis .
- **Measure serum IgG4** to exclude IgG4-sclerosing cholangitis.
- **Obtain MRI/MRCP** with contrast enhancement.
  - Normal → consider liver biopsy to rule out small duct PSC.
  - Equivocal → consider repeating MRCP or liver biopsy.
- **Avoid using ERCP** for diagnosis.
- Do not perform liver biopsy for confirmation except if there is a concern of AIH/PSC overlap.
- **Perform ileocolonoscopy** with biopsies in patients with new diagnosis of PSC without concurrent IBD.

## Surveillance

- MRI/MRCP every 3-5 years for small-duct PSC (to look for development of large duct disease).
- **Annual MRI/MRCP +/- CA19-9 for cholangiocarcinoma and gallbladder carcinoma.**
- Ultrasound every 6 months if have gallbladder polyps ≤ 8 mm.
- Cholecystectomy if have gallbladder polyps >8 mm.
- HCC surveillance only if PSC with cirrhosis.
- **Colonoscopy every 1-2 years if PSC with IBD.**

## Fibrosis Staging

- Should be done at diagnosis of PSC and regularly during follow-up but optimal frequency is unclear.
- **Transient elastography or MRE =** preferred method for measuring liver stiffness.
- Do not recommend liver biopsy for fibrosis staging.

Bowlus CL, Arrivé L, Bergquist A, Deneau M, Forman L, Ilyas SI, Lunsford KE, Martinez M, Sapisochin G, Shroff R, Tabibian JH, Assis DN. AASLD practice guidance on primary sclerosing cholangitis and cholangiocarcinoma. Hepatology. 2022 Sep 9. doi: 10.1002/hep.32771. Epub ahead of print. PMID: 36083140.

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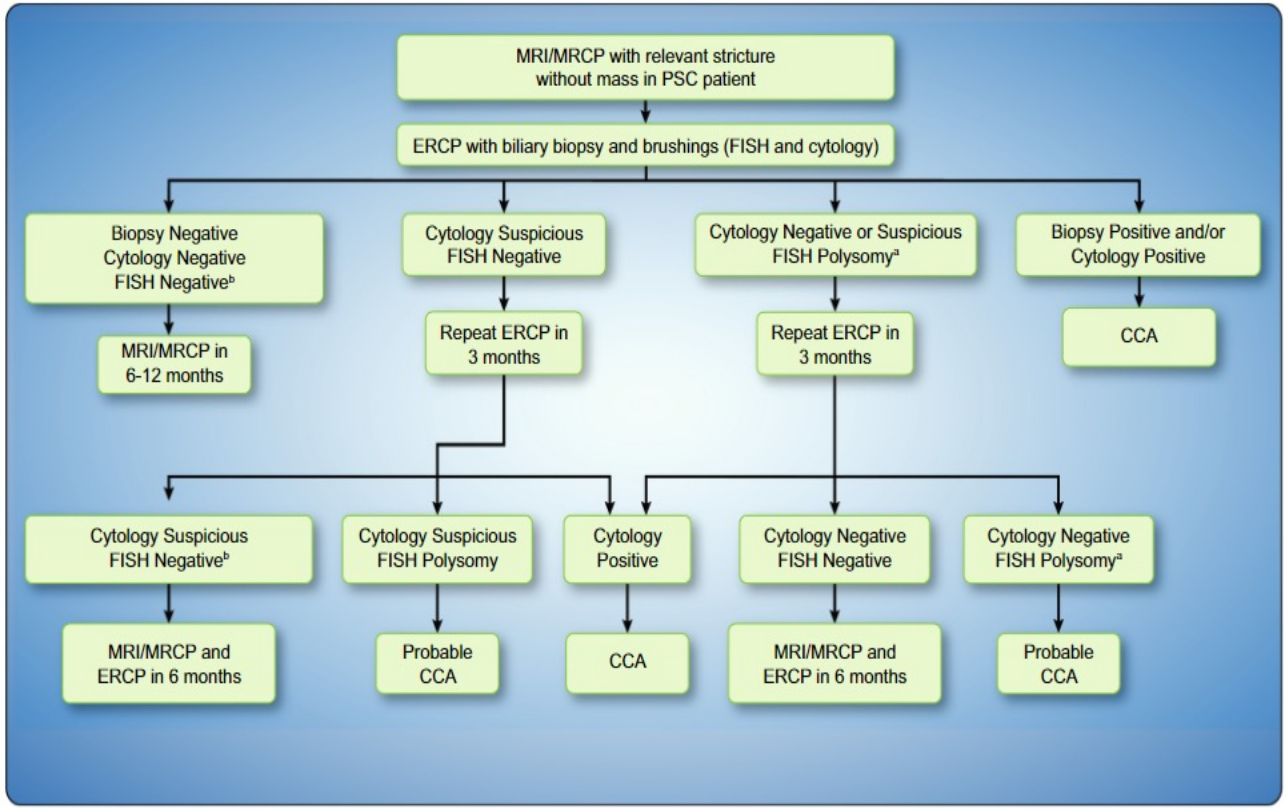
## ERCP Considerations

- **Indications for ERCP:**
  - New or worsening pruritus
  - Weight loss
  - ↑ liver enzymes
  - ↑ CA 19-9
  - Recurrent bacterial cholangitis
  - ↑ bile duct dilation
- **Need prophylactic antibiotics during periprocedural period.**
- Biliary balloon dilation +/- stenting up to discretion of the individual endoscopist.
- If plastic stent placed, usually remove within 4 weeks.
- **Sample intraductal tissue for cytology and FISH for relevant strictures.**




## Liver Transplant Considerations

- **Indications for transplant**:
  - PSC with cirrhosis
  - Recurrent cholangitis
  - Intractable pruritus
  - Early-stage hepatobiliary malignancies
- If liver enzymes ↑ post transplant, consider recurrent PSC versus rejection or biliary complications

## Management

- Consider **ursodeoxycholic acid** (UDCA, 13-23 mg/kg/day) for treatment in patients with consistently elevated ALP or GGT.
- Can continue UDCA if there is a ↓ (ALP < 1.5 x ULN, 40% reduction of ALP) or normalization of ALP or improvement of symptoms with 12 months of treatment.
- **No role for oral vancomycin** given insufficient evidence.
- Treat PSC/AIH overlap per AIH guidelines.
- Screen for varices if the liver stiffness is >20 kPA by transient elastography or the platelet count is ≤ 150,000/mm³.

- Consider **bile acid sequestrants** for pruritus if no improvement with measures such as anti-histamines, heat avoidance, or emollients.
- **Alterative therapy for pruritus** include sertraline 100 mg daily, naltrexone titrated to a dose of 50-100 mg daily, and rifampin 150-300 mg twice daily.
- Annual serum measurements of **Vit A, D, E, and K**
- **DEXA scan** at diagnosis and every 2-3 years based on risk factors.

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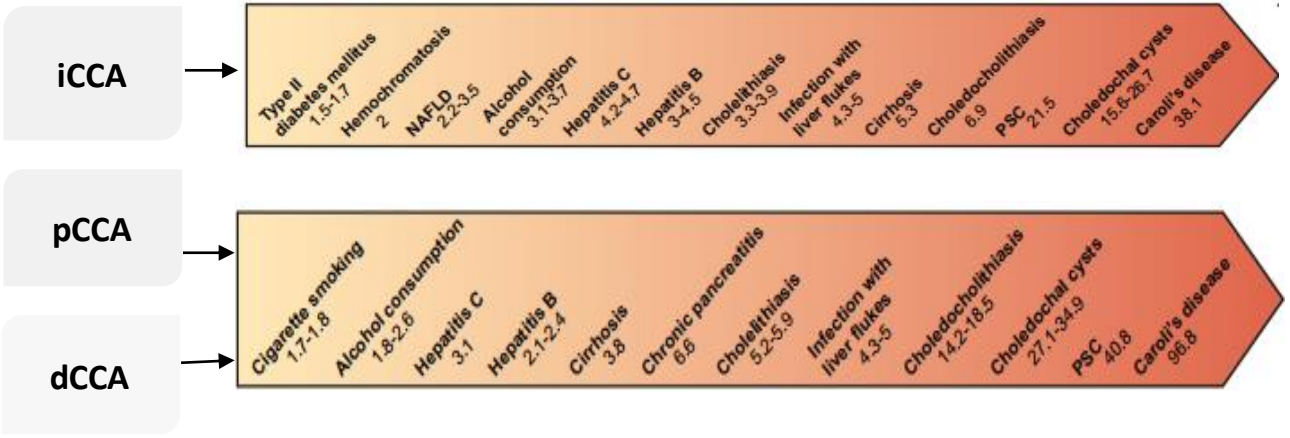
# AASLD Practice Guidance on Primary Sclerosing Cholangitis (PSC) and Cholangiocarcinoma (CCA) – Part II on CCA

Infographics Creator – Cindy Ye

## Types of CCA

| | |
|---|---|
| **Intrahepatic CCA (iCCA)** | • Arises proximal to second-order bile ducts within the hepatic parenchyma. |
| **Perihilar CCA (pCCA)** | • Arises between second-order bile ducts and the cystic duct insertion. |
| **Distal CCA (dCCA)** | • Arises in the common bile duct (CBD) below the cystic duct insertion. |

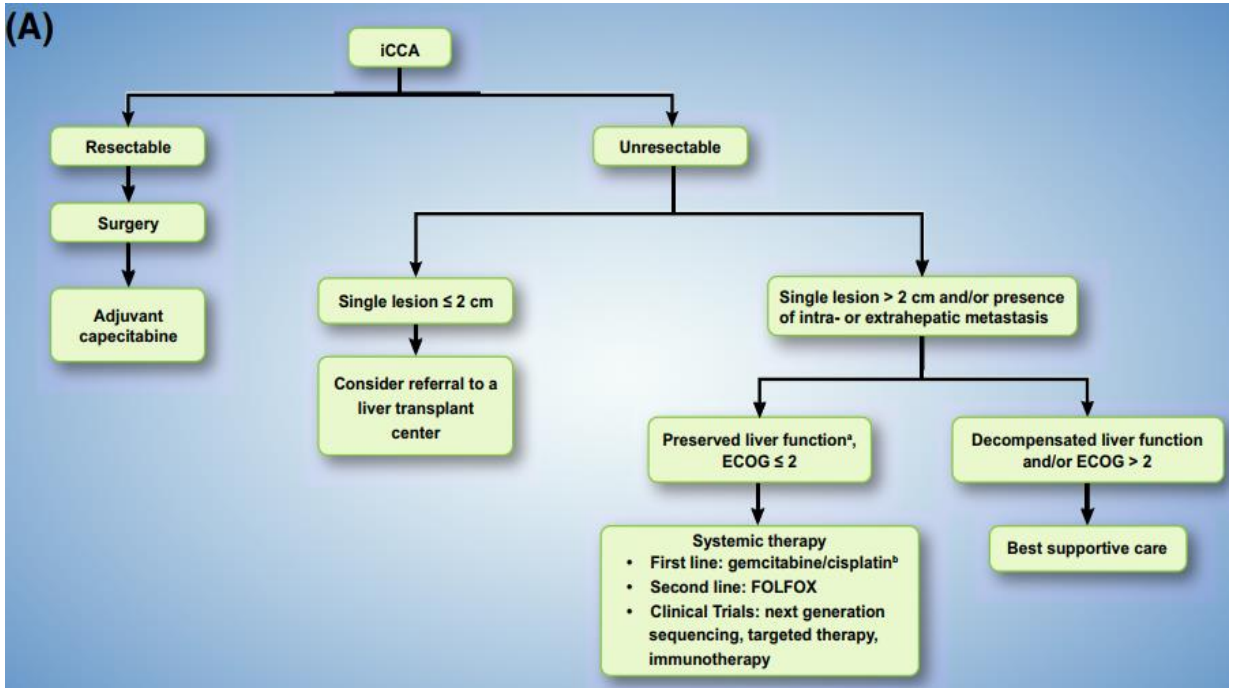
## Risk factors for CCA




## iCCA Work-Up

- CA 19-9 **not sufficient alone** for diagnosis. Levels > 1000 U/ml may allude to metastatic disease.
- **Multiphasic CT** and **MRI** are needed for assessing primary mass, detecting metastases, and staging disease.
- Requires **biopsy** for definitive diagnosis.

## iCCA Management




- Insufficient data to recommend liver directed therapies

## pCCA Work-Up

- **Multiphasic CT and MRI** are needed for assessing primary mass and vascular encasement.
- **Obtain CA 19-9** and consider IgG4 levels to exclude IgG4 sclerosing cholangiopathy.
- **Requires ERCP** with biliary brushings for cytology and FISH analysis.
- **Avoid EUS guided FNA** for diagnosis due to risk of tumor dissemination precluding liver transplant (LT).

Bowlus, Christopher L.[*,1]; Arrivé, Lionel[2]; Bergquist, Annika[3]; Deneau, Mark[4]; Forman, Lisa[5]; Ilyas, Sumera I.[6]; Lunsford, Keri E.[7]; Martinez, Mercedes[8]; Sapisochin, Gonzalo[9]; Shroff, Rachna[10]; Tabibian, James H.[11]; Assis, David N.[12]. AASLD practice guidance on primary sclerosing cholangitis and cholangiocarcinoma. Hepatology ():p n/a, October 20, 2022. | DOI: 10.1002/hep.32771

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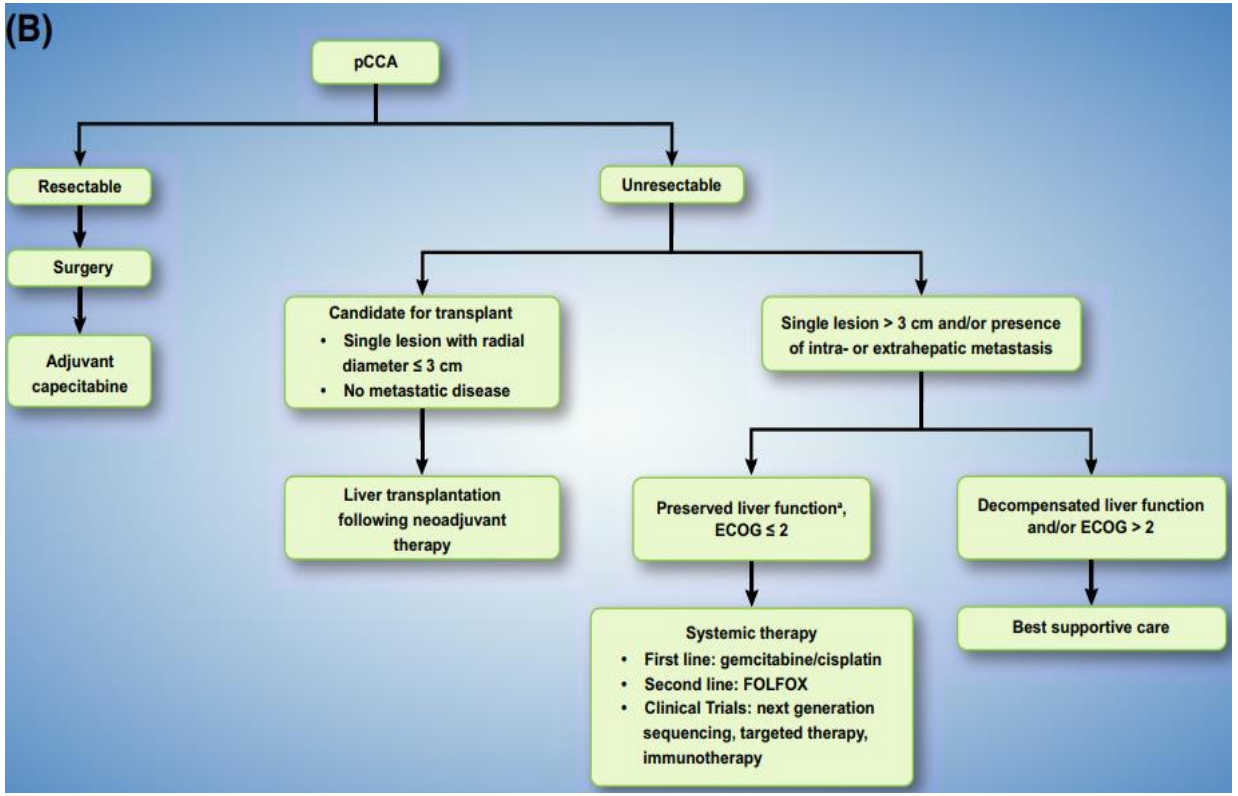
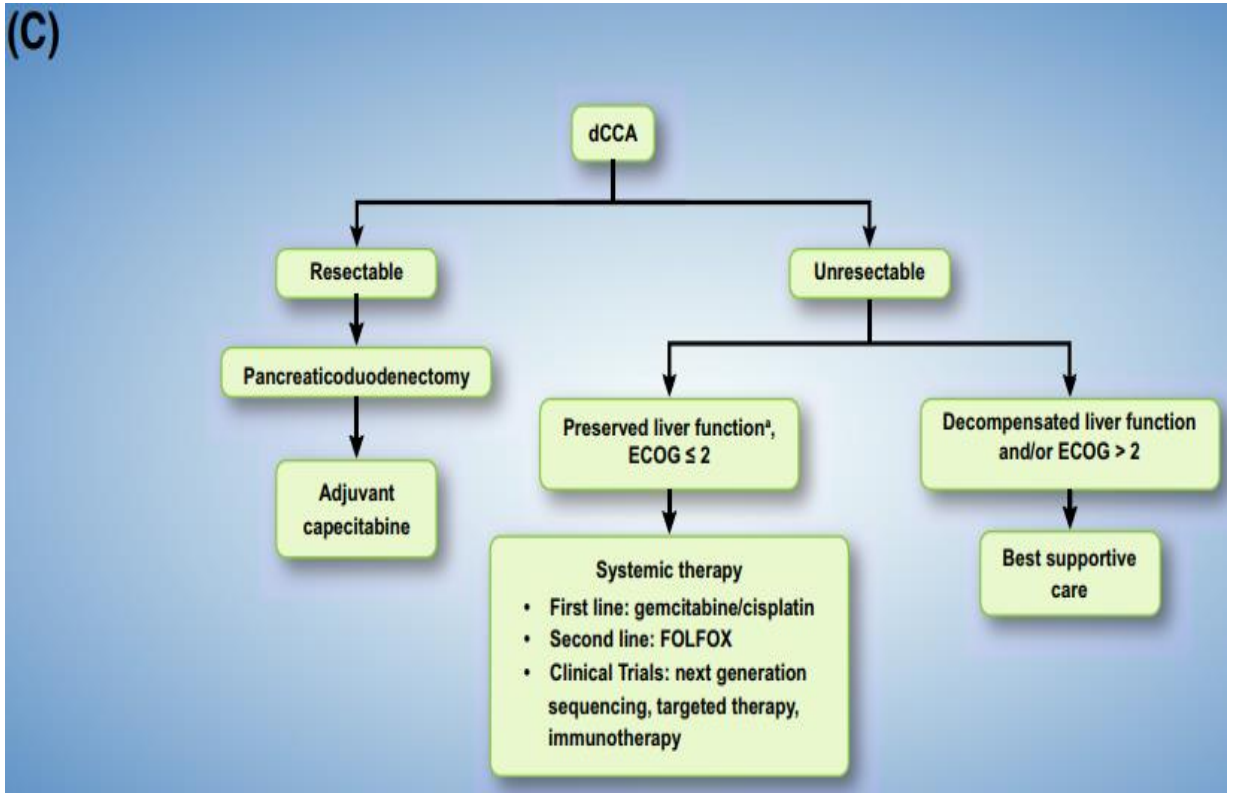
## pCCA Management




## dCCA Management




- Need **preoperative endoscopic biliary drainage** of remnant liver if obstruction is present for both pCCA and dCCA.

## dCCA Work-Up

- **Multiphasic CT and MRI** are needed for assessing primary mass and vascular encasement.
- **Obtain CA 19-9** and consider IgG4 levels to exclude IgG4 sclerosing cholangiopathy.
- **Requires ERCP** with biliary brushings for cytology and FISH analysis **and EUS** with FNA for detailed examination of the extrahepatic bile duct and tissue acquisition. EUS-FNA has higher sensitivity of detection for dCCA than pCCA.

## Systemic Therapy

- First line for advanced CCA.
- Gemcitabine/cisplatin is the first line treatment.
- FOLFOX is the second line treatment.
- Consider referral to centers with expertise in hepatobiliary malignancies and clinical trials.

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