Primary Sclerosing Cholangitis & Cholangiocarcinoma

PSC diagnosis, risk stratification, medical/endoscopic management, malignancy surveillance, and diagnosis/treatment of intrahepatic and perihilar/distal cholangiocarcinoma.

2022 AASLD Practice Guidance
Primary Sclerosing Cholangitis and Cholangiocarcinoma
Bowlus et al. · Hepatology 2023;77:659–702

FORMAT *53 numbered guidance statements — narrative consensus format (not GRADE-rated); grouped by clinical theme.*

1 Diagnosis of PSC

GS	STATEMENT
1	Suspected PSC: 3D MRI/MRCP (T1w/T2w axial + contrast) to evaluate cholangiographic features (strictures alternating with normal/dilated segments)
2	Normal high-quality MRI/MRCP: consider biopsy to rule out small-duct PSC; equivocal MRI/MRCP → refer to experienced center for repeat imaging or biopsy (repeat MRI/MRCP at 1y if still unclear)
3	Avoid ERCP for PSC diagnosis
4	Measure serum IgG4 in all suspected PSC to exclude IgG4-sclerosing cholangitis
5	No biopsy needed with typical cholangiographic findings, except if AIH overlap suspected
6	Ileocolonoscopy with biopsies at new PSC diagnosis if no prior IBD diagnosis; repeat at 5y intervals or with symptoms

2 Risk Stratification & Fibrosis Staging

GS	STATEMENT
7	Small-duct PSC: monitor with MRI/MRCP every 3–5y for large-duct progression
8	Risk stratification/fibrosis staging at diagnosis and regularly during follow-up; interpret clinical risk tools cautiously
9	Liver stiffness (TE or MRE) is the preferred method for fibrosis staging
10	Biopsy not recommended for fibrosis staging in clinical practice

3 Medical Management & Complications

GS	STATEMENT
11	Consider all patients for clinical trial participation
12	Not trial-eligible/interested + persistently elevated ALP/GGT: UDCA 13–23 mg/kg/day; continue if meaningful ALP reduction/normalization or symptom improvement at 12 months
13	Insufficient evidence for oral vancomycin in PSC
14	Concurrent AIH: treat per AASLD AIH guidelines
15	Antibiotics for bacterial cholangitis; consider MRCP to rule out relevant strictures
16	ERCP for bacterial cholangitis with inadequate antibiotic response
17	Variceal screening EGD if liver stiffness >20 kPa (TE) or platelets ≤150,000/mm³

4 Surveillance for Malignancy

GS	STATEMENT
18	Annual CCA/gallbladder carcinoma surveillance: abdominal imaging (preferably MRI/MRCP) ± CA 19-9; not needed if <18y or small-duct PSC
19	Routine intraductal tissue sampling (cytology + FISH) during ERCP for relevant strictures
20	Cholecystectomy for gallbladder polyps >8 mm (experienced center if advanced disease); polyps ≤8 mm → US every 6 months
21	HCC surveillance in PSC-cirrhosis per standard AASLD guidelines
22	PSC-IBD: high-definition surveillance colonoscopy with biopsies starting age 15y, repeated q1–2y for colonic dysplasia

5 Endoscopic (ERCP) Management of Strictures

GS	STATEMENT
23	ERCP indicated for relevant strictures, new/worsening pruritus, unexplained weight loss, worsening LFTs, rising CA 19-9, recurrent cholangitis, or progressive duct dilation; MRI/MRCP first to guide need/approach
24	Periprocedural antimicrobial prophylaxis for ERCP in PSC
25	Balloon dilation ± stenting per endoscopist discretion; remove plastic stents within 4 weeks

6 Symptoms, Nutrition, Bone Health & Transplantation

GS	STATEMENT
26	Bile acid sequestrants first-line for pruritus unresponsive to conservative measures; refractory options: sertraline 100 mg/d, naltrexone 50–100 mg/d, or rifampin 150–300 mg BID
27	Manage IBD as in non-PSC patients; continue IBD management/CRC surveillance post-transplant
28	Nutritional assessment (biometrics, lipid-soluble vitamins) at diagnosis and yearly; supplement as needed
29	Bone density exam at diagnosis and every 2–3y based on risk factors
30	Consider LT for end-stage liver disease complications, recurrent cholangitis, intractable pruritus, or early-stage hepatobiliary cancers
31	Post-transplant elevated LFTs: histologic + cholangiographic assessment to distinguish recurrent PSC from rejection/biliary complications

7 Intrahepatic Cholangiocarcinoma (iCCA)

GS	STATEMENT
32	CA 19-9 elevation alone is insufficient to diagnose CCA
33	Histopathological confirmation required for definitive iCCA diagnosis
34	Multiphasic CT/MRI required to assess primary mass, vascular invasion, metastasis, resectability
35	Chest + abdomen cross-sectional imaging required for staging
36	PET not used for primary tumor diagnosis
37	Surgical resection = treatment of choice for single resectable iCCA nodule, no metastases, adequate FLR
38	Refer iCCA to a center with hepatobiliary surgical expertise
39	Consider adjuvant capecitabine for all CCA patients
40	LT for unresectable liver-limited iCCA only under research protocols
41	Insufficient data for LRT as standard therapy for locally advanced unresectable iCCA

8 Perihilar/Distal CCA (pCCA/dCCA)

GS	STATEMENT
42	Cross-sectional imaging + cholangiographic studies required for suspected pCCA/dCCA (tumor extent, mass, enhancement, vascular encasement)
43	ERCP with biliary brushings (cytology + FISH) for suspected pCCA/dCCA
44	Avoid EUS-FNA/percutaneous biopsy of perihilar mass in pCCA (tumor seeding risk)
45	Preoperative endoscopic biliary drainage of remnant liver if obstruction present before pCCA/dCCA resection
46	Surgical resection = treatment of choice for early-stage pCCA/dCCA without metastases
47	Refer pCCA/dCCA to a center with hepatobiliary surgical expertise
48	LT after neoadjuvant therapy for pCCA ≤3 cm that is unresectable or PSC-associated
49	Pre-LT: EUS-FNA of regional LNs to exclude metastases before neoadjuvant therapy; operative staging required after neoadjuvant therapy/before LT

9 Systemic Therapy for Advanced CCA

GS	STATEMENT
50	Systemic chemotherapy = first-line for advanced CCA; gemcitabine/cisplatin is standard of care
51	FOLFOX appropriate second-line after gem/cis progression
52	Consider NGS at diagnosis to guide second-line treatment
53	Refer advanced CCA to a center with hepatobiliary expertise and clinical trial access

Bowlus CL, Arrivé L, Bergquist A, et al. AASLD 2022 Practice Guidance on Primary Sclerosing Cholangitis and Cholangiocarcinoma. *Hepatology*. 2023;77(2):659–702.

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