Primary Biliary Cholangitis

Diagnosis, AIH overlap, UDCA/second-line therapy, pruritus/sicca management, portal hypertension/HCC screening, bone health, and liver transplantation.

2018 AASLD Practice Guidance
Primary Biliary Cholangitis
Lindor, Bowlus, Boyer et al. · Hepatology 2019;69:394

FORMAT *21 numbered guidance statements — narrative consensus format (not GRADE-rated); grouped by clinical theme.*

1 Diagnosis

GS	STATEMENT
1	Diagnose PBC when ≥2 of 3 criteria met: cholestatic ALP elevation; AMA (or sp100/gp210 if AMA-negative); histologic nonsuppurative destructive cholangitis with bile duct destruction

2 AIH Overlap & Liver Biopsy

GS	STATEMENT
2	AMA-negative PBC diagnosis doesn't require biopsy if cholestatic labs + PBC-specific autoantibodies (sp100/gp210) present
3	Consider liver biopsy to exclude concomitant AIH/other disease if ALT >5× ULN
4	Suspected PBC/AIH overlap: target treatment to the predominant histological injury pattern

3 UDCA Therapy

GS	STATEMENT
5	UDCA 13–15 mg/kg/day orally for all PBC patients with abnormal liver enzymes, regardless of histologic stage
6	If bile acid sequestrant needed, give UDCA ≥1 h before or ≥4 h after the sequestrant
7	Evaluate biochemical response to UDCA at 12 months to determine need for second-line therapy

4 Second-Line Therapy

GS	STATEMENT
8	Inadequate UDCA responders: consider obeticholic acid (OCA), starting at 5 mg/day
9	Fibrates can be considered off-label for inadequate UDCA response
10	Avoid OCA and fibrates in decompensated liver disease (Child-Pugh-Turcotte B or C)

5 Pruritus Management

GS	STATEMENT
11	Anion-exchange resins as initial therapy for PBC-related pruritus
12	Refractory pruritus: rifampicin 150–300 mg BID, naltrexone titrated to 50 mg/day, or sertraline 75–100 mg/day

6 Sicca Syndrome Management

GS	STATEMENT
13	Dry eyes: artificial tears first; pilocarpine/cevimeline if refractory; cyclosporine/lifitegrast ophthalmic emulsion under ophthalmology if still refractory
14	Xerostomia/dysphagia: OTC saliva substitutes first; add pilocarpine/cevimeline if symptoms persist

7 Portal Hypertension & HCC Screening

GS	STATEMENT
15	Suspected cirrhosis: endoscopic variceal screening at time of diagnosis
16	HCC surveillance q6mo with cross-sectional imaging for men and all patients with cirrhosis

8 Bone Health & Hyperlipidemia

GS	STATEMENT
17	1,000–1,500 mg calcium + 1,000 IU vitamin D daily (diet + supplements as needed)
18	Osteoporotic patients: oral alendronate 70 mg weekly or other bisphosphonate; avoid if reflux or known varices
19	Elevated lipids + cardiovascular risk: consider lipid-lowering therapy

9 Fat-Soluble Vitamins

GS	STATEMENT
20	Treat fat-soluble vitamin (A, D, E, K) deficiencies with parenteral or water-soluble supplements

10 Liver Transplantation

GS	STATEMENT
21	Refer for LT when MELD score exceeds 14 in end-stage PBC (also consider: decompensated cirrhosis, bilirubin >6 mg/dL, Mayo risk score >7.8, or intractable pruritus)

Lindor KD, Bowlus CL, Boyer J, Levy C, Mayo M. AASLD Practice Guidance: Primary Biliary Cholangitis. *Hepatology*. 2019;69(1):394–419.