Primary Sclerosing Cholangitis

Diagnosis, UDCA therapy, dominant stricture management, liver transplantation, colon/cancer surveillance, AIH overlap, and symptom management.

2015 ACG Clinical Guideline
Primary Sclerosing Cholangitis
Lindor, Kowdley, Harrison · Am J Gastroenterol 2015

STRENGTH Strong Conditional

1 Diagnosis

STATEMENT	STRENGTH
R1 — MRCP preferred over ERCP to establish diagnosis of PSC	Strong Moderate
R2 — Liver biopsy not necessary if diagnostic cholangiographic findings present	Conditional Low
R3 — Liver biopsy recommended for suspected small duct PSC, or to exclude AIH overlap	Conditional Moderate
R4 — Antimitochondrial antibody testing helps exclude primary biliary cirrhosis	Conditional Moderate
R5 — Test at least once for elevated serum IgG4 levels	Conditional Moderate

2 UDCA Therapy

STATEMENT	STRENGTH
R6 — UDCA doses >28 mg/kg/day should NOT be used for PSC management	Strong High

3 Dominant Stricture Management

STATEMENT	STRENGTH
R7 — ERCP with balloon dilatation for dominant stricture with pruritus/cholangitis to relieve symptoms	Strong Low
R8 — Dominant stricture on imaging: ERCP with cytology, biopsies, and FISH to exclude cholangiocarcinoma	Strong Low
R9 — Antibiotic prophylaxis for PSC patients undergoing ERCP to prevent post-ERCP cholangitis	Conditional Low
R10 — Routine stenting after dilation not required; short-term stenting may help severe strictures	Conditional Low

4 Liver Transplantation

STATEMENT	STRENGTH
R11 — LT preferred over medical therapy/surgical drainage in decompensated cirrhosis to prolong survival	Strong Moderate
R12 — Refer for liver transplantation when MELD score exceeds 14	Conditional Moderate

5 Colon Surveillance

STATEMENT	STRENGTH
R13 — Annual colon surveillance (preferably chromoendoscopy) in PSC + colitis, starting at PSC diagnosis	Conditional Moderate
R14 — Full colonoscopy with biopsies at PSC diagnosis regardless of symptoms, to assess for colitis	Conditional Moderate
R15 — Consider repeat colonoscopy every 3–5 yr in those without prior colitis evidence	Weak Low

6 Cancer & Gallbladder Screening

STATEMENT	STRENGTH
R16 — Consider cholangiocarcinoma screening with cross-sectional imaging + CA 19-9 every 6–12 mo	Conditional Very low
R17 — Cholecystectomy for PSC with gallbladder polyps >8 mm, to prevent adenocarcinoma	Conditional Very low

7 Autoimmune Hepatitis Overlap

STATEMENT	STRENGTH
R18 — AIH testing for PSC patients <25 yr or with aminotransferases ≥5× ULN	Conditional Moderate
R19 — MRCP for AIH patients <25 yr with ALP >2× ULN	Conditional Moderate

8 Symptom Management & Screening

STATEMENT	STRENGTH
R20 — Mild pruritus: emollients and/or antihistamines	Conditional Very low
R21 — Moderate pruritus: bile acid sequestrants (e.g., cholestyramine); rifampin/naltrexone if ineffective	Conditional Very low
R22 — Screen for varices if advanced disease signs and platelets <150×10³/μL	Conditional Very low
R23 — BMD screening (DEXA) at diagnosis and every 2–4 yr	Conditional Moderate
R24 — Screen/monitor for fat-soluble vitamin deficiencies in advanced liver disease	Conditional Moderate

Lindor KD, Kowdley KV, Harrison ME. ACG Clinical Guideline: Primary Sclerosing Cholangitis. *Am J Gastroenterol.* 2015;110(5):646–659.