Shifting Paradigms in Primary Biliary Cholangitis

Guidelines

American Association for the Study of Liver Diseases

European Association for the Study of the Liver

Reviews

American Liver Foundation

BMJ Best Practice

National Institute of Diabetes and Digestive and Kidney Diseases

Purohit T, et al. Primary biliary cirrhosis: Pathophysiology, clinical presentation and therapy. *World J Hepatol*. 2015;7(7):926-941.

Trivedi PJ, et al. Review article: overlap syndromes and autoimmune liver disease. *Aliment Pharmacol Ther*. 2012;36:517-533.

Natural History Model for Primary Biliary Cholangitis

Mayo Risk Score

Obeticholic acid

Carbone M, et al. Predicted risk of end stage liver disease with continued standard of care and subsequent addition of obeticholic acid in patients with PBC. *Hepatology*. 2016;64(Suppl 1):184A. Abstract 361.

Hirschfield GM, et al. Long-term effect of obeticholic acid on transient elastography and AST to Platelet Ratio Index in patients with PBC. *Hepatology*. 2016;64(Suppl 1):110A. Abstract 209.

Hirschfield GM, et al. Efficacy of obeticholic acid in patients with primary biliary cirrhosis and inadequate response to ursodeoxycholic acid. *Gastroenterology*. 2015;148(4):751-761.

Institute for Clinical and Economic Review. Obeticholic acid for the treatment of primary biliary cholangitis: Comparative clinical effectiveness, value, and value-based price benchmarks. Evidence report- July 26, 2016.

Nevens F, et al for the POISE Study Group. A placebo-controlled trial of obeticholic acid in primary biliary cholangitis. *N Engl J Med*. 2016;375(7):631-643.

Pockros PJ, et al. Efficacy of obeticholic acid in patients with primary biliary cholangitis and renal impairment. *Hepatology*. 2016;64(Suppl 1):205A. Abstract 401.

Pockros PJ, et al. Analysis of the efficacy of OCA in primary biliary cholangitis by baseline biochemistry across three randomized, double-blind, placebo-controlled clinical trials. *Am J Gastroenterol*. 2016;111(Suppl 1):S337. Abstract 751.

Samur S, et al. Long-term clinical impact and cost-effectiveness of obeticholic acid for the treatment of primary biliary cholangitis. *Hepatology*. 2017;65(3):920-928.

Vierling JM, et al. Efficacy of obeticholic acid treatment in patients with primary biliary cholangitis with cirrhosis. *Hepatology*. 2016;64(Suppl 1):187A. Abstract 366.

Ursodeoxycholic acid

Boberg KM, et al. Cost and health consequences of treatment of primary biliary cirrhosis with ursodeoxycholic acid. *Aliment Pharmacol Ther*. 2013;38(7):794-803.

Corpechot C, et al. Relationships between biochemical response to UDCA and progression of liver stiffness as determined by Fibroscan in patients with PBC. *Hepatology*. 2016;64(Suppl 1):194A. Abstract 379.

Harms MH, et al. Association between ursodeoxycholic acid therapy and prolonged transplant-free survival among patients with primary biliary cholangitis- a number needed to treat analysis. *Hepatology*. 2016;64(Suppl 1):188A. Abstract 367.

Lemmers WJ, et al. Biochemical patterns of alkaline phosphatase and bilirubin levels in relation to clinical outcomes in UDCA-treated PBC patients- an international study. *Hepatology*. 2016;64(Suppl 1):197A. Abstract 385.

Poupon RE, et al. Combined analysis of randomized controlled trials of ursodeoxycholic acid in primary biliary cirrhosis. *Gastroenterology*. 1997;113;884.

Tang R, et al. Gut microbial profile is altered in primary biliary cholangitis and partially restored after UDCA therapy. *Gut*. 2017;dx.doi.org/10.1136/gutjnl-2016-313332.

Other articles

Abe K, et al. Interleukin-33 levels as predictors for disease remission and progression in patients with primary biliary cholangitis. *Hepatology*. 2016;64(Suppl 1):200A. Abstract 391.

Ali AH, et al. Varices in early histological stage primary biliary cirrhosis. *J Clin Gastroenterol*. 2011;45(7):e66-e71.

Carmel AS, et al. The 25(OH)D level need to maintain a favorable bisphosphonate response is ≥33 ng/ml. *Osteoporos Int*. 2012;23(10):2479-2487.

Cheung AC, et al. Combined ursodeoxycholic acid (UDCA) and fenofibrate in primary biliary cholangitis patients with incomplete UDCA response may improve outcomes. *Aliment Pharmacol Ther*. 2016;43(2):283-293.

Corpechot C. Primary biliary cirrhosis beyond ursodeoxycholic acid. *Semin Liver Dis*. 2016;36(1):15-26.

Czul F, et al. Novel therapies on primary biliary cirrhosis. *Clin Liver Dis*. 2016;20(1):113-130.

Floreani A, et al. Proposed therapies in primary biliary cholangitis. *Expert Rev Gastroenterol Hepatol*. 2016;10(3):371-382.

Griffiths L, et al. Pathogenesis of primary biliary cirrhosis and its fatigue. *Dig Dis*. 2014;32(5):615-625.

Hosonuma K, et al. A prospective randomized controlled study of long-term combination therapy using ursodeoxycholic acid and bezafibrate in patients with primary biliary cirrhosis and dyslipidemia. *Am J Gastroenterol*. 2015;110(3):423-431.

Jopson L, et al. Fatigue in primary biliary cirrhosis: Prevalence, pathogenesis and management. *Dig Dis*. 2015;33(Suppl 2):109-114.

Njei B, et al. Primary biliary cholangitis-associated hospitalizations in the US: A nationwide analysis, 2004-2012. *Hepatology*. 2016;64(Suppl 1):205A. Abstract 402.

Phillips JR, et al. Fat-soluble vitamin levels in patients with primary biliary cirrhosis. *Am J Gastroenterol*. 2001;96(9):2745-2750.

Rishe E, et al. Itch in primary biliary cirrhosis: a patients' perspective. *Acta Derm Venereol*. 2008;88(1):34-37.

Trivedi PJ, et al for the Global PBC Study Group. Stratification of hepatocellular carcinoma risk in primary biliary cirrhosis: a multicenter international study. *Gut*. 2016;65(2):321-329.

Yin Q, et al. Systematic review and meta-analysis: bezafibrate in patients with primary biliary cirrhosis. *Drug Des Devel Ther*. 2015;9:5407-5419.

Zhang Y, et al. Combination therapy of fenofibrate and ursodeoxycholic acid in patients with primary biliary cirrhosis who respond incompletely to UDCA monotherapy: a meta-analysis. *Drug Des Devel Ther*. 2015;9:2757-2766.

Zhu GQ, et al. Optimal drug regimens for primary biliary cirrhosis: a systematic review and network meta-analysis. *Oncotarget*. 2015;6(27):24533-24549.

Response Criteria Models for Ursodeoxycholic Acid

Barcelona

Biochemical + AST/Platelet Ratio Index

Early Biochemical Response

GLOBE Score

Paris-I

Paris-II

Rotterdam

Toronto

UK/PBC Risk Score