Preface

Cholestatic Liver Diseases

Over the past 2 decades, our understanding of cholestatic liver diseases has deepened. Genetic predisposition, for instance, is increasingly recognized as a major contributor to susceptibility or severity of various cholestatic diseases. In this issue of *Clinics in Liver Diseases*, Dr Gideon Hirschfield reviews the role of genetic determinants in both rare genetic syndromes and the more common autoimmune and drug-induced presentations. Drs Hallibasic, Baghdasaryan, and Trauner then methodically discuss the role of nuclear receptors in the pathogenesis and disease progression in cholestasis, as well as their use as important therapeutic targets. These 2 articles set the stage for a detailed survey of cholestatic liver diseases, beginning with a state-of-the-art summary on drug-induced liver injury with an emphasis on cholestatic presentations by Drs Jonasson and Bjornsson. Dr Claudia Zein examines the complex and controversial management of patients with primary sclerosing cholangitis, while Drs Czul, Peyton, and I explore the most recent advances and novel therapies for primary biliary cirrhosis. The nuances and challenges regarding the specific diagnosis and treatment of patients with overlap syndromes are lucidly discussed by Dr Marlyn Mayo. One of the most active areas of clinical investigation has been the differential diagnosis of sclerosing cholangitis. IgG4-associated cholangitis is reviewed by Dr Marina Silveira, and other secondary causes, including the newly recognized entity of sclerosing cholangitis in critically ill patients, are presented by Drs Iman, Talwalkar, and Lindor. Drs Hartley, Gissen, and Kelly highlight developments in the Pediatric arena, especially with respect to identification of causative genes and the multiple clinical phenotypes potentially associated with the same gene. Drs deLemos and Friedman describe the multiple systemic causes of cholestasis and present a very useful diagnostic algorithm for clinical practice. Drs Bolier, Oude Elferink, and Beuers discuss the current understanding of the pathogenesis of pruritus, exploring the role of the recently described autotoxin and its product, lysophosphatidic acid. We then turn to the management of extrahepatic complications from longstanding cholestasis, addressed by Andrea Gossard, as most cholestatic liver diseases do not have a definitive therapy. Lastly, for those whose condition progresses to end-stage liver disease, liver transplantation remains the only
curative option. The closing article by Drs Carrion and Bhamidimarri describes the available data on liver transplantation for both common and rare cholestatic diseases.

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