
Preface

Primary Sclerosing Cholangitis: Current Understanding, Management, and Future Developments grew out of a need I perceived within the fields of hepatology and liver transplantation. Primary sclerosing cholangitis (PSC) is a rare disease, with an incidence ranging from 0.04 to 1.30 per 1000,000 person years. Patients with PSC have variable presentations and there is significant variability in progression and prognosis. As a clinician, it has been frustrating that up until recently there has been little we could offer patients with PSC, other than liver transplantation, and little that we knew about its pathogenesis. In the liver community, much effort has been made into finding a cure for hepatitis C, a much more common chronic liver disease. With the development of successful antiviral therapies for hepatitis C, there has been a renewed interest into potential treatments of cholestatic diseases.

Indeed, it is an exciting time for PSC. Great work has been done to further clarify the role of genetics, immunology, and the microbiome with regard to the development and progression of PSC. Although liver transplantation remains the definitive treatment for advanced PSC, there are multiple new agents that are in clinical trials which will hopefully halt and even improve the fibrosis and subsequent complications associated with PSC. Endoscopic techniques have vastly improved over the past decade, and cholangiocarcinoma, a once universally fatal disease, can now be cured with liver transplantation.

In this book, recognized international experts in cholestatic diseases review the epidemiology, pathophysiology, current and future management of PSC, its variants, and associated complications. Up-to-date data regarding genetics, cholangiocyte biology, and immunology of PSC are presented. I hope that this publication will be of interest and utility to the medical and scientific community at large, with the ultimate goal of improving our understanding and treatment of this orphan disease.

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