
PREFACE

It has been recognized for centuries that cystic pathology of the liver and kidneys, and occasionally other organs, can occur in the same individual or within families and may have grave consequences. We have learned over time that the inheritance pattern seen in many of these conditions honors Mendelian genetics and that some conditions have non-cystic syndromic associations. Our ability to categorize these conditions has historically relied upon descriptive features observed postmortem, surgically, or through radiological techniques. Over the last decades, however, our understanding of the embryology, cellular pathogenesis, and genetic basis of these conditions has permitted clearer clinical diagnoses and the ability to prognosticate and offer therapies.

This is the first text to focus entirely on the fibrocystic diseases that affect the liver and the state-of-the-art research that underlies our current understanding of these conditions. We have brought together experts in the related fields of hepatic fibrocystic disease. The book provides a clear, in depth, and well-illustrated understanding of the embryology and development of the ductal plate, cholangiocyte biology, and the role of the biliary cilia in the pathogenesis of these conditions. An update review of the complex genetics of these disorders is reviewed in detail to allow further understanding of the molecular pathogenesis of the conditions as well as the clinical phenotypes encountered. Additionally, the text reviews the radiological and pathological methods important in diagnosis, the many clinical manifestations of these conditions and associated features and syndromes, the potential complications encountered in caring for affected individuals, and the treatments available to ameliorate the symptoms, progression, or complications of these diseases.

The text has incorporated a wealth of figures to illustrate the concepts described and to serve as reference for pathological and radiological findings of these conditions.

This book's purpose is to serve as a reference for those caring for patients with fibrocystic diseases affecting the liver and to offer an authoritative analysis of the cause of these conditions, their clinical manifestations, and the available strategies for managing for them. Additionally, it is the hope of the editors that this text will provide a

unique conglomeration of the knowledge to date of these conditions and hence serve as the nidus for further research advancements in the understanding and treatment of these conditions.

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