# Primary Immunodeficiency Diseases

The primary immunodeficiency diseases are a group of disorders in which the primary defect appears to be intrinsic to one or more components of the immune system.

Hans D. Ochs, MD, Dr.med,C. I. Edvard Smith, PhD, Jennifer M. Puck, MD

Primary Immunodeficiency Diseases Hans D. Ochs, C. I. Edvard Smith, Jennifer Puck, 2007 The second edition of Primary Immunodeficiency Diseases presents discussions of gene identification mutation detection and clinical and research applications for over 100 genetic immune disorders disorders featuring an increased susceptibility to infections and in certain conditions an icreased rate of malignancies and autoimmune disorders Since the publication of the first edition a flurry of new disease entities has been defined and new treatment regimens have been introduced the most spectacular being successful treatment by gene therapy for two genotypes of combined immunodeficiency. The first edition marked a historic turning point in the field of immunodeficiencies demonstrating that many of the disorders of the immune systam could be understood at a molecular level This new edition can proudly document the tremendous pace of progress in dissecting the complex immunologic networks responsible for protecting individuals from these disorders **Primary** Immunodeficiency Diseases Nima Rezaei, Asghar Aghamohammadi, Luigi D. Notarangelo, 2008-08-06 Primary immunodeficiency diseases PID are a group of disorders involving defects in one or more components of the immune system and are characterized by an increased incidence of infections autoimmunity and malignancies Although PID seem to be rare the number of patients diagnosed has increased in recent years and more than 150 different forms of PID have been identified Nevertheless because of inadequate medical awareness a significant number of patients with PID are either not recognized as having a PID or are not diagnosed as early as they should be Such delays lead to a substantial increase in morbidity and mortality among affected individuals Our understanding of PID is improving rapidly which will hopefully lead to more accurate diagnosis and efficient disease management This book contains the most recent advances in the field as well as a concise and structured review of previously identified PID Although the book s primary focus is on practical diagnosis and management the pathophysiology of PID is also discussed This book is a comprehensive yet manageable resource for physicians and nurses wishing to learn more about PID as well as a useful tool for both doctors in training and MD, Dr.med, C. I. Edvard Smith, PhD, Jennifer M. Puck, MD, 2013-11-01 Primary immunodeficiency diseases first recognized 60 years ago are inherited disorders that affect human adaptive and innate immunity In most cases affected individuals experience recurrent infections but they may also suffer from autoimmune diseases and malignancies This third edition of Primary Immunodeficiency Diseases provides readers with the historic and scientific background clinical presentations immunologic characteristics and the molecular genetic underpinnings of this rapidly enlarging class of diseases With up to date diagnostic tools and therapeutic options from prophylactic anti infective measures to hematopoietic stem cell transplantation and gene therapy this volume will remain an authoritative resource on this increasingly important area Clinical Cases in Primary Immunodeficiency Diseases Asghar Aghamohammadi, Nima Rezaei, 2012-12-13 Primary

immunodeficiency diseases PIDs are a heterogeneous group of inherited disorders characterized by different defects in the development and function of the immune system This book aims to increase the clinical awareness and knowledge of practicing clinicians regarding the diagnosis and management of PIDs In order to achieve this goal about 90 cases drawn from real life are presented along with approximately 300 related questions. The selected case reports are the result of the invaluable cooperation of more than 40 scientists in the field of immunodeficiency. They focus both on the presenting features of patients with PIDs and on the required further investigation and management Each of the numbered cases is followed by the questions their answers and additional discussion Each question focuses on a particular aspect of the PID under consideration and the topics covered include clinical diagnosis laboratory findings molecular mechanisms and therapy

Pulmonary Manifestations of Primary Immunodeficiency Diseases Seyed Alireza Mahdaviani, Nima Rezaei, 2018-12-31 This book provides a broad overview of the respiratory manifestations associated with Primary Immune deficiencies PID congenital genetic defects such as infections chronic inflammation autoimmunity lymphoproliferation allergic manifestations and rare forms of cancer Since the most common site of involvement in PID is the lung the pulmonologists pediatrics or adult Internists and General Practitioners may be among the first to recognize the pattern of pulmonary disorders leading to diagnosis of PID Pulmonary complications present a significant cause of morbidity and also mortality among patients suffering from different forms of PID With the prevalence of lung infections and disease so high in PID patients respiratory professionals will find this book to be an essential resource for diagnosing managing and referring Screening for Primary Immunodeficiency Disorders (PIDDs) in PID related pulmonary disorders in clinical practice Neonates Elham Hossny, Antonio Condino-Neto, Lennart Hammarström, Jolan Eszter Walter, 2021-02-04 Topic Editor Prof Lennart Hammarstr m holds equity in ImmunoIVD All other Topic Editors declare no competing interests with regards to the Primary Immunodeficiency Disorders Amos Etzioni, Hans D. Ochs, 2014-09-13 Primary Research Topic subject Immunodeficiency Disorders A Historic and Scientific Perspective provides a complete historical context that is crucial for students and researchers concerned with primary immunodeficiency When researchers have a poor understanding of the way we arrived where we are in research they can miss important points about a disease or miss out on how to approach new diseases This historical knowledge of research can assist greatly by showing how it was done in the past demonstrating the successes and failures so that it can be done better in the future This book provides an understanding of the process going from clinical problem to lab and back to the clinic based on historical experiences Its chapters proceed from the discovery of the T and B cell lineages through the first BMT for immunodeficiency disorder lab investigation and gene therapy for PID the discovery of the gene for AT and its function understanding cytokine defects and many other stops along the way Facilitates communication among physicians and other investigators concerned with immunological and inflammatory diseases Summarizes for the first time all the known facts from 60 years of primary immunodeficiency research and teaches how an

important field in medicine was established Provides stimulating discussions on developing new medical therapiesHighlights the importance of studying humans to understand mechanisms of disease that affect humans Primary Immunodeficiency Disorders, An Issue of Immunology and Allergy Clinics of North America 35-4 Anthony Montanaro, 2016-01-07 This issue of Immunology and Allergy Clinics of North America Guest Edited by Anthony Montanaro MD is devoted to Primary Immunodeficiency Disorders Dr Montanaro has assembled a group of expert authors to review the following topics Overview of Immunodeficiency Disorder Severe Combined Immunodeficiency Disorder Specific Antibody Deficiencies Common Variable Immunodeficiency Pulmonary Manifestations of Primary Immunodeficiency Disorders Approach to the Child with Recurrent Infections Immunoglobulin Treatment for Primary Immunodeficiency Bone Marrow Transplantation for Primary Immunodeficiency and Autoimmune Manifestations of Primary Immunodeficiency Primary Immunodeficiency Disorders Lisa Kobrynski, 2018-11-22 This issue of Immunology and Allergy Clinics of North America Guest Edited by Dr Lisa Kobrynski is devoted to Primary Immune Deficiencies Articles in this issue include Personalized therapy Immunoglobulin replacement for antibody deficiency Newborn Screening for Severe Combined Immunodeficiency Update on newborn screening and lessons learned Update on Advances in Hematopoietic Cell Transplantation for Primary Immunodeficiency Disorders Hereditary Autoinflammatory Disorders Recognition and treatment of inflammatory disorders of the immune system Use of Immunomodulatory Agents to Treat Primary Immune Deficiencies Mechanism based therapy Secondary Hypogammaglobulinemia An increasingly recognized complication of treatment with immunomodulators and post solid organ transplantation Use of Vaccines in Primary Immunodeficiency Gastrointestinal Manifestations and Complications of Primary Immunodeficiency Disorders Hyper IgE Syndromes Early Onset Inflammatory Bowel Disease and Genome Testing to Diagnose Primary Immunodeficiency Disorders and to Identify Targeted Therapy Brunner & Suddarth's Textbook of Canadian Medical-surgical Nursing Pauline Paul, Beverly Williams, 2009 This is the Second Edition of the popular Canadian adaptation of Brunner and Suddarth's Textbook of Medical Surgical Nursing by Day Paul and Williams Woven throughout the content is new and updated material that reflects key practice differences in Canada ranging from the healthcare system to cultural considerations epidemiology pharmacology Web resources and more Compatibility BlackBerry R OS 4 1 or Higher iPhone iPod Touch 2 0 or Higher Palm OS 3 5 or higher Palm Pre Classic Symbian S60 3rd edition Nokia Windows Mobile TM Pocket PC all versions Windows Mobile Smartphone Windows 98SE 2000 ME XP Vista Tablet PC

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