1. Record Nr. UNINA9910369948603321 Periodic and non-periodic fevers / / edited by Rolando Cimaz Titolo Cham:,: Springer International Publishing:,: Imprint: Springer,, Pubbl/distr/stampa 2020 **ISBN** 3-030-19055-2 Edizione [1st ed. 2020.] 1 online resource (287 pages): illustrations (some color) Descrizione fisica Rare Diseases of the Immune System, , 2282-6505 Collana Disciplina 616.047 Soggetti Fever Periodic diseases Lingua di pubblicazione Inglese **Formato** Materiale a stampa Livello bibliografico Monografia Nota di bibliografia Includes bibliographical references and index. Preface -- I From Pathogenesis to Classification -- 1 Historical notes Nota di contenuto on Autoinflammatory disorders -- 2 Pathogenesis: Genetic basis, Innate Immunity, Inflammosomes, Cytokines -- 3 Classification and nomenclature of Periodic and Non Periodic fevers -- II Specific diseases -- 4 Mevalonate Kinase Deficiency (MKD) -- 5 Tumor necrosis factor receptors associated syndrome (TRAPS) -- 6 Cryopyrin-Associated Periodic Syndromes (CAPS) -- 7 Pyogenic Arthritis, Pyoderma

gangrenosum and Acne Syndrome (PAPA) -- 8 Adenosine deaminase syndrome 2 (ADA2) -- 9 Interferonopathies -- 10 Inflammatory bone disease -- 11 Amyloidosis -- 12 Dermatologic autoinflammatory diseases -- 13 Autoinflammatory diseases with immunodeficiency --14 Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis Syndrome (PFAPA) -- 15 Proteasome. related disorders -- 16 Chronic Nonbacterial osteomyelitis -- 17 Systemic JIA and adult onset Still disease -- 18 Behcet disease -- 19 Recurrent idiopathic pericarditis -- 20 Schnitzler syndrome -- Subject Index.

Sommario/riassunto

This book, written by very well-known opinion leaders in the field. covers all aspects of periodic and non -periodic fevers, and related disorders. The expression refers to several different auto-inflammatory diseases, showing similar symptoms—the primary symptom being a recurrent fever for an infectious cause cannot be found. The opening chapters give some historical hints, explain the genetic basis of the disease and provide insights into the pathogenesis derived from recent

experimental studies and guides the reader through classification and nomenclature. A large part of the book is then devoted to a detailed description of the specific related diseases and their clinical presentations, the disease course, and potential complications in both pediatric and adult patients. The advice regarding treatment is based on the best currently available evidence in this constantly evolving area. The book is part of Springer's series Rare Diseases of the Immune System, which presents recently acquired knowledge on pathogenesis, diagnosis, and therapy with the aim of promoting a more holistic approach to these conditions. Autoinflammatory diseases are hereditary disorders that are caused by single-gene defects in innate immune regulatory pathways and are characterized by a clinical and biological inflammatory syndrome in which there is limited, if any, evidence of autoimmunity. Periodic and Non-Periodic Fevers will be an invaluable source of up-to-date information for all practitioners involved in the care of patients with these disease.