



Familial Mediterranean Fever (Rare Diseases of the Immune System)

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This book, written by very well known opinion leaders in the field, covers all aspects of familial Mediterranean fever, the most common monogenic autoinflammatory disease. The opening chapters explain the genetic basis of the disease and provide insights into the pathogenesis derived from recent experimental studies. A large part of the book is then devoted to a detailed description of the typical and atypical clinical presentations, the disease course, and potential complications in both pediatric and adult patients. Guidance is provided on the measurement of disease severity and the management of patients in daily practice. The advice regarding treatment is based on the best currently available evidence and attention is also paid to important emerging treatments.

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. Monogenic 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. Familial Mediterranean fever itself is due to a mutation in the *MEFV* gene, which codes for the protein pyrin; it is characterized by periodic fever and episodes of painful inflammation in the abdomen, chest, and joints. *Familial Mediterranean Fever* will be an invaluable source of up-to-date information for all practitioners involved in the care of patients with the disease.

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