Introduction

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This book includes the voices of clinicians, basic scientists and patients to better illustrate the most recent advances in research and care of sarcoidosis, a many-faceted disease with complex pathogenesis and various clinical manifestations https://bit.ly/32cyVUo

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Sarcoidosis is a systemic, multi-pathway disease and a clinical chameleon. Recent reports indicate that the prevalence of sarcoidosis is rising and the mortality in patients suffering from chronic sarcoidosis is increasing [1]. The high variability of manifestations, from asymptomatic to life-threatening, depends on organ involvement and disease activity, which still lacks a clear definition. About half of patients experience a chronic course. The consequences on quality of life and wellbeing can be devastating, especially if patients are not promptly referred to sarcoidosis specialists. To date, there are few approved treatments for sarcoidosis, mostly based on expert opinion, and the majority of investigational drugs have failed in clinical development. Although enormous advances have been made in the understanding of the disease pathogenesis and in the development of new diagnostic tools and management strategies, the burden of sarcoidosis, both socially and economically, is still considerable and the unmet needs persist.

It has been 17 years since the first *ERS Monograph* on Sarcoidosis, edited by Marjolein Drent and Ulrich Costabel, was published [2]. This *Monograph* is therefore timely and our main aim is to provide the reader with a comprehensive overview of the most recent advances in sarcoidosis.

We begin the book by illustrating the newest data on epidemiology [3] and cover the evidence for pathobiology of granuloma formation, including aetiological agents [4], the link between genotypes and phenotypes, and the genetic mutations occurring in familiar forms [5].

We then move on to discuss phenotyping, specific organ manifestations [6–9] and general diagnostic pathways [10], from conventional radiographic features to the use of novel modalities (*e.g.* PET scan) for diagnosis and assessment of disease activity [11]. We will also provide an update on the usefulness of circulating and imaging biomarkers for assessing disease severity and treatment response [12].

Traditional as well as innovative treatment strategies will be discussed, providing insight into unique aspects of therapy that differ between various organs. In two chapters focusing on when

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and how to treat sarcoidosis, the principles of treatment and stepwise algorithms will be critically appraised [13, 14]. The pipeline drugs on the horizon and ongoing clinical trials will be presented in a dedicated chapter [14].

Another novel and major element is represented by highlighting the patient's perspective. Besides a chapter exploring non-organ-related symptoms like fatigue and cognitive impairment [15], which all belong to the clinical picture of sarcoidosis, a chapter covering the effects of the disease on quality of life and an overview of tools used to assess quality of life is presented separately [16].

We have tried to include the voices of clinicians, basic scientists and patients to provide a better insight into this many-faceted insidious disorder. We believe that readers around the world will find this book helpful.

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