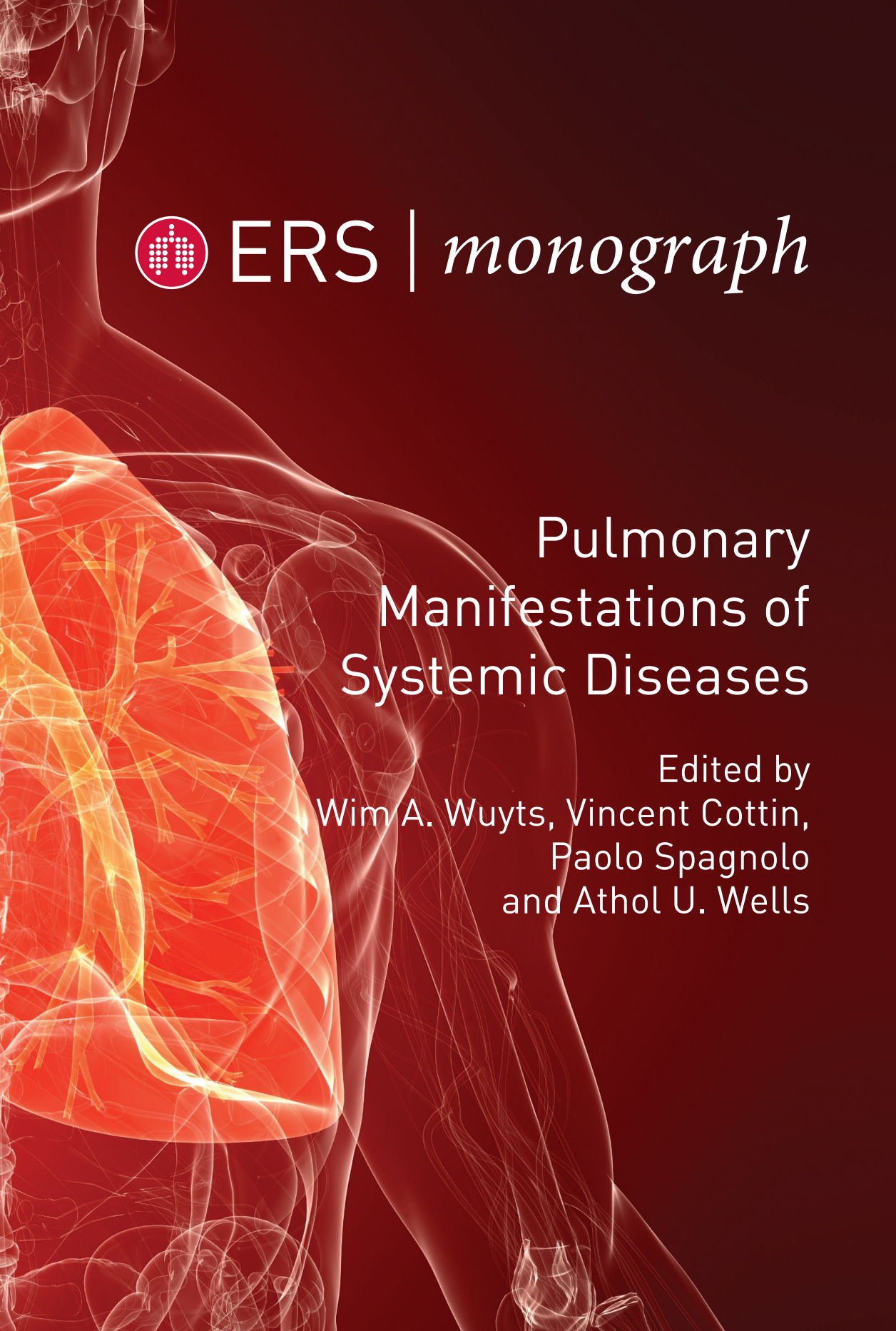
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Pulmonary Manifestations of Systemic Diseases
Edited by
Wim A. Wuyts, Vincent Cottin,
Paolo Spagnolo
and Athol U. Wells

Pulmonary Manifestations of Systemic Diseases

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Editor in Chief
John R. Hurst

This book is one in a series of *ERS Monographs*. Each individual issue provides a comprehensive overview of one specific clinical area of respiratory health, communicating information about the most advanced techniques and systems required for its investigation. It provides factual and useful scientific detail, drawing on specific case studies and looking into the diagnosis and management of individual patients. Previously published titles in this series are listed at the back of this *Monograph*.

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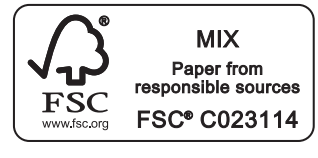

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Preface

John R. Hurst

It is a pleasure to introduce this edition of the *ERS Monograph*, focusing on pulmonary manifestations of systemic diseases.

As clinicians, we strive to care for patients by carefully evaluating the available evidence, with reference to guidelines, and by relying on our own past experiences. However, when managing people affected by rare lung diseases, this process breaks down in three main ways. First, many of us will simply not have had significant personal experience of such conditions. Secondly, guidelines may not exist and, thirdly, the available evidence may be much more limited in scope and quality.

This situation is true for many of the conditions discussed in the current edition of the *Monograph*, and I therefore have no doubt that the content will be incredibly helpful in managing some of the most difficult conditions in pulmonology. Indeed, I believe this *Monograph* will be important and of interest to specialists in other fields too, who are often the primary clinician for such patients. As respiratory clinicians and scientists, we are used to working in multi-professional teams. There is perhaps nowhere that this is more necessary than in managing the pulmonary manifestations of systemic diseases.

The society is indebted to the Guest Editors for doing an excellent job in curating the individual chapters. I would also like to pay tribute to the chapter authors and peer reviewers who together have delivered a relentless focus on quality.

Welcome to a new go-to reference work on Pulmonary Manifestations of Systemic Diseases!

Disclosures: John R. Hurst reports receiving grants, personal fees and non-financial support from pharmaceutical companies that make medicines to treat respiratory disease. This includes reimbursement for educational activities and advisory work, and support to attend meetings.

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Guest Editors

Wim A. Wuyts

Wim A. Wuyts is Associated Professor at Katholieke Universiteit Leuven (Leuven, Belgium) and is Head of the Unit for Interstitial Lung Diseases in the Department of Respiratory Medicine at the University Hospitals Leuven (Leuven, Belgium) and the Laboratory of Respiratory Medicine KU Leuven.

He obtained his medical degree and completed his postgraduate training in respiratory medicine at KU Leuven. He further specialised in the Department of Interstitial Lung Diseases at The Royal Brompton Hospital (London, UK), where he worked with the leading experts Professor Ron du Bois and Professor Athol U. Wells. He also further specialised in PAH in the department of Professor Marion Delcroix at the University Hospitals Leuven.

From 2000 to 2004, Wim Wuyts was an investigator at the Research Foundation – Flanders (FWO; Brussels, Belgium). In 2004, he earned a PhD in medical sciences at the KU Leuven with a thesis entitled "Human airway smooth muscle cells, role in chronic inflammatory disorders of the lung". He graduated in hospital management at the KU Leuven in 2004. He became a Senior Clinical Investigator at the FWO in 2011 and still holds this role.

Wim Wuyts is the driving force behind the ILD programme at the Laboratory of Respiratory Medicine of the KU Leuven. His research interests include immunology and fibrosis in ILDs, clinical research in ILDs and clinical trials in pulmonary fibrosis.

His work has appeared in peer-reviewed journals concerning ILDs, lung transplantation, PH and asthma. He is a member of various national and international task forces and scientific boards. In October 2014, he was elected as a member of the Executive Committee of WASOG (World Association of Sarcoidosis and Other Granulomatous Diseases) and he is member of the medical council of the University Hospitals Leuven.

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Vincent Cottin

Vincent Cottin is Professor of Respiratory Medicine and Coordinator of the National Reference Centre for Rare Pulmonary Diseases at the Louis Pradel Hospital (Bron, France) and the Claude Bernard University Lyon (Lyon, France). The Centre has pioneered clinical care and research in patients with rare and so-called orphan lung diseases for many years, and set up the OrphaLung network (coordinated by Vincent Cottin) of 21 expert centres throughout France. The Centre was recently recognised as the institute of its kind in France to be part of the European Reference Network for ILD (ERN-LUNG, ILD).

Vincent Cottin's research interests include rare "orphan" pulmonary diseases including IIPs and especially IPF, as well as CTD-associated ILD. More specifically, he has contributed to characterising and individualising the syndrome of combined pulmonary fibrosis and emphysema, and of IPAF. He is an investigator and member of the steering committees and data safety monitoring boards of many clinical trials on IPF, and is conducting personal research on combined pulmonary fibrosis and emphysema.

Vincent Cottin served as elected Head of the ERS Clinical Assembly from 2009 to 2012. He was Chief Editor of the *European Respiratory Review* from 2013 to 2015, and is currently Section Editor of the *European Respiratory Journal* and Associate Editor of *European Respiratory Review* and of *Respiration*. He recently edited a book on orphan lung diseases and is preparing a second edition. He is an appointed Fellow of the European Respiratory Society and has been awarded the European Respiratory Society Gold Medal of IPF.

Paolo Spagnolo

Paolo Spagnolo is Associate Professor of Respiratory Medicine and Director of the Residency Program in Respiratory Medicine at the University of Padua (Padua, Italy).

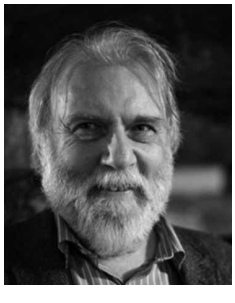
He received his undergraduate training and his MD and completed his residency in respiratory medicine at the University Hospital of Bari (Bari, Italy). In 2002, he joined the Interstitial Lung Disease Unit of the Royal Brompton Hospital (London, UK), initially as clinical research fellow under the supervision of Professor Ron du Bois, and subsequently as Honorary Consultant. In 2008, he completed his PhD at Imperial College London (London, UK) under the supervision of Professor Ron du Bois and Professor Ken Welsh, with a thesis on "Genetic predisposition to clinical phenotypes of

sarcoidosis". Between 2008 and 2013, he was research fellow and subsequently Assistant Professor in Respiratory Medicine at the University Hospital of Modena (Modena, Italy), where he joined the Centre for Rare Lung Diseases lead by Professor Luca Richeldi. Since 2015, Paolo Spagnolo has been Associate Professor of Respiratory Medicine at the Department of Cardiac, Thoracic, Vascular Sciences and Public Health of the University Hospital of Padua.

Paolo Spagnolo's main research interests include ILD and sarcoidosis with emphasis on genetic predisposition, prediction of disease behaviour and clinical trials of novel therapies. Paolo Spagnolo is the author or co-author of over 100 journal articles, review articles and editorials.

Paolo Spagnolo is a member of the editorial boards of *BioMed Research International* and *BMJ Open Respiratory Medicine*, and is a Section Editor (ILD) for *Current Opinion in Pulmonary Medicine*. He is the Treasurer of the WASOG (World Association of Sarcoidosis and Other Granulomatous diseases) and Secretary of the Study Group on Sarcoidosis of the European Respiratory Society.

Athol U. Wells



Athol U. Wells graduated at Otago University (Dunedin, New Zealand) in 1979, trained in New Zealand, and eventually moved to the UK permanently in 1999 and regrets not having done this 10 years earlier. He was given professorial status in 2005 and has focused on clinical research in ILD for the last 20 years (including diagnosis, prognostic evaluation and functional–morphological relationships). He has recently been honoured by a European Respiratory Society life-time award but does not see this as an indication that he should retire in the near future! He is very active in guideline groups and has nearly 500 peer-reviewed articles and editorials/review articles.

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Introduction

Wim A. Wuyts [1], Vincent Cottin [2], Paolo Spagnolo[3] and Athol U. Wells[4]

@ERSpublications
Pulmonary manifestations of systemic diseases must be considered forensically and not managed by focus on a single organ. This book provides expert clinical guidance on difficult diseases, aiding respiratory and nonrespiratory physicians. http://bit.ly/36HBY4i

ILDs comprise ⩾200 separate lung disorders. The interest in these diseases has risen enormously in recent years. A large body of basic and clinical research has greatly increased our understanding of the pathogenesis of IPF and, to a lesser extent, non-IPF fibrotic ILDs. Validated and emerging antifibrotic treatments for IPF are likely to be efficacious in other pulmonary fibrotic disorders exhibiting IPF-like disease progression. The ILD field is now evolving rapidly, with major implications for practical management.

However, it is important that the multiplicity of less prevalent ILDs, including those with predominantly inflammatory and mixed inflammatory/fibrotic phenotypes, are not lumped indiscriminately with inexorably progressive pulmonary fibrosis. Individual diseases must be appraised forensically. In this regard, pulmonary manifestations of systemic diseases cannot be adequately managed by focusing solely on ILDs but pose unique problems for physicians. Many systemic diseases are treated by non-respiratory specialists who do not always have expertise in assimilating pulmonary signs, symptoms and tests. Furthermore, many of these entities are extremely uncommon and this is often challenging for respiratory physicians. This *Monograph*, dedicated to the pulmonary manifestations of systemic disorders, provides background information and expert clinical guidance on these difficult diseases, which will be helpful to respiratory physicians and non-respiratory specialists alike.

The initial chapter considers the diagnostic issues in these diseases [1]. As a first step, collaboration between different experts in medicine is a requisite in these diseases; this is a challenge, but for our patients the best is not enough. In order to establish the most confident diagnosis, collaboration is necessary as data originate from different experts in the field. The next step is to find a way to communicate these results and integrate them into the specific case for the individual patient. The best way is multidisciplinary discussion. This has been extensively tested over the years and has now become the

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standard approach for evaluation of complex ILDs. We would argue that this manner of discussion should be transferred to other domains and that more non-pulmonary specialists should be added to teams. This exchange of expertise works both ways: on the one hand, it is beneficial as it raises awareness about the lungs amongst specialists in other fields, such as rheumatologists, general internists, nephrologists… On the other hand, we as pulmonologist could also learn from them in increasing our suspicion of the symptoms and signs of underlying CTD, but also by being more specific in the evaluation of only subtle biochemical abnormalities, without clinical counterpart.

Another important topic covered in this *Monograph* is PFTs, the cornerstone of diagnosis and follow-up [2]. At first sight it might seem odd that this is covered in a book mainly intended for pulmonologists. But we are convinced that in modern literature, there are few manuscripts that bring together current knowledge and particularly the pitfalls in PFTs. We also hope that this book will be picked up by experts outside the field of pulmonary medicine so that they realise the value of PFTs, but also look for the necessary expertise within their team.

The book goes on to discuss the contemporary techniques that are now in full development, such as quantitative CT analysis in ILD and the use of artificial intelligence on imaging of ILD [3]. This chapter provides a comprehensive overview of the possibilities now and in the future.

In this *Monograph*, we have tried to cover a variety of interesting pulmonary manifestations of very different disease entities, such as CTDs, systemic vasculitis and others.

The major CTDs covered are RA [4], inflammatory myopathies [5], SSc [6], Sjögren syndrome and mixed CTD [7], antiphospholipid syndrome [8] and finally, IPAF and undifferentiated CTD [9].

The chapters on systemic vasculitides comprise: microscopic polyangiitis and granulomatosis with polyangiitis [10], DAH [11], eosinophilic granulomatosis with polyangiitis [12], and pulmonary involvement in Takayasu arteritis and Behçet disease [13].

The chapters covering other diseases with extrathoracic involvement discuss bronchial and pulmonary involvement in inflammatory bowel diseases [14], the lung in liver disease [15], lung complications of neuromuscular diseases [16], amyloidosis and the lung [17], trafficking and lysosomal storage disorders (Hermansky–Pudlak, Gaucher, Niemann–Pick, Fabry, Lysinuric protein intolerance) [18], pulmonary involvement in haematological disorders and bone marrow transplant recipients [19], systemic histiocytic disorders (Langerhans and non-Langerhans cell histiocytosis, Erdheim–Chester and Rosai–Dorfman) [20], immunodeficiency [21], telomere syndrome and the lung [22], and finally, the pulmonary manifestations and management of sarcoidosis [23].

The Guest Editors very much hope that this *Monograph* will provide the reader with an interesting overview and update on the possible pulmonary involvement and specific treatment options of different systemic diseases. We sincerely hope that this book will be well received and will raise further interest in these diseases, leading to an increase in collaboration between different organ specialists and earlier diagnosis of patients confronted with very complex diseases that might turn their live upside down. We also hope that this book will serve as a catalyst to the development of more basic and clinical research in these intriguing diseases.

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List of abbreviations

BAL	bronchoalveolar lavage
CT	computed tomography
CTD	connective tissue disease
DAD	diffuse alveolar damage
DAH	diffuse alveolar haemorrhage
D_{LCO}	diffusing capacity of the lung for carbon monoxide
FEV_1	forced expiratory volume in 1 s
FVC	forced vital capacity
HLA	human leukocyte antigen
HRCT	high-resolution computed tomography
Ig	immunoglobulin
IIP	idiopathic interstitial pneumonia
IL	interleukin
ILD	interstitial lung disease
IPAF	interstitial pneumonia with autoimmune features
IPF	idiopathic pulmonary fibrosis
IPS	idiopathic pneumonia syndrome
LIP	lymphocytic interstitial pneumonia
MALT	mucosa-associated lymphoid tissue
NSIP	nonspecific interstitial pneumonia
OP	organising pneumonia
PAH	pulmonary arterial hypertension
PFT	pulmonary function test
PH	pulmonary hypertension
RA	rheumatoid arthritis
SLE	systemic lupus erythematosus
SSc	systemic sclerosis
UIP	usual interstitial pneumonia