ERS | monograph

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

[1]Respiratory Medicine, Unit for Interstitial Lung diseases, University Hospitals Leuven, Leuven, Belgium. [2]Service de Pneumologie, Hopital L. Pradel, Lyon, France. [3]University of Padova School of Medicine and Surgery, Padova, Italy. [4]Interstitial Lung Disease, Royal Brompton Hospital, London, UK.

Correspondence: Wim A. Wuyts. Unit for Interstitial Lung Diseases, Dept of Respiratory Medicine, University Hospitals Leuven, Herestraat 49, 3000 Leuven, Belgium. E-mail: wim.wuyts@uzleuven.be

 Print ISBN: 978-1-84984-111-5. Online ISBN: 978-1-84984-112-2. Print ISSN: 2312-508X. Online ISSN: 2312-5098.

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.

 https://doi.org/10.1183/2312508X.10033219

References

1. Luppi F, Faverio P, Wuyts WA. Multidisciplinary approach to systemic diseases: benefits for diagnosis and management of complex disorders. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 1–13.
2. Valenzuela C, Wells AU. Pulmonary function tests in multisystem disorders: prejudices and pitfalls. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 14–26.
3. Calandriello L, Matin T, Prosch H, *et al.* Quantitative CT analysis in ILD and the use of artificial intelligence on imaging of ILD. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 27–43.
4. Mackintosh JA, Stainer A, De Sadeleer LJ, *et al.* Rheumatoid arthritis. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 44–67.
5. Cottin V, Barba T, Mainbourg S, *et al.* Inflammatory myopathies. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 68–89.
6. Wells AU. Systemic sclerosis. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 90–105.
7. Antoniou KM, Vasarmidi E, Trachalaki A, *et al.* Systemic lupus erythematosus, Sjögren syndrome and mixed connective tissue disease. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 106–123.
8. Barratt SL, Pauling JD, Chaudhuri N. Antiphospholipid syndrome. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 124–139.
9. DeDent AM, Fischer A. Interstitial pneumonia with autoimmune features. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 140–152.
10. Pagnoux C. Microscopic polyangiitis and granulomatosis with polyangiitis. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 153–172.
11. Bonifazi M, Stanel S, Margaritopoulos GA. Diffuse alveolar haemorrhage. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 173–187.
12. Cottin V, Ahmad K, Nasser M, *et al.* Eosinophilic ganulomatosis with polyangiitis. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 188–209.
13. Alfieri V, Margaritopoulos GA. Takayasu arteritis and Behçet disease. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 210–227.
14. Camus P, Colby TV. Airway and lung involvement in inflammatory bowel disease. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 228–261.
15. Torrisi SE, Fuhrmann V, Skowasch D, *et al.* Liver disease: hepatopulmonary syndrome and portopulmonary hypertension. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 262–277.
16. Konstantelou E, Pasparaki E, Tzilas V, *et al.* Neuromuscular diseases. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 278–295.
17. Rech J-S, Brillet P-Y, Jeny F, *et al.* Amyloidosis. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 296–318.
18. Spagnolo P, Miedema JR, von der Thüsen JH, *et al.* Trafficking and lysosomal storage disorders. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 319–332.
19. Poletti V, Colella S, Piciucchi S, *et al.* Haematological disorders and bone marrow transplant recipients. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 333–358.
20. Elia D, Caminati A, Cassandro R, *et al.* Histiocytic disorders. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 359–373.

21. Bendstrup E, Vasakova M. Immunodeficiency. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 374–390.
22. Borie R, Kannengiesser C, Crestani B. Telomere syndrome. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 391–403.
23. Yserbyt J, Wells AU. Sarcoidosis: pulmonary manifestations and management. *In*: Wuyts WA, Cottin V, Spagnolo P, *et al.* Pulmonary Manifestations of Systemic Diseases (ERS Monograph). Sheffield, European Respiratory Society, 2019; pp. 404–418.

Disclosures: W.A. Wuyts reports receiving the following outside the submitted work: grants from Roche and Boehringer Ingelheim paid to his institution. P. Spagnolo reports receiving the following outside the submitted work: personal fees and non-financial support from Roche Boehringer Ingelheim, Zambon, Galapagos, Chiesi and Red-X Pharma; and grants, personal fees and non-financial support from PPM Services. P. Spagnolo's wife is employed by Novartis. A.U. Wells reports receiving the following, outside the submitted work: consultancy and speaking fees from Roche and Boehringer Ingelheim; and consultancy fees from Bayer and Blade.

 https://doi.org/10.1183/2312508X.10033219