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Bronchiectasis

Edited by R.A. Floto,
C.S. Haworth.



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Bronchiectasis

Edited by
R.A. Floto, C.S. Haworth

Editor in Chief
T. Welte

This book is one in a series of *European Respiratory 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 needed to investigate it. 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.

Contents

Number 52

June 2011

Guest Editors	v
Preface	vi
Introduction	vii
1. Bronchiectasis: epidemiology and causes <i>D. Bilton and A.L. Jones</i>	1
2. Pulmonary defence mechanisms and inflammatory pathways in bronchiectasis <i>B.N. Lambrecht, K. Neyt and C.H. GeurtsvanKessel</i>	11
3. Histopathology of bronchiectasis <i>M. Goddard</i>	22
4. Assessment and investigation of adults with bronchiectasis <i>M. Drain and J.S. Elborn</i>	32
5. Radiological features of bronchiectasis <i>P.L. Perara and N.J. Screatton</i>	44
6. Microbiology of non-CF bronchioectasis <i>J.E. Foweraker and D. Wat</i>	68
7. Allergic bronchopulmonary aspergillosis and other fungal diseases <i>B. Hilvering, J. Spiers, C.K. van der Ent and J.M. Beekman</i>	97
8. Nontuberculous mycobacterial infections <i>C.L. Daley</i>	115
9. Ciliary dyskinesias: primary ciliary dyskinesia in adults <i>L.J. Lobo, M.A. Zariwala and P.G. Noone</i>	130
10. Channelopathies in bronchiectasis <i>I. Sermet-Gaudelus, A. Edelman and I. Fajac</i>	150
11. Bronchiectasis associated with inflammatory bowel disease <i>Ph. Camus and T.V. Colby</i>	163
12. Immunodeficiencies associated with bronchiectasis <i>J.S. Brown, H. Baxendale and R.A. Floto</i>	178

13. Bronchiectasis and autoimmune disease <i>D.J. Dhasmana and R. Wilson</i>	192
14. Antibiotic treatment strategies in adults with bronchiectasis <i>C.S. Haworth</i>	211
15. Anti-inflammatory therapies in bronchiectasis <i>D.J. Smith, A.B. Chang and S.C. Bell</i>	223
16. Pharmacological airway clearance strategies in bronchiectasis <i>P.T. Bye, E.M.T. Lau and M.R. Elkins</i>	239
17. Surgery for bronchiectasis <i>D.C. Mauchley and J.D. Mitchell</i>	248
18. Conclusions and future developments <i>R.A. Floto</i>	258

Guest Editors



R.A. Floto

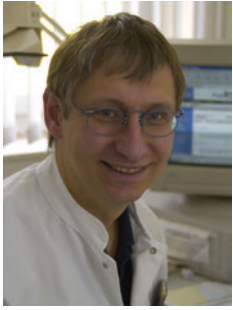
R.A. Floto is a Principle Investigator and Wellcome Trust Senior Clinical Fellow at the Cambridge Institute for Medical Research, University of Cambridge (Cambridge, UK). His laboratory, funded by the Wellcome Trust and Medical Research Council (UK), is focussed on understanding how the immune system interacts with bacterial and mycobacterial pathogens to trigger inflammatory lung damage. He is head of research at the Cambridge Centre for Lung Infection directing clinical and translational studies on CF and non-CF bronchiectasis and is an Honorary Consultant at Papworth Hospital and Addenbrooke's Hospital (both Cambridge). Recent honours received include the BUPA Foundation Researcher of the Year award (2010) and the European Respiratory Society Maurizio Vignola Award for Innovation in Pulmonology (2007).



C.S. Haworth

C.S. Haworth is Director of the Cambridge Centre for Lung Infection (incorporating The Adult Cystic Fibrosis Centre, The Lung Defence Clinic and The Immunology Clinic) at Papworth Hospital (Cambridge, UK). He is also an Honorary Consultant at Addenbrooke's Hospital in Cambridge. The Lung Defence Clinic oversees the care of more than 1,000 patients with bronchiectasis associated with primary and secondary immunodeficiency syndromes, nontuberculous mycobacterial (NTM) disease, Aspergillus-related lung disease, rheumatoid arthritis, serious childhood infection, chronic aspiration and primary ciliary dyskinesia. C.S. Haworth trained at the Royal Brompton Hospital and the Hammersmith Hospital in London (UK), before moving to Cambridge in 2003. He is a co-author of the North American Cystic Fibrosis Foundation/the UK Cystic Fibrosis Trust/European Cystic Fibrosis Society Bone Health Guidelines and is co-chair (with R.A. Floto) of the European Cystic Fibrosis Society NTM working group. He collaborates with several research groups at the University of Cambridge and is the chief investigator of multicentre, novel therapy, clinical trials in cystic fibrosis (CF) and non-CF bronchiectasis.

Preface



Bronchiectasis has been a well-known disease for a long time. Following the introduction of antibiotic treatment in clinical practice for respiratory tract infections, the problem of bronchiectasis appeared to be solved, with some exceptions, *e.g.* in diseases such as cystic fibrosis. However, bronchiectasis is associated with a number of immunological diseases and occurs as a long-term complication of chronic lung diseases. These types of diseases, mainly chronic obstructive pulmonary disease, have become more and more prevalent, which has again made bronchiectasis a disease of interest. Unfortunately, most of the evidence regarding bronchiectasis is from case series and uncontrolled studies. Bronchiectasis has not been a focus of the pharmaceutical industry and randomised controlled studies have never been performed. Specific guidelines focusing on bronchiectasis are yet to be published.

Over the past few years the scene has changed dramatically. Bronchiectasis is now a hot topic for epidemiological, basic and clinical research. A number of drugs, such as inhaled antibiotics and substances improving sputum clearance, are now available in a clinical development programme, the first results of which will be presented later this year. Therefore, now is the time to summarise the current knowledge about bronchiectasis.

The Guest Editors of this Monograph have succeeded in attracting leading experts within the field to write chapters which provide an overview from current pathophysiology, diagnostics and treatment to future developments that are on the horizon.

I want to congratulate the Guest Editors for this excellent Monograph, which will be of interest and use to basic scientists and clinicians in their daily practice.

Editor in Chief
T. Welte

Introduction

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Since its first description in the 19th century, bronchiectasis remains a clinically important, but poorly understood condition. This issue of the *European Respiratory Monograph (ERM)* brings together contributions from leading international experts on the subject of non-cystic fibrosis (CF)-associated bronchiectasis in adults. This issue of the *ERM* discusses the epidemiology and aetiology of the condition and describes the associated changes in histopathology and radiology. It explores the basic mechanisms controlling lung inflammation and immunity and how these can be disrupted to trigger bronchiectasis. In this Monograph, we define appropriate investigation algorithms, explore the role of bacteria, viruses, fungi and nontuberculous mycobacteria, and discuss the specific features of bronchiectasis associated with ciliary dyskinesias, channelopathies, inflammatory bowel disease, immunodeficiencies and autoimmune disease. This Monograph details the various treatment modalities available for bronchiectasis, including antibiotic regimens, the use of macrolides and other anti-inflammatory agents, airway clearance strategies and the role of surgery.

This issue of the *ERM* offers a comprehensive and cutting edge review of non-CF-associated bronchiectasis and provides a definitive guide to the management of this challenging condition.