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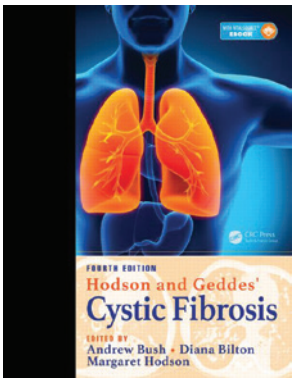
Hodson and Geddes' Cystic Fibrosis

Editors: Andrew Bush, Diana Bilton and Margaret Hodson; CRC Press; 699 pages; ISBN: 978-1444180008

Book review

Cite as: Horsley A. Book review: Hodson and Geddes' Cystic Fibrosis. *Breathe* 2016; 12: 91–92.



With the fourth edition of this now eponymous tome, the new editors have once again brought together a wide range of over 80 experts in different aspects of cystic fibrosis (CF) care. The remit of this book is both wide ranging and deep, stretching to 49 chapters over 700 pages. The history of CF care and the people who have shaped it are described in their own chapter, but are, in fact, also laid bare in the evolving editions of this book. To get a feel for the incredible advances in basic understanding of and treatments available for CF, you only have to compare the length and contents of the current (fourth) edition with that of the second edition, published only 15 years earlier (had a copy of the first edition had been available for comparison, it's doubtless that the differences would have appeared even starker). In no other field of respiratory medicine has there been such a revolution in clinical practice and outcomes. This disease of childhood, with a median life expectancy of only 24 years at the start of the century, has been transformed into a chronic disease of adulthood. Patient numbers have swelled accordingly, but with improvements in life expectancy have also come challenges created by managing a host of additional CF and treatment-related complications.

The latest edition of Hodson and Geddes' Cystic Fibrosis very much reflects this changing landscape. Much of the basics of CF medicine are perhaps covered elsewhere, including in national guidelines or other more concise texts. But this book is not one for the practitioner with a casual interest in CF healthcare. Only in a book of this magnitude can the more detailed but demanding topics be addressed. Chapters on growing old with CF, sleep, noninvasive ventilation and palliative care very much reflect the changing burden of disease, and are important topics that have so far received relatively little attention. A marker of how much things have changed is the lack of patient segregation in the earlier edition. This is a core principle of CF care today. Understanding the microbiology of CF lung disease, detecting infection early and preventing patient–patient transmission are all covered separately in three excellent and detailed chapters.

What has not changed in the intervening years is that CF remains one of the most diverse,

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challenging and rewarding areas of respiratory medicine. CF care is also an exemplar of the multidisciplinary team approach. It is refreshing to see, alongside the grandees of CF medicine, important contributions from physiotherapists, dietitians, nurses, psychologists, diabetologists, otolaryngologists, gastroenterologists, palliative care specialists and a host of others. Most notable among these are the patients and parents who have written a chapter on their own experiences of the disease. Even veteran and hardened professionals will find it hard not to melt at these first-hand accounts, and they help to put all the research and endeavour into perspective.

As our experience of the disease has evolved, CF has become very much at the forefront of clinical and basic research into the mechanisms of lung disease, work that is now filtering into other areas of respiratory medicine. This is not neglected here either, with chapters dealing specifically with trial outcomes, computed tomography and multiple-breath washout techniques. The most important and transformative recent advances in CF, however, are the development of cystic fibrosis transmembrane conductance regulator modulator and chaperone therapies, barely dreamt of 15 years ago. This is now a rapidly evolving field, with new compounds coming to trial all the time. A large volume like this, which takes time to compile, is perhaps not the best place to source up-to-date developments in this field. Indeed some chapters make reference to "future" studies that have already been published (gene therapy for example). This will always be a weakness of traditional text books, but in compensation the basic science is exhaustively covered.

Overall, this book is as comprehensive and definitive a guide to the state of clinical experience and practice in CF as it is possible to come by. It is a sweeping epic that takes in both the past and the future, and provides a reference for even the most experienced CF specialist to learn from or to reach for in times of trouble.

Conflict of interest

A. Horsley reports grants from National Institute of Health Research during the conduct of the study; grants from Vertex Pharmaceuticals and personal fees from Oxford University Press, outside the submitted work. A. Horsley is an editor and author of the ORML handbook on CF.