

gency department, post-extubation respiratory failure, weaning from mechanical ventilation, and perioperative use. The strong evidence base for NPPV in COPD exacerbation, acute cardiogenic pulmonary edema, respiratory failure in immunocompromised patients, and weaning from mechanical ventilation in patients with COPD is included and current. The rationale and evidence for NPPV in other less strongly supported situations, including asthma, pneumonia, acute respiratory distress syndrome, and post-extubation respiratory failure is discussed in detail. There has recently been an increasing amount of literature supporting the use of NPPV in postoperative respiratory failure, which I thought should have been addressed either in the chapter on hypoxemic respiratory failure or in a chapter by itself.

There is a very good chapter about setting up NPPV, which includes information about indications, patient selection, relative and absolute contraindications, and instructions for mask fitting, ventilation modes, initial settings, patient monitoring, and duration of NPPV. Cough-assist techniques and physiotherapy are detailed in a very informative chapter. The physiology of cough and the numerous techniques for cough assistance and secretion clearance are explained thoroughly. There is also a chapter on problem-solving in NPPV. Problems include persistent hypercapnia, hypoxemia, air leaks, ventilator asynchrony, confusion, and claustrophobia. Detailed action plans are described for these common situations. Those involved in starting patients on NPPV will find this chapter useful, since these problems are often challenging.

The book has several chapters on patient selection and outcomes of long-term NPPV. These sections cover the common causes of chronic respiratory failure that requires home NPPV, including neuromuscular diseases, chest wall disorders, cystic fibrosis, bronchiectasis, interstitial lung disease, and COPD. These chapters are outstanding. I particularly enjoyed reading Simonds's chapters on neuromuscular diseases and chest wall disorders. They detail the evidence base for NPPV in that patient population, the pathophysiology of the conditions, and the benefits of NPPV. Practitioners caring for these patients will find these chapters very helpful.

There are 2 chapters on discharging patients on home NPPV and problem-solving in long-term NPPV use. These chapters detail the numerous potential advantages of

NPPV over ventilation via tracheostomy. The practical issues of managing complications of long-term NPPV use and equipment maintenance are covered.

There are a few chapters on NPPV use in children. The acute indications for NPPV are discussed, although the evidence base is limited. Home ventilation in children is also covered, including the common disorders that lead to chronic respiratory failure and the evidence base for NPPV.

There is an excellent chapter on sleep-disordered breathing. This is a thorough review of obstructive sleep apnea, including pathophysiology, symptoms, diagnosis, and treatment. Continuous positive airway pressure therapy is discussed in detail, including mask selection and setting the optimal pressure. The evidence base for continuous positive airway pressure in obstructive sleep apnea is reviewed in detail, as are other therapies for obstructive sleep apnea, including weight loss, surgery, and dental appliances.

The last several chapters discuss home mechanical ventilation in Europe, and legal and ethical issues. These chapters are addressed more toward a European audience. The ethical principles of beneficence, avoiding maleficence, respecting individual autonomy, and distributive justice are always important to review, especially with regard to this patient population.

In summary, this is a very thorough, nicely written book. The reader will gain a better understanding of the expanding evidence base for NPPV in both the acute setting and the long-term setting in patients with chronic respiratory failure. The practical NPPV issues, including patient selection and monitoring, selecting an interface, ventilation mode preference, pressure settings, complications, and problem-solving, are covered in detail.

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The author of this review reports no conflict of interest.

**Alpha-1 Antitrypsin Deficiency—Clinical Aspects and Management.** Thomas Köhnlein and Tobias Welte. Bremen, Germany: Uni-Med Verlag. 2007. Hard cover, illustrated, 103 pages, €44.80.

Alpha-1 antitrypsin (AAT) deficiency is a common but under-recognized condition that predisposes to chronic obstructive pulmonary disease. It is estimated that fewer than 10% of expected individuals with AAT deficiency are clinically recognized, and there is evidence of a long diagnostic delay between initial symptoms and first recognition by health care providers. Because respiratory therapists (RTs) can play an important role in recognizing AAT deficiency and facilitating diagnosis, the book **Alpha-1 Antitrypsin Deficiency—Clinical Aspects and Management** is a welcome addition to the field and as a resource for RTs.

The book is concise (103 pages) and is a compilation of chapters written by German authors, including some very well recognized authorities in the field. The chapters include an introduction (35 pages, which presents a very nice overview of AAT deficiency, oxidant-antioxidant balance, and protease-antiprotease hypothesis), a discussion of clinical aspects (15 pages), diagnostics (11 pages), and treatment (31 pages), including usual therapy of chronic obstructive pulmonary disease, augmentation therapy, surgical approaches (eg, lung-volume-reduction surgery and transplantation), and gene therapy. The appendix (3 pages) discusses various registries and patient resources, including the Alpha-1 Foundation and the Alpha-1 Association. The book's foreword is written by an august scholar of AAT, Dr Eriksson, who, with Dr Laurel, first described AAT deficiency in 1963. The foreword is a brief description of Dr Eriksson's work in the Malmo (Sweden) cohort, which offers important insight into the natural history of AAT deficiency, and is content-rich.

In the context that the book is a concise treatise on the field, its coverage of AAT and AAT deficiency states is admirably thorough. Indeed, this book might well be used by a respiratory clinician or investigator interested in getting an overview of the field as a prelude to gaining a richer clinical understanding or entering the field as an investigator. The chapters are brief (generally 2–5 pages) and readable in short, stand-alone sessions. Citations, which are included as "Further Reading" or "References," are topical and are a resource to the reader who wishes to delve further, though the text itself certainly provides a solid foundation in basic aspects of AAT and its deficiency, as well as some of the pathobiology of resultant emphysema and liver disease.

The book is illustrated with color images, which are clear, visually appealing with striking but soothing colors, and helpful (though small, in the context of a small-format book). The histologic micrographs and radiologic images (chest radiographs and computed tomograms) are well rendered.

From the perspective of the RT reader, the book is again concise and thorough but contains both more information than might be needed for the RT clinician and some information that might be beyond reach and relevance (eg, the discussions of the regulation of serpin synthesis and some of the molecular biology). In this regard, the book might well find a place in the library of RT teaching or training programs as a reference rather than a regularly read resource, especially since a reader seeking the most up-to-date information will consult the medical literature directly, given the inevitable delays between manuscript finalization and publication of a book.

Recognizing the many strengths of this book and its great usefulness as a contribution to the field and to the libraries that respiratory clinicians use, several shortcomings warrant mention. First, as expected, and as is unavoidable for a book that summarizes a field in which new information continues to emerge at a relatively rapid pace, some late-breaking information about AAT deficiency is not included. This is perhaps more evident in the sections on therapy than in those on clinical presentation and diagnosis. For instance, the discussions on lung-volume-reduction surgery and gene therapy do not include the latest, emerging information or references (eg, current gene therapy trials underway at the University of Florida).

Second, the critical reader may quibble about some small details in this book, such as the statement on page 18 that initial pulmonary symptoms include "massive sputum production," which is not the case in my experience and is not part of the classical description of AAT deficiency, or the statement on page 17 that "for clinical symptoms to manifest, the alleles inherited from both parents must exhibit a defect." Though in that statement the authors are probably discussing the pulmonary manifestations of AAT deficiency (about which the statement is largely correct), the statement overlooks the fact that patients who are PI\**MZ* heterozygotes (who have only a single abnormal allele) may develop cirrhosis on the basis of AAT deficiency. Indeed, that fact is

recognized elsewhere in the book. Other quibbles could be made about editing errors, which are few but present. For example, on page 40, "the emphysematous changes in the lungs have been contributed [should be "attributed"] to the lack of anti-proteolytic capacity."

Certainly, these and other small quibbles, and the fact that the book does not cite the latest-breaking information, should not obscure the overall impression that this book is a very useful addition to the literature and will be a valued resource for the respiratory clinician. Its conciseness is appealing to all those who have too much information to absorb and therefore desire a single source of organized information. Finally, the book will be a useful reference addition to libraries that regard themselves as having complete respiratory collections.

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The author of this review reports no conflict of interest.

**Cystic Fibrosis**, 3rd edition. Margaret Hodson, Duncan Geddes, and Andrew Bush, editors. New York: Oxford University Press/Hodder Arnold. 2007. Hard cover illustrated, 503 pages, \$198.50

Cystic fibrosis (CF) is a common genetic disorder caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. The clinical phenotype is characterized by progressive damage to organs that express CFTR, with particular involvement of the lungs, pancreas, and hepatobiliary system. Median survival is into the mid-thirties, with over 90% of CF patients dying of respiratory failure. Life expectancy of CF patients has improved substantially because of aggressive treatment delivered by multidisciplinary CF teams in dedicated CF units.

This third edition of **Cystic Fibrosis** is a comprehensive textbook, with 34 chapters written by 66 experts, covering all aspects of CF. It has been extensively updated since the 2nd edition (published in 2000), with the addition of a third editor, which reflects the dramatic and rapid changes in the understanding and management of CF over

the past decade. The 3rd edition includes new chapters on sexual and reproductive health, palliative care, and lung transplantation, and the book is aimed at all providers of adult and pediatric CF care, including trainees, nursing staff, dietitians, physiotherapists, respiratory therapists, psychologists, and social workers.

The book has 7 sections. The first section gives an introduction to CF and a brief review of CF history and epidemiology. The chapter on the history of CF will be an enjoyable read for all CF care providers; it reviews all the major medical and research advances, broken down by decade, since Anderson first described CF in the 1930s. The detailed review of CF epidemiology includes up-to-date information on the incidence, clinical features, demographics, and survival/prognosis, as reported by international registries and epidemiological studies.

The second section has 4 chapters, each of which describes a different aspect of CF basic science, including genetics/biology of CFTR, CF pathophysiology and immunology, and the influence of modifier genes and the environment on the CF phenotype. The chapters are detailed and clear, and as they are aimed at CF clinicians, do not require extensive background knowledge of molecular biology. Citations for review articles are provided for further reading.

Section 3 deals with the diagnosis of CF. The first chapter reviews the various approaches to making the CF diagnosis and includes a section on the more detailed investigations that are often required to diagnose a patient with atypical CF. The second chapter deals with the benefits and challenges of screening newborns for CF and how to deal with CF patients identified via newborn screening. The final chapter in this section reviews the complicated issue of the role of the microbiology laboratory in the care of CF patients. I found this chapter very informative, albeit brief, and believe it will be of value to many CF clinicians. It has sections on sputum sample processing, optimal antimicrobial susceptibility testing (including synergy testing), and infection control.

The fourth and largest section of the book deals with the multitude of clinical complications and challenges of CF. This section has 12 chapters, each of which deals with a specific complication. The first 3 chapters address respiratory complications. As would be expected, considerable attention is focused on respiratory infections characteris-