

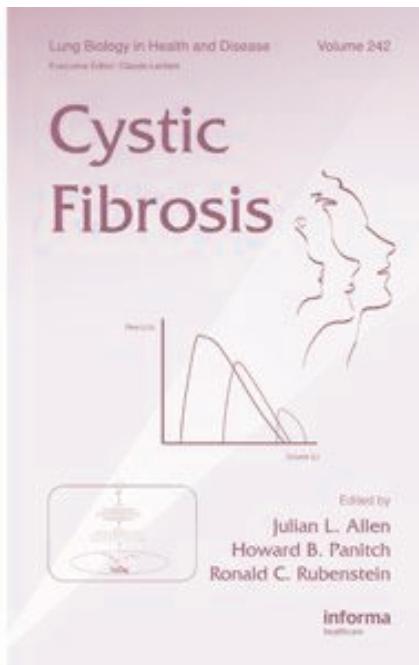
## BOOK REVIEW

### Cystic fibrosis. Lung Biology in Health Disease, Vol. 242.

Edited by J.L. Allen, H.B. Panitch and R.C. Rubenstein

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"The search for truth is in one way hard and in another very easy. For it is evident that no-one can master it fully nor miss it wholly. But each adds a little to our knowledge of nature, and from all the facts assembled there arises a certain grandeur"

Aristotle

"The big picture is all in the detail"  
Charles Darwin

The quote from Aristotle is as appropriate now as it was when it appeared in the preface to the first volume addressing cystic fibrosis (CF) in this very successful series. How far we have come, and how far we still have to go!

In 1993, we were still in the afterglow of unravelling the genetic abnormality of CF and how this related to pathophysiology and clinical syndromes. The future looked bright regarding understanding CF and having increased options to intervene and improve clinical outcomes. Over the ensuing 17 years this is exactly what happened but perhaps not in exactly the way predicted. CF outcomes have now improved dramatically, underpinned by developments in care and research at all points in the disease spectrum from early diagnosis/screening, through to nutritional and lung function maintenance, psychosocial health and advanced care, including the option of lung transplantation. If there was one standout discovery that characterises this period of improved CF care it is that enabling CF teams to focus on uncompromising excellence and vigilance in implementing, integrating and continuously incrementally improving all the "standard approaches in CF care" can have dramatic long-term benefits for CF patients.

All those involved with this book should be congratulated on achieving their aims of "giving caregivers access to an increased understanding of the molecular basis of CF, its pathophysiology and health care system approaches" and providing a platform of "detailed knowledge and insights that will

allow the generation of testable hypothesis for future basic, clinical and translational research". Despite the increased number of subsections and expert contributors (more than double that of the original volume), this tome has remained approximately the same size as its predecessor. It is divided into four distinct subsections which equally focus on pathophysiology, diagnostics, clinical manifestations/management and issues relating to psychosocial functioning, healthcare systems and quality improvement. As such it covers all important areas and is well balanced. In general, chapters follow a theme of historical background, moving on to current chapter specific information, and then variably entertaining future possibilities. Overall there is a strong descriptive focus with some repetition/overlap, which is somewhat unavoidable in a book such as this. At key junctions these "overlap" areas could have been a springboard for a deeper emergent discussion predominantly regarding genotype/phenotype and phenotype/outcome inter-relationships.

The first section on CF pathophysiology is accurate and up-to-date but perhaps leaves the reader still yearning for a 'bigger picture' unifying view. For example, the first three chapters focus on CF genetics, ion transport and mucus abnormalities/ciliary dysfunction; all reasonable overviews in their own right with a strong classification and descriptive focus but could have been significantly enhanced by better cross-chapter integration regarding concordant/discordant relationships and including more explanatory summary illustrations/diagrams, preferably in colour. This pattern is then repeated with individual strongly detailed chapters on CF airway microbiology (especially regarding laboratory detection and identification, specific organism epidemiology and antimicrobial sensitivity testing) and inflammation, emphasising the potential for "a dysregulated inflammatory response to eventually become more harmful than protective"; however, with both making the point that the links between the basic CF transmembrane conductance regulator (CFTR) defect and the propensity to airway infection and progressive lung disease remain to be fully elucidated. Appropriately, a chapter on modifier genes then follows and has the opportunity to tie this section in nicely but is somewhat lengthy.

The section on CF diagnostics begins with an excellent review of sweat testing and newborn screening (incorporating both US and European approaches) and appropriately focuses on the lifelong implications for individuals and families, the importance of concomitant genetic counselling, and cleverly uses borderline cases to emphasise critical points. This overlaps with the diagnostic approach in the following chapter on CFTR gene mutations where the full spectrum of CF- and CFTR-related disorders is evenly discussed, as are the limitations of diagnosing CF in adolescence and the central importance of an individuals clinical status in decision making. The subsequent three chapters have a strong technical flavour and focus on the very different challenges of accurately measuring lung functions at different ages from infants to young children to school aged children. Although the CF lung physiology chapters are well detailed, this section could have included a more integrated section focusing on the overarching principles of early detection and (dys)proportional change in relation to various physiological measures, clinical patterns and radiological imaging, particularly given the changing CF phenotypes at

different ages. The relatively poor correlation of chest imaging findings with the results of pulmonary function tests and clinical outcomes is again emphasised in the subsequent section on imaging. Chest imaging is presented here both with a research focus and as an outcome surrogate (the case for both being supported by the results from recent studies). Imaging of the gastrointestinal tract is also covered and overall this is a well balanced section that details both the increasing use/utility of imaging in CF and the increasingly recognised dangers of cumulative radiation exposure.

The chapters on clinical manifestations and treatment again provide excellent detail but variably introducing new information. The first four chapters in this section have a well-defined focus (pulmonary manifestations, treatment strategies for maintaining pulmonary health, mucolytic and airway clearance techniques, and pulmonary exacerbations) and are very informative and complementary, although some overlap is unavoidable. The chapter on pulmonary exacerbations in particular is concisely overviewed with an excellent discussion regarding definitions, epidemiology and evidence-based management. These pulmonary chapters are complemented by subsequent chapters on gastro-intestinal complications, liver disease, nutrition, bone health, diabetes and more general extrapulmonary complications. The information here is up-to-date, thorough and generally very well presented. The potentially difficult chapter on non-typical extrapulmonary complications is especially well structured, comprehensively covering a broad area with good use of available evidence and is very readable. The important section on chronic respiratory failure management and the roles of NIV and lung transplantation is factually stated and current but would have been well served by also including the importance of local management protocols and expertise, the role of individual risk/benefit assessments, values and choice in management decisions, and the many difficulties that we still have in predicting individual responses and outcomes. This is particularly critical given the decision making that “turns on” the analysis and interpretation of the information relating to

advanced care options in end-stage CF lung disease. The management spotlight then makes a huge leap to gene repair and CFTR function modulation, which are good basic primer chapters but remain complex areas where the detail could have been better illustrated. Although some novel information is presented and the future is full of hope, it is clear that there is still a long way to go regarding these possibilities.

The book appropriately finishes with a strong review on quality improvement, psychosocial considerations and care systems. The pivotal roles of CF foundations, patient registries, partnerships, team approaches and the chronic care illness model are well detailed. Further chapters on infection control, transition to adult care, biopsychosocial/adherence/quality of life models and palliative/end of life care in CF are all expertly discussed and flow well. Although some of these chapters are much better written than others, all have addressed many areas of change in CF care over the last two decades. This section also incorporates a succinctly written chapter on reproduction, sexuality and fertility, further reflecting the changing times and increasing life choices that people with CF now have.

Overall this is an excellent, up-to-date reference book addressing all aspects of CF that builds on the promise of the previous edition and has something for everyone from novice student to specialist clinician/researcher. The editors/contributors should be congratulated on fulfilling their primary objectives. Future tomes focusing on CF in this series could more explicitly increase the scope of objectives to include a higher level of integration between groups of highly inter-related chapters so that the “detail that doesn’t quite fit” can increase the bigger picture of CF. This would introduce an opportunity to frame a discussion for the more controversial issues that CF patients, clinicians and researchers have to deal with on a day-to-day basis.

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