who also have many years of "hands on" experience. My unit has managed over 1500 patients since 1978 in the manner described, also without any hepatic fatality or need for transplantation.³

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BOOK REVIEWS

The Thorax. Part A: Physiology; Part B: Applied Physiology; Part C: Disease (Lung Biology in Health and Disease Series, No 85). 2nd edition, revised and expanded. Charis Roussos. (Pp 2880; \$495.00 sold as set only.) New York: Marcel Dekker, 1995. ISBN 0-8247-9647-0.

The second edition of *The Thorax* is mainly concerned with the respiratory muscles and certainly has the feel of a tome, containing three parts and nearly 3000 pages. There are more than 100 contributing authors from North and South America, Europe, and Australia. Most have personal, practical experience in the field of respiratory muscle physiology and are authorities on the subject.

The book aims to provide the clinical investigator with comprehensive, up to date information of physiology and pathophysiology of respiratory muscles, and the diagnosis and treatment of respiratory muscle dysfunction. It is divided into three parts. Part 1 begins with chapters on physiology of skeletal muscle followed by information on structure, function, and metabolism of the respiratory muscles. This section provides the clinician or investigator with the opportunity to understand in depth the properties of the respiratory muscles. Part 2 illustrates clearly every method for measuring respiratory muscle function and explains function in some activities such as speech, sleep, and exercise. Some chapters are very good reviews – for example, chapter 49 on respiratory muscle fatigue by Roussos himself which includes more than 200 references up to 1994 and provides state of the art information on function and fatigue of the respiratory muscles. There is also a good chapter on respiratory muscle activity during sleep which does not include the effect of sleep disordered breathing on the respiratory muscles, but covers the outcome of a combination of sleep studies and respiratory muscle studies. Part 3 deals with the diagnosis of respiratory muscle dysfunction and pathophysiology in various clinical conditions as well as treatment. Most chapters are excellent, a noteworthy one

being that by De Troyer on respiratory muscles in COPD which deals in detail with static and dynamic function and structure of the respiratory muscles in those patients. There are also good chapters on mechanical ventilation which is useful for the clinician working in the intensive care unit.

On the whole, the standard of writing, the quality of discussion, and the level of explanation of this book are of a high order. Most of the chapters include up to date references, apart from chapter 37 on rib motion in health and disease where all the references come from before 1972. There are some trivial problems – for example, some chapters cite references using consecutive numbers and some use the authors' names and year of publication. Chronic obstructive pulmonary disease is abbreviated to COPD in one place and CAO in another. There is a fair amount of overlap and repetition between reviews – for example, measurement of maximal transdiaphragmatic pressure is illustrated in chapter 38, chapter 63, and chapter 64; mechanisms of dyspnoea are repeatedly described in chapter 43, chapter 60, and chapter 61. If some repetition had been avoided the book might have been thinner! There is some contradiction between chapters – for example, breathlessness and dyspnoea are used interchangeably in chapter 60, yet in chapter 61 they are clearly meant to represent different phenomena.

Although there are small defects, this book is still a noteworthy effort. It will provide invaluable information to the respiratory physician. At a price of \$495 it will probably be destined for the library and the respiratory muscle laboratory rather than chest physicians' shelves. – YML and SGS.

The Molecular Biology and Pathology of Elastic Tissue. DJ Chadwick and JA Goode Road. (Pp 361; £49.94). Chichester, UK: John Wiley & Sons, 1995. 0 471 95718 6.

I took my son to the Science Museum in Boston a few years ago. I think I was more impressed than he was. In one of the exhibitions they had two pumps (each representing a heart), one extruding its liquid into a solid plastic pipe and the other into an elastic tube. In one the flow was pulsatile, in the other continuous. This was a wonderfully dramatic demonstration of the importance of elastic vessels to ensure continuous perfusion of our body tissues.

The functional importance of elastic tissue has been known for many years but its composition has been poorly understood. Interest has not been helped by observations of its apparently amorphous structure (a feature which I learned from this book is unfounded and likely an artefact of conventional microscopy techniques) and its resistance to biochemists attempting to solubilise and study it. In the last 20 years or so, largely aided by techniques of cell and molecular biology, our view of elastic tissues has altered radically.

We now realise that elastic tissues such as the lung and blood vessels are characterised by a high content of elastic fibres. These fibres consist of elastin and the so-called 'elastin-associated microfibril'. The latter contains at least seven glycoproteins including fibrillins 1 and 2 and microfibril-associated glycoprotein, MAGP. Elastin gene and protein structure is now quite well understood. The protein is encoded from a single gene on chromosome seven. The

primary transcript, tropoelastin, undergoes several post-translational modifications including hydroxylation, oxidative deamination of lysines catalysed by lysyl oxidase, and stabilisation by covalent cross link formation between modified and normal lysine residues.

Abnormal deposition of elastin occurs in several respiratory disorders, including pulmonary hypertension and pulmonary fibrosis. In contrast, a diminished amount of elastin is a feature of pulmonary emphysema. There are also some rare heritable disorders in which there are abnormal elastic fibres (e.g. Marfan's syndrome and supravalvular aortic stenosis), reduced synthesis (Cutix laxa), or overproduction (pseudoxanthoma elasticum and Buschke-Ollendorff's syndromes).

This book is timely and as up to date as it can be in a field which is moving rapidly. I often judge a book like this in two ways. Has it presented what I already knew clearly and have I learned new things? By and large it succeeds on both counts. The chapters on protein and gene structure were of interest scientifically, placing the older work in historical perspective. The chapter on elastin gene structure was informative but spoiled by very poor quality diagrams for which the hatching was often indecipherable. This fault in this chapter and others was my single greatest criticism and the publishers of a book such as this owe it to their readers to get this aspect right.

There were many good chapters but some may be of particular interest to the lung research community. The chapter on elastin synthesis was of interest partly because it emphasised how little we know of the molecules regulating elastin production. The chapter on fibrillin stressed the importance of this protein in the mechanical properties of elastic tissues as manifested by the observation that Marfan's syndrome involves a defect in the fibrillin gene. The structural role of elastin has begun to be investigated using transgenic mice into which different tropoelastin gene constructs had been inserted. Of particular interest to the respiratory world was the suggestion that animals lacking exon 33 develop a lung morphology characteristic of emphysema. The parity between this model and human emphysema was argued in the discussion which followed this paper, and there was some debate as to how closely they resembled one another and whether or not the model represented a developmental defect rather than emphysema. Although this model has some limitations, it still strikes me that it may share some common mechanisms with emphysema. The publication of discussion is the tradition of this series – this should be continued as it is a great help to readers to get a feel for the current thinking of those working in the field.

Finally, on reading this book I realised I had already heard of the meeting from which it emanated. Bob Mecham visited us in London a year or so ago and told me he was on the way to Kenya for a Ciba Elastin Meeting. I remember thinking how smart these elastin people are. The last Ciba meeting I attended, although very pleasant, took place in wintry London one mile from where I work. Presumably, the environment in Kenya was a little more enticing.

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