

LETTERS TO THE EDITOR

The hyperimmunoglobulinaemia E and recurrent infections syndrome in an adult

The recent short report by Dr J-P L'Huillier and others (September 1990;45:707-8) gives a good review of the intriguing hyperimmunoglobulinaemia E syndrome that causes recurrent infections (also known as Job's syndrome). We recently presented two cases of this syndrome to the British Thoracic Society summer meeting¹ and we think that two further points are worth making in relation to the investigation and management of such patients. Firstly, the identification of the syndrome and in particular its distinction from severe atopic dermatitis is well worthwhile because these patients may improve clinically with H₂ receptor blocking drugs,¹ possibly as a result of an improvement in neutrophil chemotactic defects produced by histamine H₂ receptor blockade.² Secondly, some of these patients may show deficiencies of specific antibodies to capsulated bacteria such as *Haemophilus influenzae* and *Streptococcus pneumoniae*, and they may be helped by regular treatment with intravenous gammaglobulins.³

Because of the suboptimal immune response in these patients antibiotics by themselves may not be particularly effective, so H₂ blockade and intravenous gammaglobulins in selected cases are worth considering.

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- 1 Maxwell S, Kumararatne DS, Iles PB, Honeybourne D. Two cases of chronic pulmonary disease associated with the hyperimmunoglobulin E syndrome [abstract]. *Thorax* 1990;45:822.
- 2 Mawhinney H, Killen M, Fleming WA, Roy AD. The hyperimmunoglobulin E syndrome—a neutrophil chemotactic defect reversible by histamine H₂ receptor blockade? *Clin Immunol Immunopathol* 1980;17:483-91.
- 3 Thompson RA, Kumararatne DS. Hyper Ig E syndrome and H₂ receptor blockade. *Lancet* 1989;ii:630.

AUTHORS' REPLY We agree with the remarks of Drs Honeybourne and Maxwell. After observing an improvement in chemotaxis of neutrophils in patients with the hyperimmunoglobulinaemia E syndrome after exposure in vitro to burinamide (an H₂ receptor blocking drug) Hill *et al* proposed the use of H₂ receptor blockers,¹ and Trung *et al* suggested an H₁ receptor blocking drug with an H₂ receptor blocker and a mastocyte degranulation inhibitor²; but no controlled study has been published. Other authors have proposed antistaphylococcal antibiotic prophylaxis.³ The detailed pathophysiological mechanisms underlying this syndrome are not well known and optimum treatment has not been determined. Of the many treatments proposed (including human plasma, transfer factor levamisole, ascorbic acid, sodium cromoglycate, and plasmapheresis, etc), none is widely recognised as being effective. Our patient has been treated since October 1986 with ranitidine (300 mg/day), dexchlorpheniramine (12 mg/day), and ketotifen (2 mg/day) and, since summer 1988, amoxicillin-clavulanic acid and trimethoprim-sulfamethoxazole have been alternated for periods of

two weeks. The patient has noted increased sputum volume and fever when she stops taking antibiotics. Gammaglobulin was given in September 1986 and in June 1988, but was stopped soon afterwards because of lack of efficacy and the difficulty of giving an intramuscular injection.

We could not include these points in our paper because of shortage of space.

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- 1 Hill HR, Estensen RD, Hogan NA, Quie PG. Severe staphylococcal disease associated with allergic manifestations, hyperimmunoglobulinemia E and defective neutrophil chemotaxis. *J Lab Clin Med* 1976;88:796-806.
- 2 Pham Huu Trung, Oury C, Daumling S, Buriot D, Delohradsky BR, Griscelli C. Syndrome de susceptibilité aux infections avec hyper IgE. A propos de 19 nouvelles observations. *Arch Fr Pédiatr* 1982;39:353-8.
- 3 Schopfer K. Le syndrome de susceptibilité aux infections avec hyper IgE. In: Doin J, ed. *Déficits immunitaires congénitaux et acquis*. Paris, 1984:141-56. (Progrès en hématologie 5.)

BOOK NOTICES

Occupational Exposure to Silica and Cancer Risk. L Simonato, AC Fletcher, R Saracci, TL Thomas (Pp124; £19.) Oxford University Press, 1990. ISBN 92-832-1197-9.

This book comprises a collection of scientific papers describing the results of mortality studies of the possible relationships between occupational exposure to airborne silica and risk of lung and other cancers. It is the result of the work of a study group activated by the International agency for Research on Cancer to produce new epidemiological information on some of the less investigated aspects of the lung cancer-silica dust relationship. The papers comprise an introduction by Simonato and Saracci and 11 other papers, three of which describe previously unreported work, so far as I can determine; the other seven are updates of longitudinal studies or reworkings of published data. All are in English, and are written for readers familiar with epidemiological concepts and, in some cases, specialist terms. The critical reader will quickly spot the problems of potential bias in selection or recruitment of some study populations, and in case identification and diagnosis when the subjects are known to have silicosis or are receiving special medical surveillance; two studies, however, seem to me to avoid these problems. One, by PD Winter and colleagues reports new (or new to me) findings in pottery workers, and a relationship between lung cancer risk and cumulative exposure to silica that I find impressive. Additionally, Koskela and colleagues describe updated findings of an excess lung cancer risk in granite workers. The book marks an advance in the debate on silica and lung cancer, and will be a required purchase by libraries and specialists.—CS

The Lungs (Vol 5 of **Systemic Pathology**). 3rd ed. B Corrin. (Pp 467; £75.) London: Churchill Livingstone, 1990. 0-443-03094-4.

The series of pathology texts under the

general editorship of W St C Symmers has evolved from two volumes covering all pathology to a set of 15 monographs, each covering one system. This is the fifth in the new third series. In most chapters the authors discuss aetiology and pathogenesis in detail, usually with a good clinicopathological section. Limitations of space do not allow the authors to dwell on rare conditions covered in larger monographs but they have mentioned almost everything a pathologist is likely to see more than once in a practising lifetime. Part of this miracle of fitting a quart into a pint pot is achieved by the clarity and economy of the writing, which makes the book easy to read and understand. I particularly like the chapter on pulmonary fibrosis, which dismisses the complexities of usual interstitial fibrosis and the like in favour of a logical discussion of the pathological mechanisms and a descriptive histological diagnosis. The book is well illustrated with a mixture of line diagrams, radiographs, macroscopic and microscopic pathological preparations, and electron micrographs, with a slight bias in favour of electron micrographs that I did not like. The illustrations are generally of high standard but occasional light micrographs are difficult to interpret because of poor contrast and lack of definition. The previous two editions of Symmers have been written predominantly for histopathologists. This book will appeal to a wider market, particularly clinicians and radiologists who wish to have access to a good text on the general pathology of the lung. As such it would have been of value to have had a chapter describing the uses and limitations of various types of biopsy and, for trainee histopathologists, discussion of an approach to gross surgical specimens. In summary, the authors have sacrificed some of Symmers' pedigree as a first report bench book, producing a text that will appeal to a wider market; the result is the best of the middle length, moderately priced monographs on pulmonary pathology now available.—MW

Diagnosis of Diseases of the Chest. Vol 3. 3rd ed. RG Fraser, JAP Paré, PD Paré, RS Fraser, GP Genereux. (Pp 704; £60.) Philadelphia: Saunders, 1990. ISBN 0-7216-3872-4.

Most respiratory physicians recognise the primacy of the chest radiograph as a diagnostic aid. It was around this principle that Fraser and Paré developed their textbook. This is now in its third edition and runs to four volumes. Volume three deals with pulmonary vascular diseases, airways disorders, and the effects of inhaled foreign material. The chapter on embolic and thrombotic lung diseases is an example of the comprehensive approach of the authors. It is meticulously indexed at the beginning, as are all chapters in this volume. There follows a consideration of the incidence, aetiology and pathogenesis, pathology, radiographic features, and aspects of diagnosis. The methods of ascertaining pulmonary embolism are discussed at some length and pulmonary angiography and scintigraphy are compared for their usefulness. This is followed by clinical aspects and then less common examples of embolic diseases. This chapter is elegantly illustrated with many radiographs, computed tomograms, and scintigraphic images. There are also excellent examples of histology and whole organ pathology. The 783 references provide both an up to date literature review and a historical perspective. The chapter on diseases of the airways covers obstructive sleep apnoea, asthma, chronic obstructive airways

disease, McCleod's syndrome, bronchiectasis, and cystic fibrosis. There are also chapters on the pneumoconioses and on aspiration of foreign material. One can find areas of deficit—for example, the lack of discussion on the radiographic scoring methods used in cystic fibrosis, such as the Crispin Norman score, which are in widespread use. The text as a whole, however, is detailed and broad and is not just about the chest radiograph and imaging techniques in respiratory medicine. This book is a must for any respiratory unit.—DJS

Diagnostic Imaging of the Lung. (Vol 46 of *Lung Biology in Health and Disease*). (Pp 769; \$198.) New York: Dekker, 1990. ISBN 0-8247-8318-2

This volume is written by a group of mainly American radiologists who are eminent in their field. The book has the professed objectives of "reviewing the role of conventional radiographic techniques" and "providing a reasonable diagnostic approach for the utilisation of more advanced technologies." It is aimed at a wide medical audience ranging from pulmonary physicians and radiologists to general practitioners. It starts with a good overview of the modalities currently available in radiology for the investigation of the chest. It describes the benefits of the high kv chest radiograph, emphasises the growing importance of high resolution computed tomography, compares the relative merits of magnetic resonance imaging and computed tomography and has sections on digital radiography, nuclear medicine, ultrasound, and interventional radiology. The introduction to each subsequent chapter covers the same ground again in a more limited form. The topics included range from diseases of the trachea and other airways through vascular disease, malignancy, and pulmonary oedema to diffuse and focal lung conditions. It finishes with a chapter on interventional radiology and another on future goals in chest radiology. For a book concerned with imaging, there is rather too much description of the clinical aspects of the disease processes, particularly in the chapters on diffuse lung disease. The space would have been better spent on pursuing the radiology in greater depth. Much of the introduction to the section on intervention would seem too obvious to need inclusion. Generally, the radiographs are reproduced well and the points that they illustrate are clearly seen. There are in addition some line drawings, particularly in the section on pulmonary oedema, and these are helpful. Each chapter ends with a long list of references, the more recent ones being not later than 1988 but covering the subject of the chapter well. The book is unlikely to appeal to a radiologist who has experience in thoracic disease but would be useful to readers who are seeking an overview of the range of imaging modalities available and the ways in which they interrelate. The chapter on pulmonary vascular diseases is a good summary for physicians on the approach to pulmonary thromboembolic disease as this condition is often rather poorly investigated from a radiological perspective.—ARM

Modern Drug Treatment for Tuberculosis. 7th ed. (Pp 102; £5.) London: Chest, Heart, and Stroke Association, 1990. ISBN 901548-47-2

The seventh edition of *Modern Drug Treatment of Tuberculosis* has been updated from

the last edition in 1983 to cover new developments, particularly the impact of AIDS on tuberculosis. The book title is a little misleading for, although over half of the book covers the drugs used to treat tuberculosis (both first line and reserve drugs and drug regimens for developed and developing countries), other topics are included. The first chapter covers the pharmacology of the drugs, together with side effects and interactions. Treatment regimens, including daily, partly intermittent, and fully intermittent regimens, are discussed for both pulmonary and extrapulmonary disease. Costs and suitability for different economic and social situations are well covered, recommendations being tailored to the money and health facilities available, and are consistent with the IUAT guidelines. Short chapters on generalised drug reactions, special situations, chemoprophylaxis, the management of acquired drug resistance, opportunistic mycobacterial disease, tuberculin testing, BCG vaccination, and tuberculosis control programmes are also included. The emphasis in these latter sections is on practical advice based on the author's wide experience, covering the limitations and advantages of certain techniques—for example, tuberculin testing—and dispensing sound advice on the clinical topics. The price of £5 is very reasonable. The book will be suitable for three groups of doctors. Firstly, those who treat tuberculosis only occasionally will find firm and concise advice. Those who treat tuberculosis frequently will also find helpful advice on the less common problems. Finally, because the different needs and resources of developing countries are borne in mind throughout the book, doctors in developing countries will find much of practical help in setting up and running a tuberculosis service.—PO

Kendig's Disorders of the Respiratory Tract in Children. Eds V Chernick, EL Kendig Jr. (Pp 1055; £116.) Philadelphia: Harcourt Brace Jovanovich, 1990. ISBN 0-7216-2214-3

Although it is always a pleasure to place a bulky and expensive textbook on the office bookshelf, there must be doubt about the value of large tomes on any topic. Smaller monographs are often more up to date and better. This work is considered by many to be the bible of paediatric chest medicine. The fifth edition has been extensively revised and updated, only seven years after the last edition. None of the alternative large textbooks on this subject have managed to keep up to date; but smaller books have been published recently, the best by far being *Respiratory Illness in Children* from the Children's Hospital in Melbourne. *Kendig* is a multi-author textbook written predominantly by North American authors, with a few from other countries. The bibliographies contain very few non-North American references and some chapters are clearly unsuitable for European consumption. The chapter on asthma is particularly unsuitable: isoprenaline is still given more prominence than any beta₂ specific agonist, theophyllines have more space than sodium cromoglycate, and inhaled corticosteroids are demoted to two sentences buried in the section on oral steroids. The list of "useful" investigations for asthma starts with a full blood count followed by cytological examination of sputum and serum determination of immunoglobulins G, A, and M as well as E. Some chapters have been revised but really not updated, particularly

those on congenital defects. There is no classification and disorders are discussed in haphazard fashion, pulmonary sequestration in particular. The latest reference on this topic was 1983 (there are two references from the 1980s, six from the 1970s, 19 from the 1960s and 17 the 1950s). Other chapters are excellent, however, with very recent references. Outstanding among these are the chapters on sudden infant death syndrome and apnoea and pulmonary disorders in paediatric acquired immunodeficiency syndrome. A host of omissions might be picked out. There is no mention of the primary ciliary dyskinesia syndrome as a cause of transient tachypnoea or neonatal pneumonia, no mention of extracorporeal membrane oxygenation in the chapter on intensive care, and no mention of the peak expiratory flow meter in the section on lung function testing. The ciliary dyskinesia syndrome chapter does not comment on the measurement of ciliary beat frequency from nasal brushing or the saccharin clearance screening test. The chapter on antibiotics misses out ciprofloxacin and all monobactams. The chapter on foreign bodies does not mention the dangers of nut aspiration or the need for long term follow up, but suggests without references that steroids should be given for 48 hours postoperatively. It is fascinating that all paediatric respiratory textbooks say that taking a neck radiograph in a child with epiglottitis is dangerous, yet show a radiograph (twice in this book). The general format of the textbook follows that of many others with an initial section on anatomy, physiology, and clinical examination and investigations. The first two chapters are particularly good. The chapter on host defence concentrates almost exclusively on immunological mechanisms with little comment on non-specific defence mechanisms. The chapter on age and lung disease fails to discuss the changes that occur during male adolescence, which has a profound effect on form and function and manifestations of disease. Subsequent sections include neonatal disease and a very useful chapter on infections. Overall I am glad that I possess this textbook, which has some excellent sections and good bibliographies interspersed with poor chapters. It could be useful as a first port of call reference book when one is beginning to study a specific topic in paediatric respiratory medicine.—JOW

NOTICES

British Sleep Society

The British Sleep Society's third annual meeting will be held at Worcester College, Oxford, on 1-3 September 1991.

Free communications are invited. Information and abstract forms from: Dr J Stradling, Osler Chest Unit, Churchill Hospital, Headington OX3 7LJ (tel 0865 225236; fax 0865 225221).

Symposium on cardiopulmonary emergencies

The 7th International Symposium on Cardiopulmonary Urgencies and Emergencies will be held on 19-22 November 1991 in Rotterdam.

Details from Dr O Prakash, Thorax Centre, Erasmus University, 3000 DR Rotterdam, The Netherlands (fax 31-10-463 5240).