

## 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 meeting<sup>1</sup> 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 H<sub>2</sub> receptor blocking drugs,<sup>1</sup> possibly as a result of an improvement in neutrophil chemotactic defects produced by histamine H<sub>2</sub> receptor blockade.<sup>2</sup> 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.<sup>3</sup>

Because of the suboptimal immune response in these patients antibiotics by themselves may not be particularly effective, so H<sub>2</sub> 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 H<sub>2</sub> receptor blockade? *Clin Immunol Immunopathol* 1980;17:483-91.
- 3 Thompson RA, Kumararatne DS. Hyper Ig E syndrome and H<sub>2</sub> 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 H<sub>2</sub> receptor blocking drug) Hill *et al* proposed the use of H<sub>2</sub> receptor blockers,<sup>1</sup> and Trung *et al* suggested an H<sub>1</sub> receptor blocking drug with an H<sub>2</sub> receptor blocker and a mastocyte degranulation inhibitor<sup>2</sup>; but no controlled study has been published. Other authors have proposed antistaphylococcal antibiotic prophylaxis.<sup>3</sup> 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