Drs Ogilvy-Stuart and Shalet suggest that our results indicate a sexual dimorphism in thalassaemic children in terms of persistence of gonadotrophin deficiency secondary to iron deposition in the pituitary. The aim of our study was to assess whether conditioning treatment before bone marrow transplantation (BMT) with busulphan and cyclophosphamide may cause gonadal damage in thalassaemic patients, which would have a deleterious effect on the quality of life. While our data did not demonstrate obvious gonadal damage in prepubertal boys, it did so in girls. As a consequence, the discussion was focused on this last aspect, which was crucial to the aim of the study. However, we did recognise the possibility that the reduced gonadotrophin response after gonadotrophin releasing hormone in prepubertal thalassaemic males may be a consequence of iron overload which could conceal germinal epithelial damage. There is no doubt that long term follow up studies are necessary to demonstrate if the gonads of male thalassaemic patients are damaged by cytotoxic drugs.

This possibility is an additional reason for considering gonadal damage when discussing the pros and cons of BMT and this aspect was emphasised in our discussion. We agree that the patterns of gonadotrophin in response to gonadotrophin releasing hormone, found in prepubertal thalassaemic males, seems to suggest a higher sensitivity to iron overload. However, we feel that other studies must be made to explain this dimorphism in thalassaemic patients, because this hypothesis is not supported by the findings of a multicentre study on endocrine complications in 3200 thalassaemic patients followed up in 41 Italian hospitals. The result of our survey has shown an absence of puberty in 41% of the males and 39% of the females, over the age of 15 years (V De Sanctis *et al*, unpublished data).

We thank Drs Ogilvy-Stuart and Shalet for pointing out that the serum testosterone values reported in table 2 and in the text are not correct. We are very sorry for this mistake, which was caused by the change made from ng/ml to nmol/l. The values given should be reduced by a factor of 100.

Equipment requirements for community based paediatric oxygen treatment

SIR,—The bare list of ingredients required for domiciliary oxygen treatment is of little value without accompanying instructions on how they should be assembled.¹ Deficiencies in the current system of provision for community based oxygen treatment need to be tackled by national recommendations and the provision of appropriate devices for young children on prescription.

To this end, a Working Party on Domiciliary Oxygen Therapy for Children was convened under the auspices of the Committee for Thoracic Medicine of the Royal College of Physicians (London) in order to provide the Department of Health with the concerted recommendations of a number of organisations. It met in January and its recommendations have been submitted to the Department of Health.² Briefly the document recommends the types of equipment that are needed, the means whereby the equipment should be supervised, and the level of clinical support for families receiving domiciliary oxygen treatment. The responsibilities of health professionals within and without the hospital and

the role of the equipment industry were addressed.

It is hoped that these recommendations will provide the basis for the provision of domiciliary oxygen treatment for children to match the system which has evolved over a number of years for adults with chronic obstructive airway disease.

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- 1 Angell C. Equipment requirements for community based paediatric oxygen treatment. *Arch Dis Child* 1991;66:755.

*Members of the working party represented: Royal College of Physicians, Royal College of General Practitioners, Medical Gases Industry, Cystic Fibrosis Trust, British Paediatric Association, British Association of Perinatal Medicine, Royal College of Nursing, Department of Health, and British Thoracic Society.

Copies of the recommendations are available from Dr Silverman on receipt of an A5 stamped, addressed envelope.

Computerised information systems

SIR,—I read with interest Dr Spencer's recent article on neonatal information systems.¹ While admirably covering a large topic in a few pages, there were two points that I feel warrant greater emphasis.

'Local' databases that are set up by enthusiasts—and I have been involved in four such systems in different hospitals—often fall into relative or absolute disuse after a period of time. While this may be due to fundamental weaknesses in the systems, it is more commonly due to one of two factors.

Firstly, the enthusiast who initiated the project may well move to a different hospital. This leaves a void, often with direct control passing to someone who was not immediately involved in the development. Any problems that later arise (and they will!) may be difficult to fix, and further development of the system often ceases.

Secondly, and associated with the first point, is the importance of clear documentation. Dr Spencer mentioned this in his last sentence as 'an asset', but this understates its crucial role. Interest in the database is likely to wax and wane with time, as workload alters, research fellows come and go, or as the deadline for yearly reports comes round. Without comprehensive and clear documentation for a system, including details of trouble shooting and support available, the system will gradually deteriorate. Clear manuals must be an essential aspect of any computerised system.

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- 1 Spencer SA. Computerised neonatal information systems. *Arch Dis Child* 1991;66:823–6.

Septicaemia and adrenal haemorrhage in congenital asplenia

SIR,—The incidence of congenital absence of the spleen is said to be one in 2000, according

to the one postmortem series.¹ Dyke *et al* report five cases of asplenia including two otherwise normal infants.² We have recently seen a 1 year old infant who, in his first year of life, has had pneumococcal meningitis twice and osteomyelitis (culture negative) once. Several ultrasound scans and a technetium labelled sulphur colloid scan failed to reveal a spleen. Numerous Howell-Jolly bodies were present in the erythrocytes. He has no other apparent congenital abnormalities; immunoglobulin, white cell, and complement studies are normal. We concur with Dyke *et al* that congenital asplenia is an under recognised entity, and recommend the use of pneumococcal and *Haemophilus influenzae* vaccine at diagnosis, despite the lack of demonstration of efficacy in infants less than 24 months. The potential benefits of vaccination appear to outweigh possible adverse effects. Surely these infants need not wait until aged 18–24 months for potentially preventive immunisation?

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- 1 Shinohara Y, Komiya S, Nakashima A, *et al*. Asplenia and polypenia syndrome. *Acta Pathol Jpn* 1982;32:505.
- 2 Dyke MP, Martin RP, Berry PJ. Septicaemia and adrenal haemorrhage in congenital asplenia. *Arch Dis Child* 1991;66: 636–7.

SIR,—I read the paper by Dyke *et al* illustrating the role of *Haemophilus influenzae* infection and adrenal haemorrhage in causing overwhelming infection and death in patients with congenital asplenia with great interest.¹ I would like to bring to the attention of your readers a further case that recently presented to the Adelaide Children's Hospital.

Case report

A white girl aged 2 months was taken late at night to a country hospital with a three day history of breathing difficulties and cough. Apart from a non-prescription cough medicine no other medication had been given before admission. The initial examination showed that she had a temperature of 38°C and was tachypnoeic with laboured respirations. A provisional diagnosis of pneumonia was made and a single dose of intramuscular penicillin was given. After admission she appeared to settle initially but was found dead in bed four hours later. Her body was transferred to the Adelaide Children's Hospital for postmortem examination with a provisional diagnosis of death from pneumonia or possible sudden infant death syndrome.

At postmortem examination there was situs inversus of thoracic and abdominal organs with dextrocardia but no other significant cardiac abnormality. There was no spleen but two splenunculi (combined weight 3.4 g) were found in the right upper quadrant. Both adrenal glands were appreciably haemorrhagic. The lungs were atelactatic but there was no evidence of pneumonia. Bilateral otitis media was evident. Microbiological cultures including blood culture and culture of lung and middle ears were negative, but penicillin given before death may well have eliminated a sensitive organism.

This case further emphasises the points made by Dyke *et al* of the need to look for

evidence of anatomical or functional hyposplenism and of instituting measures such as antibiotic prophylaxis, vaccination against pneumococcal and *H influenzae* infection, and early treatment with antibiotics at the first sign of infection.

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1 Dyke MP, Martin RD, Berry PJ. Septicaemia and adrenal haemorrhage in congenital asplenia. *Arch Dis Child* 1991;66: 636-7.

Dr Langdon Down

SIR,—Peter Dunn's cameo is a helpful reminder of a distinguished physician.¹ However, Down published a good deal more than the article implies. In the 1887 Lettsomian lectures and a series of case studies Down described a diverse range of conditions. These included pseudohypertrophic muscular dystrophy ('by the aid of a harpoon I extracted specimens of the muscle from both gastrocnemii') of which he gives an accurate pathological description. He also describes patients with what is now called Asperger's syndrome, and children with Prader-Willi syndrome, fetal alcohol syndrome, hypothyroidism, dysgenesis of the corpus callosum and others.

There were some themes in which he took particular interest ranging from two articles on the mouth in people with mental retardation, the exaggerated criticism of traumatic childbirth as a cause of disability, the unsuitability of sitting schools for mentally handicapped children in hospitals for the mentally ill, and the relief experienced by parents when told that their children's disabilities predated the process of birth. He was also an advocate for full educational opportunities for women.

This material, first collected and published by the Medical Society of London in 1887, has been reprinted by MacKeith Press as the fifth in their excellent series of *Classics in Developmental Medicine*.²

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1 Dunn PM. Dr Langdon Down (1828-1896) and 'mongolism'. *Arch Dis Child* 1991;66:827-8.

2 Down JL. *On some of the mental affections of childhood and youth*. Oxford: MacKeith Press/Blackwell, 1990.

A new edition of what was Jelliffe's textbook of 'tropical paediatrics' is very welcome. Jelliffe in his foreword to the 4th edition talks of two main routes to be balanced in the effective promotion of child health, that is, specialisation, and child health and community. This book encompasses both routes. There are six sections each with its own editor and 15-20 contributors. The first three general sections cover maternal and child health, maternal and neonatal care, growth and development. Two disease specific sections follow, that is, infectious disease and diseases of systems. There is a final section on practical aids.

The contents contained everything it should. Although it is invidious to praise specific contributions, my favourites were: immunisation, protein energy malnutrition, and tuberculosis; other people might choose others. One problem I found was that some subjects were divided so that at times it was difficult to track down the relevant parts in the book, for example, gastroenteritis is dealt with in maternal and child health (oral rehydration services) and in infectious disease (diarrhoeal diseases). Nutritional supplementation programmes appear in the maternal and child health section, and nutritional rehabilitation is in the nutrition section of growth and development. In such a large book it was difficult to get an overall feel for the major medical causes of child mortality, and morbidity in the topics. The top three, that is, respiratory infection, diarrhoeal disease, and protein energy deficiency are all there and are dealt with effectively but get lost to some extent in the pith of the book. The remainder of the top 10 such as malaria, anaemia, tuberculosis, and so on are again dealt with well but their relative importance to overall child health is not clear. The introductory chapter might have set the scene better. Informative as it is, the chapter contains very selected mortality statistics but little breakdown by age and cause, and few measurements of morbidity. Perhaps my difficulties show I have become wedded to the medical model while the authors have escaped this straight jacket. The chapters/sections on cultural factors, delivery of individual services, the doctor as teacher, and child care in refugee situations are fascinating. They would not have been included in a more traditional approach and this book is much stronger as a result.

The editors say in their preface 'A balance has to be struck between the assembly of the information and instruction needed by the paediatrician in the reference centres of excellence, and the study and practice of management at the level of primary care'. The authors have achieved a commendable balance of emphasis in this very wide field.

BRIAN WHARTON
Rank professor of human nutrition

chapters on specific infections, namely measles, ehrlichiosis, and human herpes virus 6 (HHV-6) and four chapters addressing more general issues such as viral encephalitis, infected burns, skeletal infections, and the screening of immigrant children.

As the entire book emanates from US based authors, inevitably not all material presented is applicable to British or Australian paediatric practice. The authorship, as with previous volumes, comprises several recognised leaders in paediatric infectious disease.

True to its objective, this volume covers issues of contemporary concern with references right up to 1990. This is exemplified by the chapter on new developments in vaccines. It documents the change in US policy regarding the recommended age of immunisation against *H influenzae* type b. In March 1990, universal immunisation was recommended at 15 months of age instead of 18 months, but by October last year, this was again revised to recommend immunisation from 2 months of age, a change which was hinted at as pending in the text.

The infectious aetiology of several paediatric syndromes has been elucidated in recent years. This gratifying shift in understanding is reflected in the chapter on HHV-6, the cause of roseola infantum, also known rather appropriately as sixth disease.

The ongoing problems presented by pertussis and measles are expertly covered, though it is odd that the issue of pertussis should be covered again since it appeared in the previous volume. Also surprising was that in the authors' presentation of their contrasting views on the timing of licensure of acellular pertussis vaccines, one argued anecdotally regarding the reactogenicity of whole cell pertussis vaccine.

In the chapters on viral encephalitis and screening immigrant children, the authors have taken a practical approach in which they document their own strategies of management. The topics of infected burns and skeletal infections are dealt with ably and comprehensively.

The book is attractively presented and well referenced and apart from confusing staphylococcus with streptococcus in two tables dealing with paediatric skeletal infections, there are few typographical errors. It is of relevance to those practicing in the paediatric infectious disease field. Given that infection is the 'bread and butter' of general paediatric practice, most chapters in this volume will be appreciated by the general paediatrician who is keen to keep abreast of advances in this interesting field.

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Childhood Cancer: Understanding and Coping. By Henry Ekert. (Pp 185; \$16 paperback.) Gordon and Breach Science Publishers, 1989. ISBN 2-88124-356-8.

When a child has cancer what are the needs of those around him as regards information? First and foremost, of course, they need a sympathetic paediatric oncologist who has the time to sit down and explain the problem in detail. As there are so many factors that are peculiar to the individual child it may be misleading to turn to books for information without guidance.

Dr Ekert's book is intended for parents, relatives, and friends of children and adolescents with cancer. Of the 22 chapters, the first

Advances in Pediatric Infectious Diseases. Volume 6. By S Aronoff, W Hughes, S Kohl, W Speck, and E Wald. (Pp 200; £36.50 hardback.) Mosby Year Book, 1991. ISBN 0-8151-0308-5.

The editors of this series have again provided a useful and interesting update of important topics in paediatric infectious disease.

This volume covers new developments in vaccines against four major paediatric pathogens: *Haemophilus influenzae* type b, pertussis, rotavirus, and varicella. There are also three

BOOK REVIEWS

Diseases of Children in the Subtropics and Tropics. 4th Ed. Edited by P Stanfield, M Brueton, M Chan, M Parkin, and T Waterson. (Pp 1076; £90 hardback.) Edward Arnold, 1991. ISBN 0-340-50633-4.