

relative roles of pulse oximetry and skin electrodes in monitoring oxygenation both with and without an indwelling arterial catheter. There are too many abbreviations; 77 are listed in the glossary.

The publishers claim it is the first book to deal specifically with the baby under 1000 g. This book would be enjoyed by any paediatrician who looks after very preterm babies.

B W LLOYD
Consultant paediatrician

Celebration. By Margaret Spufford. (Pp 121: £2.95 paperback.) Collins, 1989. ISBN 0-00-627449-8.

This is a remarkable book. Written by the mother, it describes the discovery and progress of the daughter's cystinosis and the mother's own agonisingly painful idiopathic osteoporosis. It shows the need despite these afflictions to provide as loving and as normal a family environment both for the son's childhood and one in which, despite the family's extra tribulations, the father could function and work as the breadwinner. It is written with the impartial clarity of her profession as an academic historian. Even when describing severe stress and extreme suffering there are discerning and important observations about the medical world including occasions when despite the most stressful circumstances, the family could see with wry humour the absurdity of the predicaments in which they found themselves.

While the doctors sought to establish the diagnosis of cystinosis in her daughter by a series of invasive tests 'now I discovered, as I held a baby who was increasingly only capable of screaming "Mummy" that my instinct to defend her threatened to override my instinct to protect her in the long term, and therefore to get a diagnosis'. 'We were long past wanting her to live at any cost for possessive reasons, because we had so much wanted this child. What was intolerable was watching her learn fear'. 'She learned fast. I am never going to be able to forget the sound of her screams'.

It is of the greatest importance at this time when reorganisation of the NHS is being considered to listen to Dr Margaret Spufford when she says 'I have come seriously to doubt whether life should be sustained when it can only be done through massive and repeated medical intervention, especially when there is no community care, apart from the family, to which these young adults can suitably move on'. It is a message of the greatest medical and social significance that this girl, who on three occasions had been prevented from dying by major medical procedures, had in her adolescence become friendless, and acutely lonely. Sadly the parents realised that while they had been able to help a baby and a small child that they were much less able to help a girl of 18. They realised that she had lost her peer group as a result of the innate effects of the disease and of too much isolating medical experience. 'By definition she needed such a group above all things. As parents we had become less resilient to conquer the next obstacle in our fifties than in our early thirties. We had learned by now that there always is a next obstacle'.

Why then call her experiences 'Celebration'? In this short review it is not possible to describe the difficult pathway by which Dr Margaret Spufford reached the stage when she could see that the family's experiences con-

tained genuine achievement and reason for celebration as well as so much pain and suffering.

While this book describes how one individual family won through and how her religious faith was crucial to the mother's personal contribution, there is help here for people of many different beliefs as well as practical advice for both those needing care and those who provide it whether they are religious or non-religious. This is an important book for all those involved with medical ethics as well as those concerned with the future of our health services both inside and outside hospitals.

F BRIMBLECOMBE
Honorary professor of child health

Health. By the children of Walsall.

This little booklet was written by children of varying ethnic origins who attend a school in Walsall. It deals with their reports of their illnesses and accidents: it reveals their attitudes, priorities, and imagination.

The first priority would appear to be 'How many days off school for this'—they can tell you almost to the day!

Heroism—its here. 'I fell down my grandad's stairs—a four inch nail went through my head. Nobody knew for three weeks'. You'd have thought they would have noticed when combing her hair!

Misfortune—'When I was two, I was crying because I had fallen over. My mother took me upstairs to cheer me up. After a while I fell downstairs'. Shades of Gerard Hoffnung!

Attention to detail—'Over in America I had an accident in a car, I went in the car and I put my gear on and the car started to move. It hit a restaurant'.

Frustration—'When I was born, I turned blue. For two weeks I was stuck in an incubator with half a plant pot on my head'. The seeds of an idea?

Children as therapists: tummy upset—'Drink brandy and wine and go to the toilet' (seems like a good idea!) and warts—'Rub some bacon on it'. Lumps—'If I had lumps, I will go to the doctor and get some medicine and lie down. In two days I will feel better'. Such precision. Something in my eye—'I would tell my brother to blow it'. Why? What had he done wrong? Cannot sleep—'I imagine a sheep and I say this sheep is green, it is 11 years old it is 7 stones and its name is Samantha and I just close my eyes when I'm saying it'.

We should all read this booklet. It is refreshing, joyful, and unstuffy. It reaffirmed my decision to look after children.

G MCCLURE
Consultant paediatrician

Paediatric Hepatology. By Stuart Tanner. (Current Reviews in Paediatrics.) (Pp 363; £29.50 hardback.) Churchill Livingstone, 1989. ISBN 0-443-03393-5.

This latest edition in the series of *Current Reviews in Paediatrics* published by Churchill Livingstone aims, along with the preceding editions, to provide an overview of a current topic relevant to acute care paediatrics. These books are published by single authors and represent the experience of someone at the forefront of the field. In this sense Dr Stuart Tanner is a more than suitable choice for this particular edition.

The book starts with a description of the assessment of liver function, including both physical examination and history taking, and the current developments in biochemical investigation and imaging. The chapter is a well drawn out introduction to the topic and contains many of the latest advances that are currently available. The succeeding chapters are of particular relevance to neonatologists in the dealing of both cholestatic jaundice and hepatitis in the neonate and infant. These are set out in an extremely practical way and furnished with a lot of useful, up to date references and quick and easy to follow tables. The section on hepatitis has at last made clear to me the complexities of antigen and antibody rises and falls and their aid in the diagnosis of, particularly, hepatitis B infections and have certainly cleared up several of the problems surrounding hepatitis B screening in the antenatal population.

The latter part of the book deals successively with metabolic diseases, more specifically Reye's syndrome, and the role of trace elements within the liver. In this section Dr Tanner has done an excellent job in calming the fears of the non-biochemists among us in providing a suitable and easily readable account of some of the more complex disorders of enzyme deficiencies within hepatology and I certainly found this section very helpful.

After the discussion of topics including cystic fibrosis, liver failure and cirrhosis, the book ends with a chapter on the current state of play in liver transplantation. This is an extremely important chapter with the current focus of media attention upon liver transplantation in young children and, in my opinion, almost justifies the reading of the book. The latest figures are presented in an easily accessible way and both the complications and indications for liver transplantation are well set out.

In conclusion, Dr Stuart Tanner, in producing the latest addition to this series has kept up the excellent tradition maintained by his predecessors. The book is moderately priced and as such should be available in most general paediatric departments and is certainly well worth reading for those people about to embark on the Membership examination.

N MEADOWS
Consultant paediatrician

Surfactant Replacement Therapy. Edited by Donald L Shapiro, Robert H Notter. (Pp 321; \$69.50 hardback.) Alan R Liss, 1989, ISBN 0-8451-4281-X.

In the past few years a deluge of knowledge of surfactant has opened on the unwary, jobbing paediatrician. The effect has been to excite, confuse, and alarm but, most importantly, to expose our ignorance. This volume of over 300 pages sets out to rectify this last problem while maintaining our excitement; these aims have happily been achieved.

The volume consists of two sections dealing with the fundamentals of lung surfactant and the clinical aspects of surfactant replacement therapy. The first is probably of more interest to the clinician as this is the area with which we are less familiar. Some little part will be above the heads of most of us but most is written lucidly and, whereas the subject is complicated, the reader will finish with a profound knowledge of the complexity of the subject.

The volume begins with an overview of surfactant, the historical background, and takes us forward to the concept of gene cloning. There follows a treatise on the physical chemistry and physiological activity of pulmonary surfactants and this chapter is succeeded by one on surfactant proteins, those mysterious molecules that are, at once, fundamental to the function of surfactants while being unwilling to yield their secrets.

The chapter on surfactant and lung liquid absorption at birth is revealing, at least to the reviewer, but one wonders if the only function of fetal lung liquid is 'a template for fetal lung growth'.

Alan Jobe takes us through the metabolism of lung surfactants explaining what is known, nudging us towards the unknown, raising questions along this way. One example—it would appear that exogenous surfactant does not impair endogenous surfactant synthesis but isn't this odd in a biofeedback system?

The chapter on immune complexes formed in response to surfactant administration is disturbing. Whereas no clinical effects of such complexes has been described, they may pose problems in the longer term.

The chapter by Bengt Robertson on microscopy both without and with surfactant replacement lends weight to the argument that such treatment improves lung function. Again, he raises problems of lung poor size and protein fluxes which will stir the reader from any intellectual lethargy.

The second half of the book deals with the variety of surfactants so far used in clinical trials. Much of the work has already been published so the reader is able to relax more into this section. Again, the authors are at pains not to be too hopeful, to draw attention to some potential disadvantages.

Altogether, I found this book stimulating and well worth reading. My major criticism is that, overall, it may be a little too enthusiastic. I doubt whether surfactant will prove to be a panacea and would have liked to see a discussion of surfactant failures. My other criticism is that, yet again, little is mentioned of European work—an all too common complaint of American books.

G MCCLURE
Consultant paediatrician

Paediatric Secrets. Edited by Richard A Polin, Mark F Ditmar. (Pp 447; £35.95 paperback.) Hanley and Belfus, 1989 (C V Mosby). ISBN 0-932883-14-1.

As medical literature continues to emerge at an overwhelming rate, it is refreshing to find a new book which is original, well presented, and a pleasure to use. The concept is simple: those questions commonly asked by American paediatric residents have been collected and then answered by a group of paediatricians associated with the Children's Hospital of Philadelphia. The areas covered include pathophysiology, clinical management, diagnostic tricks, specific diseases and definitions, useful physiological data, and a few delightful oddities (Ondine's curse is compared with 'intern's curse', mortals who are affected by not sleeping when they breath!).

The title is misleading and does not do justice to the contents. Although it may be aimed at the resident (or senior house officer) who needs to have all the answers during a ward round, it has a much wider potential use. Candidates for the MRCP examination will use it for gathering information and paediatricians

will find it valuable for teaching sessions, *viva voce* examinations, and for clinical management information. Each major system has a separate chapter, and each question is followed by succinct, up to date information or advice. The information is presented as a discussion, or in lists and tables. There are a few line drawings and useful flow diagrams.

There is an inevitable American bias, and although some treatment advice is less useful in the UK (for example, the management of acute asthma or immunisation schedules) most information presented is pertinent and valuable. Some areas are particularly well covered, for example the treatment for necrotising enterocolitis and the discussion of Kawasaki disease are very good. It was wonderful to find precise information that is sometimes difficult to track down elsewhere.

There are a few errors and inconsistencies but the scope and accurate detail of the areas covered are remarkably comprehensive.

This is a fascinating, useful book and I would urge all paediatric departments to have a copy. Junior paediatricians can use it to outwit their seniors, or examiners and consultants can dip into it at leisure, remind themselves of the answers to 1819 questions, or use it as a valuable reference for clinical problems.

J STROOBANT
Consultant paediatrician

The Parents' Directory. Compiled by Fiona McDonald. (Pp 260; £5.95 paperback.) Bedford Square Press, 1989. ISBN 0-7199-1235-0.

Eight hundred voluntary organisations are listed in this helpful directory for parents. The variety of potential help is enormous. There are five main categories dealing with education, family welfare, handicap, health, and leisure. Finding one's way around this directory is slightly irksome. The rather incomplete index does mean that the reader has to wade through several categories before finding certain entries. One could quibble about some listings in the health and handicap categories. Despite these shortcomings, this book does provide a useful reference source for parents. Any professionals working with children should have access to a copy.

B M MACARDLE
Consultant paediatrician (community child health)

The Fragile X Syndrome. Edited by Kay E Davies. (Pp 135; £15.00 paperback.) Oxford University Press, 1989. ISBN 0-19-261836-9.

This book draws together in a compact work the increasing volume of information on this the second commonest cause of mental retardation after Down's syndrome. The fragile X syndrome, recognisable cytogenetically under certain conditions by a gap (fragile site) at Xq27 on the X chromosome, is unusual among X linked disorders in that as many as one third of obligate carrier females are mentally retarded. Even more fascinating is the fact that the condition may be transmitted through clinically and cytogenetically normal males.

Five authors, distinguished in their respective fields, cover the important aspects of the clinical manifestations, epidemiology, behaviour and treatment, and DNA and cytogenetic studies.

Paediatricians and clinical geneticists alike will find Jean-Pierre Fryns' clinical descrip-

tions and accompanying photographs useful; the classical clinical triad of moderate mental retardation, long face with large everted ears and macro-orchidism often seen in the post-pubertal fragile X male is less evident in the prepubertal male in whom relative macrocephaly is more striking and macro-orchidism is uncommon. Randi Hagerman emphasises the important correlation of the fragile X syndrome with autism and describes the characteristic behavioural patterns of affected individuals, which not only provide important clues for the diagnosis but also allow appropriate treatment and educational programmes to be devised. Useful comments are included on seeking and treating associated problems, such as pes planus, strabismus, hernia, scoliosis and mitral valve prolapse, which is said to affect as many as 50% of males. Treatment with folic acid is perhaps a more controversial issue than the author suggests.

The syndrome occurs in all ethnic groups and Tessa Webb's chapter on epidemiology emphasises the need to define the populations for which prevalence figures are being derived.

At the molecular level, a number of theories have been proposed by various authors to explain the basis of this unique disorder; these are lucidly summarised by Ted Brown. The gene itself is proving to be elusive and it has been difficult to identify DNA markers which are consistently close to the fragile X locus. Ted Brown describes, however, how in some families, use of flanking DNA markers may help in carrier detection studies.

Neils Tommerup highlights the cytogenetic 'catch 22'; males and females with non-specific mental retardation ideally should be tested cytogenetically for the fragile X marker but this adds a heavy workload to the laboratories, as many cells need to be screened (only a proportion express the fragile X marker) and special culture conditions must be met.

One minor criticism is that most of the authors refer to the 'female heterozygote' without defining whether this refers to obligate carriers on pedigree grounds or those who manifest the fragile X marker, or both. One small point of omission is that no specific mention is made of the fact that, unlike in other X linked disorders, most mothers of a boy with fragile X are thought to be carriers.

This book will be of great value in promoting awareness and understanding of this important disorder among paediatricians, who may well wish to purchase their own copy. The genetic idiosyncrasies of the disorder (for example, only 50% of obligate carrier females express the fragile X marker) highlight the need for referral to a clinical geneticist for carrier detection studies in females (and males) at risk, and for prenatal diagnosis counselling. Clinical geneticists themselves will find the book useful; Neils Tommerup's check list for prenatal diagnosis being an example. Cytogeneticists and molecular geneticists will also find a helpful 'state of the art' summary of the fragile X syndrome.

E M THOMPSON
Consultant clinical geneticist

Sudden Infant Death Syndrome. Edited by J Culbertson, H Krous, R Bendell. (Pp 264+xix; £29.95 hardback.) Edward Arnold, 1988. ISBN 0-340-49381-6.

In contrast to the recent preoccupation with the role of suffocation in the sudden infant death syndrome (SIDS) both in the medical



Surfactant Replacement Therapy

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