Candidates for the MRCP examination will round, it has occurred to one sur- factant protein that this is not only un- willing to yield its secrets. The chapter on surfactant and lung liquid absorption at birth is refreshing, 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 have 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 pub- lished so the reader is able to relax more into this section. Again, the authors are at pains not 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 dis- cussion of surfactant failures. My other criticism is that, yet again, little is mentioned of Euro- pean work, which is all too common complaint of American books.

G MCCLURE Consultant paediatrician


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 defini- tions, useful physiological data, and a few delightful oddities (Ondine’s curse is com- pared 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 book as it may be alluring to 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, entry to examinations, and for clinical management information. Each main system of the book (cardiovascular, etc.) 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 valu- able. 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 out- line their sessions, and seniors who 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 STROOANT Consultant paediatrician


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 educa- tion, family welfare, handicap, health, and leisure. Finding one’s way around this direc- tory 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 missing voluntary organisations in the headings, but 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 MACARDEL Consultant paediatrician (community child health)


This book draws together in a compact work the increasing volume of information on this the second commonest cause of mental retar- dation 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 men- tally retarded. Even more fascinating is the fact that the condition may be transmitted through clinically and cytogenetically normal males.

Five authors, distinguished in their respec- tive fields, cover the important aspects of the clinical manifestations, epidemiology, behav- iour and treatment, and DNA and cyto- genetic 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 is often seen in the prepubertal fragile X male is less evident in the prepubertal male in whom relative macro- cephalia is more striking and macro-orchidism is uncommon. Robert Harper emphasises the important correlation of the fragile X syn- drome with autism and describes the charac- teristic behavioural patterns of affected individuals, which not only provide important clues for the diagnosis but also allow appropri- ate 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. Treat- ment with folic acid is perhaps a more con- troversial 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 developed.

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.

Neil 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 labora- tories, 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 oblig- ate carriers on pedigree grounds for those who manifest the fragile X marker, or both. One small point of omission is that no specific men- tion 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 promot- ing 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 coun- selling. Clinical geneticists themselves will find the book useful; Neilis Tommerup’s check list for prenatal diagnosis being an example. Cytogeneticists and molecular gene- ticists will also find a helpful ‘state of the art’ summary of the fragile X syndrome.

E M THOMPSON Consultant geneticist


In contrast to the recent preoccupation with the role of suffocation in the sudden infant death syndrome (SIDS), the medical community is only just beginning to understand the pathophysiology of the condition. SIDS is the sudden, unexpected death of an apparently healthy infant under one year of age which cannot be attributed to a cause other than the syndrome (usually defined as death after a period of normal feeding and otherwise without obvious cause). A cause is defined as ‘obvious’ if it is the ‘immediate cause of death’ and ‘Brought to you by guest. Protected by copyright.