
Dr Serjeant is Director of the Medical Research Council Laboratories in Jamaica where there are 2000 cases of sickle cell disease under study with an additional 550 children followed from birth in a special cohort study. He describes the structure of haemoglobin, the distribution and different types of the disease, its pathophysiology, the effects on different organs, and its treatment and epidemiology. There are 90 pages of references and good illustrations—in particular, some excellent pathological photographs by Dr L W Diggs of Memphis.

Paediatricians who look after cases of sickle cell disease will find out all about features as disparate as nutrition and growth, the hand-foot syndrome, cardiac function, and immunological state. Dr Serjeant is not an enthusiast for folate supplements. Prevention of pneumococcal sepsis in Jamaican toddlers and infants called for regular monthly injections of penicillin at home by nurses. Pneumococcal vaccine is recommended at 2 and 4 years.

The book is nicely set out and well written, with a good selection of figures and tables. I had expected a personal account of sickle cell disease in the West Indies but found an extensively referenced monograph on the clinical and scientific aspects. Sickle cell disease is, however, not only a physical illness; it produces major effects on the social and emotional lives of its sufferers. These are hardly touched on. In a chapter on the painful crisis only a couple of pages cover treatment, with two skimpy paragraphs on pain relief. Dr Serjeant acknowledges that the management of pain is heavily influenced by social and cultural factors; but even so, it seems that Jamaican patients get very little analgesia, and what there is is very simple. In this country control of painful crisis is a major problem for those affected and their doctors. Another subject of current interest is antenatal diagnosis; but its value is only briefly discussed. Its role in Jamaica is dismissed in four lines. What this book does best, and does well, is to describe the physical effects of sickle cell disease.


This book is designed to help paediatricians and others to diagnose and manage inborn errors of metabolism. It summarises the clinical and biochemical features of over 200 disorders grouped according to the area of metabolism affected. The biochemistry is well explained and the results of recent experimentation authoritatively discussed. Clinical descriptions are in general adequate, although some helpful points are omitted—for example, mouth ulceration and recurrent otitis in type 1B glycogen storage disease, hypotonia with hyperreflexia in some of the organic acidemias. Differential diagnosis is seldom discussed, and if you require a list of biochemical tests you should consider in a child presenting with recurrent encephalopathy or with developmental delay and coarse facial features you will have to construct your own as you read this book.

Advice to the paediatric chemical pathologist is patchy. In the chapters on mucopolysaccharidoses and disorders of glycoprotein catabolism there are details of which substrates are to be preferred for enzyme assays, but the urea cycle disorders are discussed without any reference to the pitfalls of estimation of plasma ammonia concentration, and urinary organic acid determination is deemed too complex to be discussed at all. Treatment is covered, but details of management of the acutely ill metabolic patient are lacking. Prenatal diagnosis is authoritatively discussed, and the book has a very comprehensive reference list (occupying 185 out of 670 pages).

Comparison with The Metabolic Basis of Inherited Disease by Stanbury et al is inevitable. The authors have succeeded in covering most of the same ground in a smaller and cheaper volume, thus answering the prayer of every new junior doctor on the metabolic unit. I will suggest they read it, but I continue to nurture the hope that someone will write a problem orientated metabolic book. Such a book would be more likely to increase the number of diagnoses we make than this one.


Hyperactivity is a tricky subject.

In the first part of this book the authors describe their understanding of hyperactivity and its relation to other disorders, suggested causes, and contributory factors. The second part discusses management techniques, with a good deal of practical detail. Professor Rutter's introduction commends the book but highlights many of the uncertainties in dealing with the child. The point is well made that young children almost never refer themselves for diagnosis and treatment but are seen because someone else finds their behaviour unacceptable and disturbing. It would be interesting to know more of the children's views.

Attempts to distinguish hyperactivity from a situation dependent conduct disorder do not seem fruitful. The hyperspecificity of the term 'attention deficit disorder' is also unsatisfactory. On the whole, children are more active than adults would like them to be.

There is a useful discussion of adolescence and prognosis into adult life. The references are extensive, up to date, and international, being mainly drawn from the psychiatry and psychology published works.

Although the authors have been diligent and it is difficult to disagree with much of the content, the style is slow moving. It sent me to sleep!