
This book is designed to provide information for patients with neuromuscular diseases and their families “so that they may circumvent some of the ‘road blocks’ in the health care system and so profit from the many medical advances available today”. Professor Ringel is Director of the Neuromuscular Disease Clinic in Denver, Colorado and he has written a short readable book which is packed with insights and tips about the management of neuromuscular disease. The first 40 or so pages are brief descriptions of nerve and muscle and the diseases which affect them. There follows a chapter on genetic counselling. The following 70 pages concern therapy (imagine that distribution in a neurology textbook!) and include chapters on physical therapy, bracing and surgery, aids to daily living, respiratory and speech therapy (the former including a discussion on the indications for ventilation), nutrition and exercise, drugs and emotional adjustment to disability. Two short sections on educational opportunities and various government and community resources complete the book—these latter chapters will be of limited interest to the UK reader since they apply specifically to the US.

Who can benefit from this book? Some of our patients and their relatives (notably the parents of boys with Duchenne dystrophy) will certainly learn more about their condition. For example, sound advice is often lacking about the type of foods most appropriate in neurogenic dysphagia; patients battle to find out by bitter experience and the alternatives to choking may never be discussed. Relatives are frequently not taught how to do a Heimlich manoeuvre; it may not always work but it often does and it relieves that intense feeling of helplessness which relatives feel when faced with acute aspiration.

The decision about whether or not to be ventilated is an issue nowadays—one which it is appropriate to talk about to the patient and family. Doctors and “health care providers” too may find much valuable advice here.

This book is helpful because it discusses many issues of therapy in an open and informed way. It will raise the expectations of patients as to what they may expect of their “specialist” doctors and the para-medical departments. It will help to banish finally the notion that any neuromuscular disease is untreatable and I commend it to all who look after patients with these conditions and to the many patients who wish for more information but cannot get it in the clinic.

CM WILES


This is obligatory reading for every neurologist. To quote the editor: “Physicians have a responsibility to care for patients with incurable diseases. The negative emotional responses of avoidance, rejection, inadequacy, frustration and feelings of impotence and loss of control that ALS brings out in physicians have resulted in the common practice of telling the patient with ALS: ‘There is nothing that can be done. Go home, and prepare to die.’”

It is because neurology is more than diagnosis that this book is essential reading. Of the 21 chapters, only four are concerned with purely medical considerations. The chapter on ethical issues by the editor puts in a nutshell what should be done regarding telling the patient about this fatal disease. Three of the chapters concern swallowing and feeding problems; four are concerned with physiotherapy and the various devices that can be used to help, while other chapters concern the psychological, sociological and nursing requirements. There is also a chapter on communication problems, the final chapter being one on voluntary organisations that help with this difficult disease. There are two chapters on the use of machines when patients develop respiratory failure, and these are perhaps not strictly relevant to practice in the United Kingdom. This difference in management between the United States and the United Kingdom is of great interest, and probably not dependent solely on the financial pattern of health care.

The editor, who tragically died shortly before its publication, was in charge of a special clinic for MND patients at Mount Sinai Hospital in New York, and devoted his life to this one disease. There are other similar centres in Chicago, Philadelphia and Miami, all supported by charitable bodies. The book is soft-backed and costs less than £20. For anyone dealing with this sad disorder, it is the best buy on the market.

F CLIFFORD RENSE


This book about sleep in children is equally about parents fighting with their children at bedtime and again in the middle of the night. Most of the 20 authors work in sleep laboratories or departments of psychiatry, not paediatric clinics. There is a slant towards American children. The book is in two parts—part 1 “presents normative data” and part 2 is dedicated to pathological phenomena”—which, translated, means there are 128 pages on normal sleep habits, and 179 pages on colicky infants and problems such as the sleep of children with epilepsy, mental retardation and “brain impairment”—whatever that may be.

A lot of the book is valuable, with excellent and full documentation of maturation of sleep and breathing patterns through infancy and childhood to adolescence. The establishment of circadian control is discussed in detail, and paralleling these studies there are sections about normal psychologi- cal development, the give-and-take relationship between infants and parents as well as about social and psychiatric factors that disturb the sleep of children. Sections on disease are especially well done, including those on narcolepsy, sleep apnoea and other respiratory disorders in children, although here the causes, pathophysiology and management are not very different from those in adults. Guillemainault’s wide experience of infantile sleep apnoea and SIDS, as well as his studies of infective mononucleosis in adolescence, deserve special recognition.

There are a number of problems. The overall standard of writing is laboured, with a fair amount of jargon, making detailed reading difficult. Also, the viewpoints of some of the authors is a little narrow. Clear advice on treatment is seldom given. What do statements such as “improved sleep
hygiene and sleep schedule, sleep restriction therapy, relaxation training, biofeedback, chronotherapy and psychotherapy have a "place" (in the treatment of adolescent insomnia) amount to in real terms? What do the authors actually do? Is it surprising that a history of colic 'significantly co-varies with the parents' judgment that night waking is a current problem (44) (\( \chi^2 = 7.4, p = 0.007 \))? Diagnostic polysomnographic features are not common among adults with complaints of significant insomnia, and neither H reflex studies nor CSF measurement of amine metabolites, are clinically useful in the diagnosis of childhood narcolepsy and cataplexy. And what to make of the Lochower phenomenon, described as "a pre-sleep experience including blurred distinctions between parts of the body, tactile and vestibular sensation of fullness, turnings or pressings, and sounds of murmuring or visions of indistinct roundness"? Does the statement that somnambulistic children have significantly more inhibited aggression (\( p = 0.05 \)) than non-somnambulists on Rorschach tests have any meaning? There is a lot more like this.

So this book is a strange mixture. It needs more paediatric input and more linkage with general medicine. However, it is a useful source of data about both normal and abnormal sleep in children.

JDParkts


In this age of multi-authored texts it is refreshing to read one written by an individual. The psychosocial consequences of brain injury, which occurs most frequently in young adults, are often profound and lasting and it is an indictment of our society that so little provision has been made for this group of patients. As neuropsychologist at the Kensley Brain Injury Rehabilitation Unit, Dr Wood has contributed to the pioneering work in the management of severely brain damaged individuals with behavioural disturbances carried out there. This book is largely descriptive of that experience. The theories of causation of behavioural disturbances after brain injury and the terms used to define them are critically discussed, the case being made for a neurobehavioural rather than a traditional psychiatric approach. The treatment strategies for aggression, sexual disorders, unacceptable habits and attentional problems are outlined with frequent case illustrations. The author argues cogently for the single case design method for the measurement of the efficacy of the interventions used. The nature of the complex disturbances after brain damage justifies this approach but some attention to the overall numbers, the failures as well as the successes and the longer term results of the treatment methods used would have been interesting. It would also have been useful to have the author's views on the applicability of the neurobehavioural approach in settings other than the specialised unit devoted to brain damage rehabilitation as the majority of such patients continue to be managed in surgical, psychiatric or general rehabilitation wards.

For its price the book is not particularly attractively produced. It will be of interest primarily to psychologists but should be read by other neuroscientists including nursing and remedial therapy professions involved in brain injury rehabilitation. Unlikely though it is, it would be pleasing to think that health care planners and community medicine specialists would read it as it would give them insight into the problems these patients exhibit and the cost in terms of human effort and facilities needed to help them.

Brian Pentland


This book is the first in a series on neurochemistry and neuropharmacology. The next three volumes are to discuss iron metabolism in the brain, depression and the neurochemistry of alcohol. Each book will be for a rather specialised audience and the current volume which is excellent will be read and bought by all those interested in pterins, as there has previously been a dearth of books on this subject, particularly one that combines both basic and clinical aspects. For clinicians perhaps only those with an interest in neurochemistry, particularly that of dementia, Parkinsonism and dystonia, will have any great interest here although paediatric neurologists seeing patients with phenylketonuria or its rarer variants will wish to read at least some of the chapters.

Unconjugated pterins were first discovered a hundred years ago as a new pigment in butterfly wings. In 1963 tetrahydrobiphenol (\( BH_4 \)) was discovered to be the naturally occurring essential cofactor for phenylalanine hydroxylase. Later studies demonstrated its key position in the regulation of the synthesis of serotonin and the catecholamines and led to the current interest in this substance. Subsequently rare forms of hyperphenylalaninemia were discovered that are separate from classical phenylketonuria and are due to faults that lead to under-availability of \( BH_4 \). These include a deficiency of the enzyme crucial to salvaging the cofactor after it has been oxidised during hydroxylase activity and two forms where the synthesis of \( BH_4 \) is deficient. The biochemical and clinical consequences of these different forms of hyperphenylalaninemia are discussed and are of importance as their current treatment is somewhat different and may become very different in the future. In neurological disease of adult life the role of \( BH_4 \) is somewhat more speculative but the data concerning its abnormalities in other diseases, particularly dementia, Parkinsonism, generalised dystonia, lead and aluminium poisoning, depression, ageing and oestrogen therapy are all discussed critically. The bulk of the book discusses the chemistry, regulation of biosynthesis and distribution of pterins and this will chiefly be of interest to neurochemists who I am sure would all speak very highly of the chapters, their authors, and the general readability of their contributions.

AC Williams


The first edition of this book was published in 1985. The need for a second edition now speaks well of its reception and also hints at changes in therapy during such a short interval. In reviewing the first edition, I warned that it was not a book to be used by initiates without supervision. This comment still pertains because the policy which makes each section so interesting (the expert contributor being advised to state his personal method of management) leads to contentious statements which could mislead the novice. The authors have been changed to provide new viewpoints and 17 new chapters added.

As in the first edition, coverage is remark-