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Book reviews

Duchenne Muscular Dystrophy. By A E H Emery. Revised edition. Pp 317: £17·50 paperback. Oxford Medical Publications, 1988. ISBN 0-19-261798-2.

These are exciting times for those involved with Duchenne muscular dystrophy. Now that the gene has been isolated and sequenced, and the protein 'dystrophin' which it encodes identified, we are at last established on the road to understanding the pathogenesis of the dystrophic process. Considering the tremendous interest in this disorder, there is clearly a need for a book that draws together all the various strands of current knowledge, and this is ably provided by Emery.

The author has covered every conceivably relevant aspect with chapters on the clinical features, methods of investigation, differential diagnosis, biochemistry, pathogenesis, genetics, molecular pathology, management, and more besides. It starts with a fascinating chapter on the history of the disease, from ancient Egypt to modern times, and ends aptly with Duchenne's obituary in the *Lancet.* The book contains many excellent illustrations. The text is commendably clear, particularly when explaining difficult concepts such as the use of DNA probes and the mathematics of risk calculation.

Inevitably one looks to see if the author has been able to keep the book up to date in this revised edition. He has included a section on dystrophin including important 1988 references. He has also discussed the various animal models, including the MDX mouse and the retriever dog, but was not able to indicate fully their relevance to the human condition. The only other omission is the theory of mechanical damage in the evolution of the dystrophic pathology, which is looking increasingly more relevant as time goes by. These comments notwithstanding, this excellent book is of potential value to any physician or basic scientist involved with this disease.

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Prader-Willi Syndrome. Edited by M Caldwell and R Taylor, Pp 110: hardback. Price not stated. Springer-Verlag, 1988. ISBN 3-540-96699-4.

The mere mention of Prader-Willi syndrome engenders in most doctors a blank look of incomprehension, but it is heartening that paediatricians are increasingly aware of the syndrome. This book presents 'selected research and management issues'; in fact it is mainly a review of the literature flavoured with personal experience of the two editors, both North American educationalists, and a psychologist, paediatrician, paediatric surgeon, and a geneticist.

The clinical features are first outlined, but is height really 'within normal limits during the first ten years of life'? Most surprisingly though, no mention of the characteristic facies is included although 'dysmorphic facial features' are referred to in a later useful chapter on aetiology. This deals mainly with the role of chromosome 15q12 stressing the importance of special chromosome preparations examined by experts accustomed to the syndrome. The hypothalmic disorder in the syndrome might usefully have been included here. 'Surgical Considerations' tackles problems that exercise both doctors and parents—well illustrated by an opening sentence '. . . non-surgical procedures usually succeed at least to a better level of satisfaction than surgery . . .'. Intestinal bypasses to prevent and cosmetic surgery to excise obesity both have complications which have to be balanced against possible benefit. Although poor testicular development at biopsy is quoted, orchidopexy is generally favoured, but I wonder whether this is justified? After all, testicular malignancy in association with the syndrome seems never to have been reported and cosmetic considerations seem to be more of an anxiety of parents than of patients. The necessary cooperation of the patient to benefit from surgery for scoliosis is discussed as are problems of intubation and hypothermia during anaesthesia, which sometimes complicate any operation in this syndrome.

Two chapters are about cognition and behaviour and its management but hypotheses outrun practical advice. Similarly, 'Management of Problems of Infancy' resolves into that of a hypotonic baby whatever the cause. Finally a chapter on 'Parent Concerns', based principally on an analysis of a questionnaire sent to only 12 families, emphasises the anxieties of parents and the need for all carers to communicate with them.

Most of all though, it is early diagnosis and a diagnostic 'label' that helps. The Prader-Willi Association in America is the most useful solace for parents, reflecting the experience of its counterpart in the United Kingdom. For those interested in the syndrome this is a useful book, but I could not give it a high priority on a medical library list.

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Perinatal Events and Brain Damage in Surviving Children. Edited by F Kubli, N Patel, W Schmidt, and O Linderkamp. Pp 338: DM 120 hardback. Springer-Verlag, 1988. ISBN 3-540-18111-3.

'The book of the conference' is often in danger of being a disappointment because it is usually published so long after the meeting that prospective readers feel it must surely be out of date, and the written word seems to lack the impact and immediacy of the spoken papers. In this case the editors have avoided these defects and achieved success with a book on a conference held in Heidelberg in 1986, organised by the committee on Perinatal Mortality and Morbidity of the International Society of Gynaecology and Obstetrics (FIGO). The conference was designed to explore the relationship between events in pregnancy and neurodevelopmental handicap in the offspring with the aim of 'separating scientifically based knowledge from myths'. The majority of the 62 contributors came from Europe and Scandinavia with a handful from North America and Uruguay. They covered the specialties of paediatrics, obstetrics, pathology, physiology, and epidemiology and the list of their names reads like an international Who's

Who in perinatology, consequently each of the papers has an air of authority about it.

The book itself is in seven sections each containing four to six papers, these have been written not as transcripts of the spoken word but as short chapters mainly in the form of a review of the current state of the topic with some also including the author's original data. The first section on the epidemiology of neurodevelopmental disorders is followed by sections on the pathogenesis of handicap, intrauterine growth retardation and chronic hypoxia, and acute intrapartum asphyxia and birth trauma. The conference then turned to the preterm and very low birthweight baby with papers on intraventricular haemorrhage and ischaemia, and reviews of the changing outcome for these infants showing improved survival and decreased morbidity. Finally, there are reports of four recent follow up studies from Dundee, Uruguay, Finland/Bavaria, and Hamburg.

I found this book to be much more than just the report of a conference, it contains a wealth of information of relevance to both paediatricians and obstetricians providing clear up to date views on an important area of perinatal care. The papers are well referenced but unfortunately there is no subject index, consequently there is no alternative but to read the book from cover to cover.

One of the contributors started his chapter with a quotation from Freud (1897) 'Difficult birth in itself in certain cases is merely a symptom of deeper effects that influenced the development of the fetus'. We are just beginning to appreciate how right Freud was, perhaps the lawyers ought to read this book as well, certainly all paediatricians should.

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Manual of Hospital Paediatrics. By Garry Hambleton. Pp 360: £14·95 paperback. Churchill Livingstone, 1988. ISBN 0–443–03512–1.

The Concise Oxford Dictionary defines 'manual' as a small book for handy use. As a description of the author's goal it would be difficult to better. Rest assured that Dr Hambleton achieves it, and in so doing invites favourable comparison with another English Midlands paediatric institution's handbook with a latin title.

The manual is well laid out, defining or describing each condition, listing causes, symptoms and signs, differential diagnosis, investigation, and treatment. Where relevant, simple practical advice on management and prognosis is offered, giving clear guidance as to what to expect and, most importantly for a junior paediatrician, what to tell the parents. Its scope ranges from the emergency situation to the common or garden complaints that all too often present in the small hours, such as a blocked nose or a painful ear, though sleep disturbance is not mentioned!

Five appendices include normal values, drug dosages, fluid and electrolyte needs, blood taking, and how to do a cut down. Commonly available growth charts are generously allotted 15 pages. Despite being barely large enough to use, they are infinitely preferable to tables of anthropomorphic data.

In two situations only could one find contentious advice: hyposensitisation is recommended in favour of systemic antihistamines and in suspected meningitis and established coma no warning is given about the danger of coning in the absence of papilloedema. This is a distinctly useful addition to many a houseman's pocket, and at an affordable price, therefore I shall be recommending it to my junior paediatric staff.

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