

## 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 conceivable 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 hypothalamic 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