Duchenne Muscular Dystrophy

It showed great foresight on the part of Alan Emery and the publishers to produce a book devoted entirely to a disease that has probably had more written about it in the 1980s than any other condition. It was a wise decision: the monograph is not just an account of what everyone knows. It gives a comprehensive and critical assessment of every aspect of Duchenne muscular dystrophy, to suit research workers, clinicians, and physiotherapists and others who are concerned with the management of muscular dystrophy.

After a brief introduction to the physiology and anatomy of muscle, there is a fascinating history of the first descriptions of Duchenne muscular dystrophy. Here we learn that the first diagnosis was made by Dr Meryon of St Thomas's Hospital, who also reported an X linked pedigree, about 17 years before Duchenne. It is interesting later in the book to read a reprint of the obituary of Duchenne which appeared in the Lancet of 1875. Photographs of neurologists of the last century are accompanied by a modern one of P E Becker who described the more benign type of X linked muscular dystrophy during the life time of many patients alive today. Chapter 3 is remarkable for some valuable tables presenting the variation in the natural history of the disease, based on Professor Emery's large series of personal cases. These are followed by a critical account of investigations used in the diagnosis of Duchenne muscular dystrophy, and in its distinction from other neuromuscular diseases. Emery discusses biochemistry and possible pathogenetic mechanisms, and later there is a long and useful chapter on management, in relation both to individual patients and to the liaison between community services and lay organisations.

The chapters on genetics, genetic counselling, and molecular biology are particularly interesting to readers of this Journal. These chapters are very clear, practical, and up to date. The approach is wide ranging and non-controversial, although Professor Emery does point out some features of Duchenne muscular dystrophy that are not fully explained. He asks, for example: Why is the disease so common? Do postzygotic mutations occur? Why are some boys mentally retarded? Why is the proportion of female carriers detected through creatine kinase estimations different from that estimated from the proportion of affected sons? Why do DNA deletions fail to correspond with the clinical picture? Why do some families have more than one new mutation for Duchenne muscular dystrophy?

This book is timely and useful; it stimulates as well as teaches. I can recommend it for all those doctors, scientists, and paramedical staff who work with muscular dystrophy, and probably some parents would also find it interesting.

Sarah Bundy