PostScript

BOOK REVIEW

Huntington's disease

3rd edition. Editors Gillian Bates, Peter S Harper, and Lesley Jones. Oxford: Oxford University Press. 2002. ISBN 0 19 851060 8 (Hbk)

Peter Harper's first two editions of Huntington's disease have been widely regarded as key reference works for those working on this disease and related polyglutamine disorders. For the third edition, he has teamed up with Gillian Bates and Lesley Jones to produce a worthy successor to the previous editions.

This book covers a wide range of topics relevant to this condition with sections devoted to clinical features, genetics, neurobiology, structural biology, cell biology, animal studies, care of HD patients, and therapeutic approaches. The editors have assembled an authoritative group of authors with wide ranging expertise. In general, the chapters are meticulously researched, extensively referenced, and provide a careful documentation of the field.

I enjoyed reading this informative book and will certainly refer to it frequently. The chapters that were particularly memorable were those dealing with the history of Huntington's disease (Harper), the cell biology of HD (Jones), a comparison of HD with other polyglutamine diseases (Bates and Benn), and comprehensive care in HD (Nance and Westphal). The latter chapter is an excellent distillation of the experiences and strategies used by a group with expertise in long term management of HD. For those of us doing basic research, this chapter serves as a reality check for the great difficulties faced by HD patients, families, and carers. On the other hand, this chapter has useful advice for those involved in long term care of HD families.

In general, I found the reviews to be accurate. However, there were a few details that I did not agree with that may be of relevance. For instance, the chapter on HD genetics gives the impression that the penetrance of mutations in subjects with 36-39 CAG repeats may be predicted by their penetrance in the preceding generation. I am not aware of any robust data supporting this assertion. The chapter on the cell biology gives the impression that wild type huntingtin is not cleaved. My interpretation of a number of recent data is that wild type huntingtin is efficiently cleaved in vivo and in cell culture. There was also an apparent difference in opinion between the authors of chapters 2 and 5 as to whether the CAG repeats influence the progression of disease.

This book will be of great value to those working on HD and related polyglutamine diseases, including basic scientists, clinicians, and those involved in long term care. A major value in this book is that it provides the reader with the opportunity to see the disease from a number of facets. When I recommend this book to my colleagues in the laboratory, I will certainly highlight the chapters dealing with the clinical and care aspects of HD as well as those dealing with the basic science.

David C Rubinsztein

NOTICE

British Human Genetics Conference

The British Human Genetics Conference will be held on 15-17 September 2003 at the University of York, England. There will be Symposia on: Genetics of complex disorders, Unusual genetic mechanisms, Heterozygote - advantages and disadvantages, Duplicons and breakpoints, and Mechanisms of DNA repair and cancer predisposition. The Carter Lecture will be given by Professor Sir Alec Jeffreys on "Genetic fingerprinting and beyond: exploring human genome diversity and instability".

Further information from the Conference Office - British Society for Human Genetics, Clinical Genetics Unit, Birmingham Women's Hospital, Edgbaston, Birmingham B15 2TG, UK. Tel/fax: 0121 627 2634. Email: york2003@bshg.org.uk Website: http://www.bshg.org.uk