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BOOK REVIEW

Huntington's disease, third edition: Oxford monographs on medical genetics

Edited by Gillian Bates, Peter Harper and Lesley Jones. Oxford University Press Inc., New York, 2002

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Since its first edition, published in 1991, 'Huntington's disease' has been a classical source of information for all of us interested in the multiple aspects of Huntington's disease.

Published by Oxford University Press, the third edition has been renovated in design and content and integrated into the Oxford Monographs on Medical Genetics.

Editors G Bates, P Harper and L Jones included in the new edition an outstanding list of different authors, while keeping the spirit and highest quality of the former editions.

Giving the reader an overall and at the same time profound approach into the current knowledge of Huntington's disease, the different chapters reflect the great advances which occurred since the mapping of the HD gene about a decade ago; the practical influence this finding had on diagnosis, management and genetic counselling of patients and their families; the multiple lines of research aiming to determine the underlying etiopathology of the disease and the efforts to develop therapeutic solutions.

Notwithstanding a multiauthor book, it gives a sense of unity when reading through the different chapters.

The book is divided into six sections containing 17 chapters:

Section 1, which deals with the clinical aspects of Huntington's disease includes four chapters: Chapter 1 gives a historical perspective allowing for a more dynamic comprehension of the milestones in the development of clinical, epidemiological and genetic knowledge of the disease since first described almost 130–150 years ago. Chapter 2 presents the spectrum of clinical variability of the HD phenotype, its onset and progression with a helpful discussion on differential diagnosis. Chapter 3 introduces the neuropsychological and neuropsychiatric aspects of HD with an excellent review on the basis of behavioral problems in HD. Chapter 4 discusses applications of structural and functional imaging, its importance in the clinical assessment of patients and its role in research.

Section 2 on the genetics of HD includes three essential chapters covering the individual, familial and population genetic implications of HD.

Chapter 5 deals with basic Mendelian to non-Mendelian inheritance; CAG repeats, genomic imprinting, genotype–phenotype correlation and supporting evidence for the existence of genetic modifiers. The indications and limitations of molecular testing are exhaustively reviewed. Chapter 6 includes a comprehensive epidemiological picture of HD and it is very well complemented by Chapter 7 on medical, psychological, social implications of genetic counselling and presymptomatic testing at the individual and familial levels.

Section 3 is based on Neurobiology. This challenging section presents the newest research results from animal models and studies of the human brain in the field of neuropathology, neurochemistry and energy metabolism. Chapter 8 discusses the findings of huntingtin-related agreggates and cell loss; neurochemical alterations occurring in the brain of HD patients are presented in Chapter 9 and the evidence of defective energy metabolism in Chapter 10.

Section 4 deals with the structural biology, cell biology and mouse models of HD. Chapters 11–12 complement each other by giving a thorough and exciting review of current experimental results and directions for future research, while Chapter 13 summarizes the great success of mouse models and its role in developing effective therapeutics.

The inclusion of an excellent chapter on polyglutamine diseases – Section 5 – adds to the didactic importance of this book.

Section 6 on therapeutic interventions, comprises Chapter 15 on comprehensive care, Chapter 16 on therapeutic trials and Chapter 17, on cell and tissue transplants, offering matter for thought and hope, although there are still so many open questions.

An excellent updated bibliography, good illustrations and useful addresses included in all chapters add to the value of this book as an excellent resource for clinicians and basic researchers in human genetics and neurology sciences, as well as for the general reader.

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