

I will nitpick on a few points. The chapter on alpha-synuclein and PD was well written and referenced, however the conclusion was not a summary but a brief statement of recent advances and did not allow for proper closure. Another chapter had no article titles in the references. There was actually more clinical information on PINK1 parkinsonism in the first chapter on Neuropathology and Staging than in the chapter devoted solely to PINK1 parkinsonism. There is some overlap between the chapters, which isn't necessarily a bad thing.

The figures are in black and white which undoubtedly lowers the production cost. While colour photos or figures are nice, the lack of this does not significantly detract from the quality of the book.

This book is not intended for a practicing general neurologist. Even for a movement disorders clinician without particular basic science interest, I would suggest reading a review article on genetics and Parkinson's rather than purchasing this book.

I recommend "Parkinson's Disease – Genetics and Pathogenesis" for those with a particular interest in basic science, genetics, and animal models and how those interact in PD and PD models. It also makes for a great Neurology library reference book that may inspire future clinician-scientists to further our understanding of Parkinson's.

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MONOGRAPHS IN HUMAN GENETICS NEUROFIBROMATOSIS. VOLUME 16. 2008. Edited by Dieter Kaufmann. Published by Karger. 192 pages. Price C\$190.

"Neurofibromatosis" is one volume in a series of Monographs in Human Genetics focusing on Neurofibromatosis. It provides a detailed and timely review of our current knowledge of Neurofibromatosis Type 1 (NF-1), Neurofibromatosis Type 2 (NF-2) and Schwannomatosis, and specifically of the recent genetic advances in identification of mutations and molecular mechanisms of these conditions. The major focus of this book is on NF-1, given that this is, by far, the most common clinical entity of the three. The book's editor and most chapter authors are geneticists, so the main thrust is on the genetic aspects.

The first two chapters provide a comprehensive overview of the current classification, treatment and management of NF-1. The first chapter focuses on the definition and classification of these syndromes, and succinctly outlines the clinical features of NF-1. Included is a detailed table listing the various features and complications of this disease, and the age at presentation of those features, which would be very useful for the clinician caring for NF-1 patients. While specific subtypes of NF-1 are described which have much more uniform findings in families, the authors note that these account for only 5% of cases. For most NF-1 patients, there remains considerable variation in presentation among affected members. The second chapter focuses on management, and provides

good reviews of the challenges of surgical therapies for plexiform neurofibromas, and treatment of optic nerve gliomas, pseudoarthroses and malignant peripheral nerve sheath tumours.

The NF-1 protein, neurofibromin, is a tumour-suppressor gene, which downregulates the biological activity of the RAS pathway, thus regulating cell growth and differentiation. The third chapter reviews other disorders affecting the RAS system and outlines some of their overlapping phenotypes, as these conditions may provide clues on possible disease-causing genes and better therapies for malignancies associated with NF-1. The chapters focusing on gene structure, genotype/phenotype correlations (or lack thereof), NF-1 mutation analysis and NF-1 gene evolution were difficult reading and would be more relevant to a molecular geneticist than a clinical neurologist. The last two chapters in the NF-1 portion focus on the composition of neurofibromas and NF-1 expression and somatic NF-1 mutations in tumours and other tissue, and provide a useful overview of the pathogenesis of these lesions.

Two chapters are devoted to NF-2 and the role of merlin in tumor formation. The various mutation types, the higher prevalence of mosaicism (20-30%) in non-familial NF-2 cases and challenges of genetic counseling in such individuals, and the therapeutic challenges are well summarized. The final chapter is devoted to Schwannomatosis, and provides a brief overview of distinguishing features from NF-2 and the genetics of this condition.

In summary, this volume provides a timely update, mostly focusing on the recent advances in genetics and molecular mechanisms in these disorders. However, it is clear that for most patients, we are still far from being able to predict the phenotype based on the genotype. Given its main focus is on genetics and molecular mechanisms, this book would be most useful to Neurogeneticists or Neurologists with a strong interest in Neurogenetics.

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NEW ANIMAL MODELS OF HUMAN NEUROLOGICAL DISEASES. BIOVALLEY MONOGRAPHS. VOLUME 2. 2008. Edited by P. Poindron, P. Piguet. Published by Karger. 100 pages. Price C\$109.

This monograph is the second in a series of BioValley monographs; the BioValley being an area bordered by Basel, Fribourg, Strasbourg and Mulhouse. The text includes five monographs and an introductory chapter for a total of 100 pages. On the whole, it is well written although the exact aim of the text is a little unclear, particularly as 4 of the 6 chapters are scientific monographs while 1 (the second chapter) is a review article.

Chapter 1 provides a general overview of the development of animal models of human disease and gives a succinct summary of the relative pitfalls in over-interpretation of animal models. In particular, I found the emphasis on isomorphic versus homologous models useful and a nice framing of the subsequent chapters.