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.

Alex Rajput
Saskatoon, Saskatchewan, Canada

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

"Neurofibromatosis" is one volume is 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.

Elaine Wirrell
Rochester, Minnesota, USA

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 Bassel, Frieburg, 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.

Chapter 2 is a traditional review of the models of motor neuron degeneration and specifically focuses on amyotrophic lateral sclerosis. The tables are informative and the references parsimonious. The various transgenic models are dealt with well, including the relative strengths and weaknesses of each. A minor point, but one which all such texts must by definition suffer from, is the evolution of literature during publication. In this case, the reference to the minocycline trials in ALS is dated; we now know the drug was ineffective.

The remaining chapters are scientific papers. Chapter 3 describes a rat model of MOG-induced EAE. It would have benefited from a more robust comparison amongst the currently existing models. Chapter 4 describes the development of a new technique for the middle cerebral artery occlusion (MCAo) model of stroke using a detachable silicon plug. While also a scientific report, it does provide a robust comparison to other models and indeed, in the experimental design, two more traditional models of MCAo are included. It will be useful as a review of this area. Chapter 5 deals with the modeling of pyridoxine toxicity in which a sensory neuropathy with ataxia is induced. The introduction provides a brief but appropriate review of the major models of sensory neuropathy. It is of interest that this specific neuropathy is sensitive to nerve growth factor and hence will be of interest to those studying neurotrophic treatments for the peripheral neuropathies. Again, it is better suited to a scientific publication. The final chapter, number 6, speaks to a novel spontaneous mutation (frissonnant) which gives rise to locomotor instability with ataxia. The syndrome is responsive to dopaminergic therapies, but bears none of the hallmark pathology of such. Hence, it will be of interest to those studying the pharmacology of the dopamine depletion syndromes.

In summary, this is a relatively highly specialized monograph. It will be of interest to those with specific focus in modeling neurodegeneration, but of limited utility to a more general readership.

Michael J Strong
Arthur J Hudson Chair in ALS Research
London, Ontario, Canada