

## Dementia and Motor Neuron Disease

Traditional teaching was that motor neurone disease (MND) or amyotrophic lateral sclerosis (ALS) was a disorder exclusively of the motor system and that patients were cognitively preserved and hence all too horribly aware of their progressive neurological predicament. Occasional reports of MND with dementia (the first possibly by Alzheimer, in 1891; p 133) and of frontotemporal dementia (FTD) complicated by MND have now been succeeded by more systematic studies suggesting that overlap is common and that this may be a spectrum or multisystem disorder with purely motor and purely cognitive boundaries but with extensive overlap, shared pathophysiology and neuropathology.

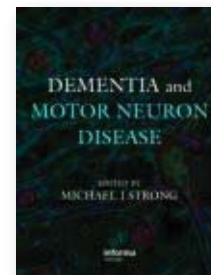
This book is based on papers presented at the First International Research Workshop on Frontotemporal Dementia in ALS held in London, Ontario, in 2005. Whereas workshop proceedings can often be somewhat arid for non-participants, the chapters in this volume do engage, in part because a work on the convergence of these two fields is timely. Individual chapters cover clinical and cognitive features of MND and FTD, neuroimaging findings and neuropathology, including a welcome piece on primary lateral sclerosis (8), with chapters devoted to disease in Japanese (7,15) and Guamanian (14) populations. To be sure, the multiplication of terminology (e.g. Tables 2.1 and 5.1, p 10 and 63) which may bewilder non-initiates, and which has to some extent inhibited taxonomy, has yet to be resolved, but the neuropathological division into disorders with tau positive inclusions (e.g. Pick's disease, FTDP-17) and disorders with ubiquitin inclusions (FTD-U) may help (Table 12.1, p

148). On the other hand, genetically defined FTDP-17 with tau gene mutations may encompass cases clinically defined as FTD, progressive supranuclear palsy, corticobasal degeneration, and Alzheimer's disease; furthermore P (= parkinsonism) may be absent. The role of tau in disease pathophysiology is discussed but generally there is little on mechanisms (e.g. apoptosis, ubiquitin-proteasome system).

This is a well-produced volume with high quality illustrations (odd, though, that throughout the book pathological images are without scale bars or magnifications, excepting some in chapter 12). From a clinical standpoint, it would have been interesting to learn how entities such as Mills syndrome (progressive hemiparesis) and pure hippocampal sclerosis (mentioned only in passing, p 170) fit into the spectrum. Unluckily for the editor, the recent discovery of progranulin mutations causing some cases of FTDP-17 already dates the book, although the dynactin mutation associated with familial ALS/FTD is mentioned (p 49). John Hardy, doyen of the genetics of neurodegenerative disease, suffers an unfortunate meltdown in his clinical knowledge at p 202 when he confuses dementia with bone cysts (Nasu-Hakola disease) with the syndrome of inclusion body myositis, Paget's disease, and frontal dementia (IBMPFD), suggesting VCP mutations occur in the former, rather than the latter. Easily done!

Nonetheless, minor reservations aside, neurologists with an interest in either MND or dementia may profit from reading this book.

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## Mc Alpine's Multiple Sclerosis - 4th Edition

*McAlpine's Multiple Sclerosis* has been the most authoritative text book on MS since the publication of its first edition in 1985. It was edited by Nigel Compston, father of the current editor. But its origins go back to 1955, to Douglas McAlpine's, *Multiple Sclerosis*. The book has had remarkable success and has remained the comprehensive gold standard reference text for MS.

The current edition (2005) has been extensively rewritten and updated. This is evidenced by the 1008 pages (previous edition had 592 pages) and the numerous high quality and visually appealing figures and tables. There are 4 new very experienced authors who are thought leaders in MS (Christian Confavreux, Ken Smith, David Miller and John Noseworthy). The book itself is divided into 4 sections: (1) The story of MS (2) Cause and course of MS (3) Clinical features and diagnosis (4) Pathogenesis and treatment. Each section is further divided into chapters - 19 in total. Each section ends with a thought provoking chapter that aims to identify and debate unresolved issues.

It has always been the style of the book to have a limited number of authors covers all aspects of MS. The natural concern of 'depth', when such a huge task is handled by few, is easily allayed as one cruises through the comprehensive and thoroughly referenced chapters written by authors who have contributed immensely and guided the course of MS research into the 21st century. However it's important to know at the outset, and as stated in the preface, that the book is not intended as a compendium of research published elsewhere alone, but the authors have declared or upheld their own personal positions on many topics.

There is a paucity of subheadings. Topics often run into pages without a sub heading e.g.: Environmental factors

in MS: Infections- run into 5 pages and finding the contribution of herpes virus or chlamydia, without reading a substantial portion can be daunting. Though the interested reader, searching desperately for a particular fact will plough through, the casual reader may be intimidated and may opt to skip.

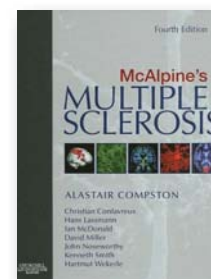
The references for all the sections are placed conveniently at the end of the book which is very good. However even when arranged alphabetically and chronologically, each author has more than a handful (e.g. Kurtzke-47) and finding the right one can take more than a few minutes. A numbered approach may have been easier for the reader.

The chapter on differential diagnosis is comprehensive. But it was surprising to not to find Neuromyelitis optica there, though it has been mentioned elsewhere in the section on symptoms and signs. I would think that the distinction between MS and NMO is now sufficiently clear to consider it a different disease.

In the editors words "The aim of the book is to summarise everything of importance relating to MS from the time the disorder was first recognised to the mid 2005 and to make this synthesis useful for the interested lay person and the fully informed professional".

The book succeeds brilliantly in this. It is a 'must have' for any physician with an interest in MS and any library that boasts a section on neurology.

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