Prion Diseases of Humans and Animals

Ed S B Prusiner, J Collinge, J Powell, B Anderton
Ellis Horwood, £80, pp 583
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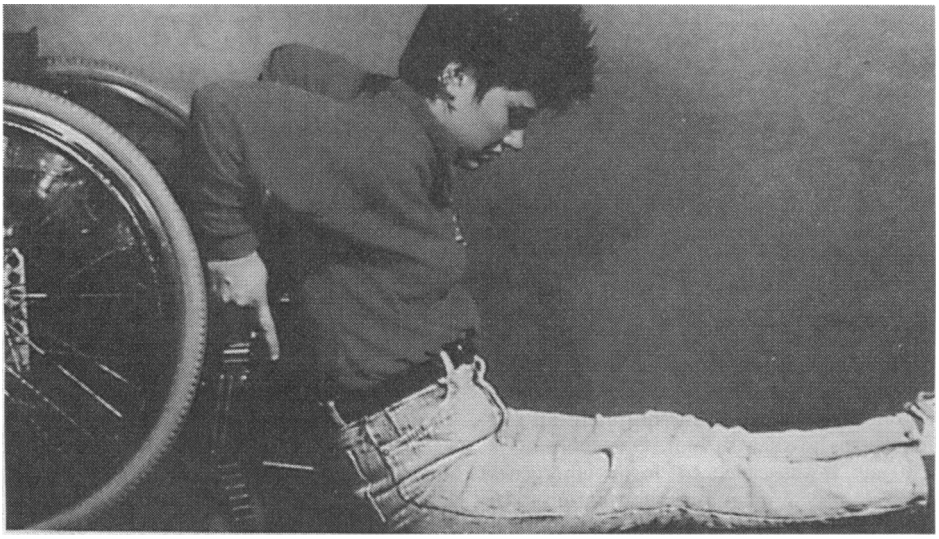
Since the pioneering studies of Carlton Gajdusek and his colleagues in the 1960s showing intracranial transmission of the spongiform encephalopathies kuru and Creutzfeldt-Jakob disease to chimpanzees, this group of diseases, which includes scrapie of sheep and transmissible mink encephalopathy, has continued to be the focus of much clinical and scientific interest. This interest has recently been intensified by the recognition of bovine spongiform encephalopathy (BSE) of cattle (mad cow disease). The arrival of this large volume on prion diseases of humans and animals is therefore particularly timely and topical. I was at first a little taken aback at the list of no less than 136 authors (which reads like a *Who's Who in Prions*) and 45 chapters, but despite the inevitable variability of style and content in such a multiauthored work, the various reviews show a uniformally high standard of presentation.

Just about every aspect of the subject is covered, including the epidemiology of BSE, molecular genetics of human familial prion diseases, the cell biology and biochemistry of prions in animals and humans, and the more recent transgenic models of prion diseases. It is clear that a unique feature of prion diseases is that they are both genetic and infectious. There is also a fascinating series of historical chapters starting with the scrapie story and including some original correspondence between W J Hadlow and Carlton Gajdusek in 1959 in which the remarkable similarities between scrapie and kuru were first appreciated. There are also reflective chapters by other pioneers in the field including Clarence Gibbs, Michael Alpers, Elisabeth Beck, and P Daniel. I found this section highly entertaining and it will certainly appeal to those readers with an interest in scientific discovery and the development of scientific ideas.

Although it is not always easy reading, the book will certainly enable the non-specialist clinician or scientist to gain a clear understanding of the subject, one which is rapidly advancing thanks to the application of the most recent molecular genetic techniques. For instance, it is explained how the term "prion" came into general usage. First proposed by Prusiner in 1982, a prion is defined as "a small proteinaceous infectious particle that resists inactivation by procedures which modify nucleic acids." Although this whole subject has been steeped in controversy for many years, it is now clear that the infectivity of scrapie depends on an abnormal form of the "prion protein" (PrP). In prion diseases such as Creutzfeldt-Jakob disease, Gerstmann-Straussler-Scheinker syndrome of humans, scrapie of sheep, and BSE, the transmissible "prion particle" is an abnormal isoform of the PrP (designated PrP^{sc}) which seems to be a biochemically modified version of the cellular protein PrP present in normal brain (designated PrP^{c}). Although it is clear that they are different from viruses, it is not known whether prions are composed entirely of PrP^{sc} molecules or contain an additional component, and much current effort is directed towards unravelling the methods of conversion of PrP^{c} to the infectious PrP^{sc}.

As explained in the excellent final overview by Collinge and Prusiner, a key question is whether this conversion process will have more widespread implications for normal cells or whether its significance is confined to neurodegenerative diseases. No doubt time will tell. Meanwhile, the editors have done a really splendid job in persuading such a galaxy of talented individuals to translate their oral presentations at a London meeting into a cohesive series of concise chapters. I thoroughly recommend this book and will personally find it very useful as an information source. Indeed, perhaps its greatest asset is the fact that it is so comprehensive.—P G E KENNEDY, *professor of neurology, University of Glasgow*



Task-specific training to enable a paraplegic woman progressively to increase her lifting capacity, in *Key Issues in Musculoskeletal Physiotherapy* (Butterworth-Heinemann, £19.95, ISBN 0-7506-0177-9).

Clinical Neurology: A Modern Approach

A Hopkins
Oxford University Press, £50, pp 486
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"Neurology should not be separated from the main stream of general medicine," writes Dr Hopkins. No neurologist should argue with this. However, he takes it much further and suggests that in so far as neurology should remain within the mainstream of medicine so should its exponent, the neurologist, who should be responsible for the care of acute emergencies in a medical rotation. To do otherwise, he advises, is to suffer "an invidious distinction of standing apart." He would evidently have the neurologist responsible for the care of emergencies; perhaps diabetic ketoacidosis, to instance but one condition well outside the experience and expertise of today's trained specialist.

Clinical Neurology, reflecting the author's view that most neurological textbooks overemphasise the abstruse and rare disorders, gives more space to a discussion of the commonplace. In assessing the place of expensive neuroradiological and neurophysiological investigation in the modern economic setting of the health service, Dr Hopkins rightly declares that the neurologist is responsible for deciding which symptoms warrant the use of expensive imaging techniques and shows how careful clinical judgment can play an important role in medical economics.

The book is eloquent, error free, and well illustrated. The author does not single out a specific readership, but I think this has to be a student textbook, for the general nature of the layout and discussion of the various nosological entities are aimed at that level. As a general view of clinical neurology, it will allay the fears of many students that the understanding of complex anatomy and neurophysiology is essential to the proper understanding of the clinical subject. However, it will have to be explained to these students that beyond the final examination and the practice of neurology in general medicine there lies a detailed and fascinating clinical specialty, the basic grammar of which