
Considerable advances have been made in our knowledge of the chemistry of gangliosides during the past ten years, but these have not been matched by a concomitant understanding of their functional role. Distribution of gangliosides is known to be wide throughout most tissues in the body but the high concentrations shown to be present in nerve cells have suggested to many observers that they are probably of functional importance in the nervous system.

This book constitutes the proceedings of an International Symposium held at Columbia University in 1980 and represents an attempt to gain insight into such biological function. The main areas covered are development and ageing of the brain, modification of enzyme activity, seizure control, cell growth in tissue culture and regeneration of damaged nerve tissue. There is a brief but useful introductory review chapter on the basic aspects of ganglioside chemistry, but the later chapters, most of which are also short, present original experimental data, with some attempt at speculation in terms of function. Inevitably, however, the volume consists of a series of discrete articles which will be of interest to scientists wishing to enter the specialised fields of ganglioside neurobiology. There is therefore no overall critical assessment of the present state of knowledge as a whole and the orientation of the book is not primarily clinical.

The volume is well produced, the diagrams and figures clear and the photomicrographs of a high standard. The bibliography is very adequate and contains an occasional reference even as late as 1981. This book will be of interest mainly to neurochemists, neurobiologists and experimental neurologists.

PT LASCELLES


Those interested in Huntington's disease will welcome this excellent monograph, which has evolved out of the author's original study of the illness in South Africa. Hitherto we have relied heavily on the chapter on Huntington's disease by GW Bruyn in the Handbook of Clinical Neurology, volume 6 (Bruyn writes the foreword to Hayden's monograph), and on the two volumes in the Raven Press series Advances in Neurology (volumes 1 and 23) which reported the proceedings of the Centennial Symposium in Columbus, Ohio, in 1972, and of the later symposium in San Diego in 1978. Hayden's text will take its place alongside these other publications as amongst the essential modern reading and reference works on Huntington's disease.

A brief historical introduction takes the reader from the dancing manias of the middle ages to modern concepts of the illness, which include the tendency, particularly in North America, to replace Huntington's chorea with Huntington's disease. Such a change of emphasis shows that chorea is neither the crucial hallmark of the illness (many patients present with the akinetic-rigid variant of Westphal), nor the most disabling feature of the illness (the personality and mental changes are the overriding deficits in most individuals). Subsequent chapters on Genealogy and Geographical Distribution, Epidemiology, Natural History, and Genetics are particularly strong, reflecting the author's background in clinical genetics. The origins of the defective gene responsible for the disease, and its world-wide distribution are discussed in detail, all available literature up to about 1979 being reviewed. It is particularly nice to see the extensive, but unpublished studies of David Stevens on the epidemiology of the illness in Yorkshire receive due recognition, along with the more recent investigations of Harper and his colleagues in South Wales. In each of these chapters, the available original data is tabulated for ease of reference, and this material alone will prove of inestimable value to research workers. There seems to be general agreement that the prevalece of Huntington's disease is of the order of 30 to 70 per million of the population in those countries whose origins lie in Europe. No obvious areas of increased prevalence, that cannot be explained by the "founder effect" in small populations or by enthusiastic ascertainment, emerge. However, the illness undoubtedly is uncommon amongst the Japanese and the American and African Blacks. "All genealogical studies indicate that emigration from north-western Europe was primarily responsible for the spread of the gene for Huntington's chorea around the world. A diminished frequency of the disorder would then be expected in those races who have their origins outside Europe." A disturbing feature is the apparent increase in the frequency of the illness in the United Kingdom, as judged from mortality statistics. It is to be hoped that genetic counselling will reverse any such trend, provided that the incidence of spontaneous mutations remains negligible. Stevens, for example, has calculated the spontaneous mutation rate, expressed as the number of mutations per million gametes, to be as low as 0-7. This compares with figures of about 10 for tuberous sclerosis and up to 100 for neurofibromatosis. In fact, the number of mutations in which parentage has been examined closely is remarkably small, and some would go so far as to say that no convincing example of such a mutation exists.

Another interesting aspect of the genetics of the disease, namely the predominance of paternal descent in juvenile Huntington's disease receives appropriate attention, as does the tendency for familial aggregation of juvenile patients. As is to be expected, the section on practical genetic counselling is handled well, and the principles of conditional probabilities of developing the disease at different ages are discussed in an Appendix. A graph of the residual probability for developing the disease at different ages might have been of practical help here.

The sections devoted to clinical features, diagnosis, and neuropathology are somewhat briefer. The description of the neurological deficits of the disease is adequate, but neuropsychiatrists would view the section on the mental sequelae as a short summary of a large and complex subject. These mental problems pose the major challenge to the management of the individual with Huntington's disease, and usually represent the greatest strain upon the family of the sufferer. Diagnostic techniques, such as they are, are mentioned, but pathophysiology is not discussed at all. The section on differential diagnosis is helpful, but few of us would have the confidence to say that "tardive dyskinesia is a unique entity with specific motor abnormalities." The differentiation of chronic psychiatric disease with tardive dyskinesias from Huntington's disease is a common and very difficult problem, for patients with the former nearly always can give no adequate family history.

The two chapters on management of patients with Huntington's disease and living with the illness are carefully constructed and valuable. The author obviously has had great personal contact with affected individuals and their families, so
speaks with sense and sensitivity about the problems they face. Current drug therapy is reviewed, with the conclusion that, despite the many studies undertaken of alternative treatments, dopamine antagonists such as perphenazine or tetrabenazine remain the drugs of choice for controlling chorea. Details of the nursing approach to the problems of the illness are presented and will prove valuable. Psychological defence mechanisms for coping with the illness are discussed, and the stresses of “being at risk” are realistically presented. The author takes the view that the role of the clinician is to present the facts, but does not take a stance on the vexing problem of the role of predictive testing, if and when such a test becomes available. No predictive test exists at present, but much research is being undertaken to produce one. Should it become available before an effective treatment for the disease is discovered, agonising ethical dilemmas will ensue. The author presents these potential problems, but perhaps sensibly refrains from passing any judgement on their solution.

The final chapter is entitled “Current Trends in Research”. The location of the abnormal gene for Huntington’s disease has not been established and the altered gene product has not been identified. The studies claiming abnormal membrane structure and fibroblast behaviour are reviewed, but the general consensus at present is that definite proof that these changes are specific for Huntington’s disease is lacking. The extensive biochemical changes in the brains of those dying of Huntington’s disease discovered in recent years are presented in detail, but most if not all must represent the consequence of neuronal loss—the “smoke rather than the fire”. But this general ignorance is not peculiar to Huntington’s disease. “Of the 2811 described genetic diseases, a total of 1489 disorders are inherited as autosomal dominant traits (McKusick 1978). In contrast to the tremendous advances in the understanding of autosomal recessive diseases, there is a dearth of knowledge concerning the basic defects in autosomal dominant conditions. Elucidation of the primary defect in Huntington’s chorea would provide a conceptual framework for the investigation of other similarly inherited disorders.” Dr Hayden has done a great service to all those interested in this disease by providing an invaluable monograph to turn to as a source of reference to all aspects of Huntington’s disease.


The trouble in writing any textbook is knowing when to stop. As a general rule, the longer the book becomes, the less use it is to medical students and the more use it is to doctors as a source of reference. This book is positively encyclopaedic, and combines all the usual advantages and disadvantages of such standard texts. It is not an attractively produced book: apart from being large and physically very heavy (which makes it difficult to read on the bus), the diagrams are rather poor and in some chapters a little thin on the ground. In fact, in a book which provides as much detail as this, there are far too many diagrams without scales or axes indicating, for example, the duration of action potentials or the size of sense organs in the ear, eye or skin. Much more use could have been made of published diagrams and data from recent papers.

It is not a book to be recommended for teaching purposes, because although there is a wealth of detail in all the chapters, there is little or no attempt to point out fundamental concepts. You would not, therefore be able to understand the working of the inner ear by reading page 316, nor the columnar organisation of the visual cortex by reading page 331. But on the other hand, you would certainly be able to refresh your memory about how cortical orientation columns may be demonstrated by the 2-deoxyglucose method. This is really a book for reference only, and it has a useful index to help in this. There are perhaps a little too much pure anatomy in some places, which readers will find better described in other books, and there is certainly too little emphasis placed on neuropharmacology—which usually is relegated to a few paragraphs at the end of a chapter or section. Nevertheless, despite its faults, I have found this to be a useful book which I am glad to have on my shelves.

**J ROTHWELL**


This book gives an account of electrical methods of treatment and diagnosis for physical therapists. It introduces the principles governing the application of electric currents in patients and reviews the areas in which success seems to have been achieved. It also gives an outline of the principles of electromyography and evoked potential recordings for therapists who might be thinking of adding diagnostic procedures to their repertoire. It is written by (and for) a group of people who are not encouraged to exist in the British Health Service, namely therapists who are not medically qualified but have an interest in basic science and a reasonably sophisticated understanding of clinical neurophysiology. On the British scene it therefore falls between two stools, the level of discussion being too simple for clinical neurophysiologists, while most of the techniques are regarded over here as a medical prerogative.

It provides a useful, if somewhat uncritical, source of references to the application of electrical treatment to promote bone growth, control infection and strengthen skeletal muscles. Lucinda Baker’s chapter stands out as a thoughtful, intelligent and critical account of the value of neuromuscular electrical stimulation in the restoration of purposeful limb movements. Two chapters on transcutaneous electrical stimulation give a useful summary of its possible mechanisms and practical applications, emphasising the shaky experimental basis of most present clinical practices. Stimulation of the spinal cord and brain is not discussed.

“Electrotherapy” provides an optimistic account of electrical stimulation with enough basic information to whet the appetite of therapists or physicians interested in finding out more. I would not recommend it for students because it is too cavalier in its treatment of basic science and there are some errors (such as the diagram that purports to show a sine wave) in this department. In Britain, I suspect that it will be read mainly by private therapists looking to extend their range of gadgetry but others who, like the authors, are concerned to strengthen the clinical bias of electrical treatments will find this a useful short review.

**DL MCELLENN**


The book under review—the third volume of the series “Persistent Pain: Modern Methods of Treatment”—is edited by Sampson Lipton and John Miles of the
Huntington's Chorea

CD Marsden

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