Gangliosides in Neurological and Neuromuscular Function, Development, and Repair. Edited by Maurice M Rapport and Alfredo Gorio. (Pp 296; \$44.88.) New York: Raven Press, 1982.

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 are 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

Huntington's Chorea. By Michael R Hayden. (Pp 192; DM98, US \$45.60.) Berlin: Springer Verlag, 1981.

Those interested in Huntington's disease will welcome this excellent monograph, which has evolved out of the author's original study of the disease 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 emphasises 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 disease (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 prevalence 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 mutants 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 carry 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,