
Clinically, and as a problem in biology, the neurofibromatoses represent something of an adventure playground for clinical scientists. The impact which these diseases make on the lives of affected individuals and their families calls for all the practical and pastoral skills on which clinical medicine is based.

Several candidates, other than von Recklinghausen, claim priority for description of the NFs. The most recent is the unknown sculptor of a Hellenistic votive offering, catalogued in 1921, but not seen since the second world war. Much has since happened. Molecular geneticists know about the neurofibromin locus on 17q11-2 and the fragment of 22p12-1 which encodes the NF2 gene and cell biologists have shown that defects in tumour suppressor genes lead to uncontrolled Schwann cell proliferation. The differential diagnostician can sharpen up on the eponymously challenging syndromes of McCune-Albright, Deumer, Jaffe-Riley-Roberts-Kipple-Trenanuary-Weber, Maffucci, Bannayan-Zonana, Jaffe-Campanacci and Arskog and for the unwary, the spots of NF1 may turn out to be a LEOPARD. Even if the frequency of tumours affecting the central or peripheral nervous system in the NFs has been exaggerated, their diversity has not and for the neuropathologist there are tumours galore—dermal, nodular and plexiform neurofibromas; neurofibrosarcomas, Schwannomas, cafe au lait macules, Lisch nodules, astrocytomas, meningiomas and hamartomas. On the topic of macrocephaly, heads may swell even more at the news that Lock's of London now stock larger hat sizes than in the days when Admiral Lord Nelson had his 57 cms cranium hatted out for the battle of Trafalgar. The most celebrated person not to have neurofibromatosis was Joseph Merrick. The editors have here arranged an intellectual exhumation of his remains, and preference is given to the diagnosis of Proteus syndrome, named after the old man of the (Aegean) sea who was given the power to change his shape at will. Sadly, those bits of Merrick which might have allowed modern molecular genetics to settle the issue of diagnosis were destroyed after dry rot infested the pathology museum at the London Hospital in the 1940s.

Although The Neurofibromatoses. A pathogenetic and clinical overview is the product of 25 pens, the editors have produced a text which maintains an easy narrative; Susan Huson and Richard Hughes bring to their subject a wealth of knowledge, common sense and infectious enthusiasm, drawing on considerable experience of the neurofibromatoses in all their clinical, social and psychological manifestations. The right topics are selected and the quality of reproduction for radiographs, clinical photographs and the few colour illustrations is uniformly (and unusually) high. In The Neurofibromatoses. A pathogenetic and clinical overview, Susan Huson and Richard Hughes have presented a scholarly and coherent account of one of medicine's most fascinating problems; it is an important book.

ALASTAIR COMPSTON

The Central Nervous System in AIDS: Neurology, Radiology, Pathology, Ophthalmology. Edited by J ARTIGAS, G GROSSE, F NIEDORFER. Published by Springer-Verlag, Heidelberg. Pp 237 Illustrated DM283.00 sFr 280,00 ISBN 3-540-55839-X.

Many practising neurologists in the United Kingdom still have little exposure to AIDS in their clinical practice. However, this subject has become a major topic at most International Neurological Conferences. AIDS must be considered in the differential diagnosis of disease at all levels of the neuroaxis and may be considered as the modern "giant mimic". This compact book in five chapters and 236 pages covers clinical pathology of AIDS, diagnostic imaging, neuropathology and clinical and pathological ocular features. The three editors of this volume, from the Auguste-Viktoria-Krankenhause, produced this intensive third chapter on the neuropathology.

As a clinical neurologist I was interested in the initial chapter of fifteen pages which covers the statistics of neurological involvement in the disease. Although short, it is well laid out with useful tables, which will make helpful teaching slides. The therapy of the complications of AIDS covered and the chapter is supported by more than one hundred and fifty references.

The second chapter involving the diagnostic imaging of intracranial manifestation of AIDS is clearly laid out. It considers radiological features of all the manifestation of AIDS and its complication. It is well illustrated by good figures, some showing serial changes in the evolution of AIDS in the nervous system. This chapter is also supported by in excess of four hundred and sixty references.

The centre of the book is dominated by an extensive chapter on the neuropathology of AIDS, which is covered in one hundred and seven pages. The text is punctuated by very useful tables. The introduction covers practical problems of collecting specimens from AIDS autopsies. The introduction and, indeed the chapter as a whole, emphasizes one of the opening statements that "the neuropathological finding in AIDS autopsies reveal a very broad spectrum of findings". These refer to the classical neuropathological features of disease, as well as the special AIDS-associated changes. The chapter takes us through HIV, encephalitis and leukoencephalopathy, the opportunistic infections and the CNS tumours. Again the chapter is well supported by more than six hundred references. The final two chapters cover the clinical ophthalmology of AIDS. This is enhanced by good illustration. I think it would be helpful if the retinal pictures could be in colour, though I suspect that this will add excessively to the cost of the volume.

The book is consistent in its style, well laid out, provides clear script and a good bibliography. It lies between a book to read and a reference book. It is clear from the literature that the bibliography on AIDS is changing rapidly; however, this volume presents a comprehensive selection of references up to 1993. The production of the volume is good quality. I think many may find the price of 283 DM somewhat excessive, but I would recommend it.

LESLIE FINDLEY


Paediatric epileptologists have a difficult time. About 75% of patients with epilepsy start to have seizures when they are between one and six years of age. Seizures are more common in the first month of life than at any other time. In children seizure types change with age and so do the underlying causes for the seizures.

The classification of seizures and epilepsy syndromes changes with the years and it now seems increasingly important to identify the syndromes accurately—some of them (eg, benign familial neonatal convulsions) may be the result of a single gene defect. Life becomes more complicated.

Perhaps adult epileptologists have a more difficult time. They may have to deal with the medical, psychological and social consequences of seizure disorders and syndromes that have started in childhood. Are paediatricians inventing these complicated stories to keep themselves occupied? The best way to find out is to read a good book on the subject. Aicardi's book may be the best one to get. Professor Jean Aicardi is the ideal man to write a text book about epilepsy. He has been a clinical epileptologist for 28 years and has a wide knowledge of the literature, he is intellectually rigorous and he produces better written English than most of us. His book is a good length—about 530 pages—and it is packed with information. The text is lucid despite its density.

The book is divided into 4 parts. The first is general and deals with definitions and the classification of epileptic seizures and syndromes. The second describes the major types of epileptic seizure in childhood and the corresponding epileptic syndromes; this takes the reader from infantile spasms to the Landau-Kleffner syndrome via the Lennox-Gastaut syndrome and others. The third deals with clinically important seizure problems in children—neonatal seizures, febrile convulsions, status epilepticus etc. Finally there is a discussion of aspects of diagnosis, prognosis and treatment. Each chapter is a distillation of much research and clinical experience and the author provides both clinical advice, historical advice. The references occupy 100 pages at the end of the book and there are 2552 in all. References relating to a particular subject have to be winkled out of this list with some effort.

DAVID HARDY

Book reviews


This is a valuable book for neurosurgical trainees and for those engaged in the care of patients with brain tumours. It is well produced and is replete with clinical, histological and radiological data. It contains numerous tables and diagrams and at the end of each chapter are a number of useful tables.