The possibility that von Willebrand's disease is a monogenic or connective tissue disorder enforces the plea by Dr. Osenbach and his colleagues that structural vascular lesions should be ruled out in all patients with von Willebrand's disease who develop intracerebral haemorrhage upon minor trauma.

Osenbach et al's patient had successful surgical extirpation of the lesion after two weeks of medical therapy. Administration of the synthetic vasopressin analogue 1-desamino [8-D-arginine] vasopressin (DDAVP) may have been considered in their patient with type I von Willebrand's disease. This type of von Willebrand's disease is characterised by decreased plasma levels of qualitatively normal von Willebrand factor: antigen (vWF:ag). vWF:ag strongly promotes platelet-vessel wall interaction. DDAVP has been shown to stimulate the release of factor VIII and vWF:ag, shorten or completely normalise bleeding time, and provide surgical haemostasis in patients with von Willebrand's disease and other haemostatic disorders.12,13 The drug is administered at doses of 0.3-0.4 μg/kg body weight by intravenous infusion over 20 minutes. DDAVP was approved by the United States Food and Drug Administration in 1984 for the treatment of the haemostatic defect of von Willebrand's disease. After an adequate response to the drug has been shown before surgery,10 DDAVP is recommended for early prophylactic therapy for patients with type I von Willebrand's disease undergoing surgery. Endogenous vWF:ag released by DDAVP into plasma has been shown to be haemostatically more effective than exogenous vWF infused with plasma concentrates, allowing safe performance of surgical procedures.14 Moreover, prolonged bleeding time in patients with severe von Willebrand's disease can be further shortened by DDAVP administration.14

Treatment with DDAVP avoids the risks associated with the administration of plasma-derived products, for example, viral transmission and allergic reactions. DDAVP administration is associated with very few adverse effects.14 Mild facial flushing, probably caused by dilation of the skin, is most frequently encountered. Other less common side effects are mild and transient headaches, a 10% increase in heart rate, and minor decreases in blood pressure. These reactions can easily be attenuated by slowing the rate of DDAVP infusion. DDAVP administration can be repeated at intervals of 12 to 24 hours although some patients treated with this drug at closely spaced intervals may become progressively unresponsive over a period of approximately five days.

WOUTER J SCHIEVIK, MARCEL M LELY Department of Neurology and Centre for Thrombosis, Haemostasis, and Atherosclerosis Research. The Academic Medical Centre, 1105 AZ Amsterdam, The Netherlands

BOOK REVIEWS


We doctors love names, and the more confusing and meaningless the better. In the past we could keep patients, lay people, junior staff and even the faculty puzzled for what now names have taken on a totally new life. Speaking as a Consultant Neurological Pathophysiologist with neuroanatomical undertones and an interest in neurological rehabilitation and a working day of neuro-epileptology, neuro-psychotherapy, neuro-genetics, neuro-dyna and neuro-everything else, I wish this excellent book—which was originally called (or, as the editors changed it, “published by the appellation”) “Neurological Pathophysiology” and is now called “Neurobiology of Disease” and has first class introductions of normal functioning systems—was called “Neurology” (ie that branch of science and medicine which deals with the nervous system, both normal and in disease), or is this wish in early manifestations of somatodemenoria a loss of neuro-adaptive behaviour, and neuro-intelligence?

The Editors of this book have brought together 28 eminent contributors who have, together with the 2 editors, brought us one of the most readable and worthwhile introductions to neurology on sale today. The book, in 2 sections, dealing with functional and anatomical systems, and disease processes, is successful in its purpose of providing both an introduction to “the scientific basis of neurology” for medical students and “the expression of fundamental mechanisms for neuroscientists in neurobiology, (ie it provides an introduction—an excellent introduction—to the study of neurology). The first part, entitled “Functional and Anatomical Systems” consists of 12 chapters dealing with normal and demyelinated axons, peripheral nerve, neuromuscular junction, muscle, the somato-sensory system and pain, the auditory system, the visual system, eye movements and vestibular system, and “demyelinating diseases”. The second part, “Disease Processes” deals with the genetic disorders (almost entirely about Huntington's disease), seizures and epilepsy, and dementia, demyelinating disease, stroke, metabolic encephalopathy, cerebrospinal fluid, blood brain barrier, and brain oedema, brain tumours, infections and Parkinsonism.

It is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory. For example, muscle is dealt with in the first section on “function and neuroanatomical systems” and “disease processes”, is not entirely satisfactory.
One of the many excellent features of this book is the incorporation of "case classics". These arereeze membranes throughout the appropriate sections of the book, and include such superb accounts as Cerebellar Injury described by Gordon Holmes in 1917, Alzheimer's own description of his disease, the first description of lumbar puncture, Huntington's (1872) description of his chorea etc etc. All these are followed by an explanatory comment. This is a most interesting way of adding depth to a subject and it also adds a nice touch of intellectual tone.

The recent developments in molecular biology, particularly as they relate to neuromuscular conditions, are well covered. This (as with many) is reasonably priced, the illustrations are clear, and I thoroughly recommend this book for both undergraduate and graduates. I think, however, that it would be immensely improved by not attempting the artificial separation mentioned above.

L S ILLIS


Books such as this are produced to help keep up to date the overworked clinical neurologist who is failing to keep abreast of the modern neurological literature and current trends in the field of neuroscience. Clearly only a limited number of topics can be covered and in preparing this review I have specifically looked to see if the topics are appropriate and if they have been adequately covered with up to date information.

The selection of topics is bound to be personal and I think that Chris Kennard has chosen well. Two chapters that immediately caught my attention were those on Neural Transplantation and Myalgic Encephalomyelitis. These are areas that are receiving widespread publicity in the popular press and we neurologists need up to data information to be able to answer our patients' questions. The authors in addition provide a balanced view of the available clinical information and I now feel much better armed to answer questions from patients with Parkinson's disease and other degenerative disorders regarding "Brain transplants". I particularly appreciate the conclusion which reads "there exists at present no intracerebral transplantation therapy for any neurodegenerative disorder".

Another chapter that tackles a difficult issue is that on the Chronic Fatigue Syndrome which is described as "viral fatigue". The introduction refers to the Myalgic Encephalomyelitis Association as being Britain's fastest growing charity and hints at the difficulties created by the Association for clinical neurologists having to deal with patients who think that they might have the disorder. The chapter carries the authority of the much respected neurological scientist, Professor P K Thomas, and the conclusions therefore are important and hopefully will receive widespread support. The most important of the conclusions are that the chronic fatigue syndrome is a heterogeneous disease and a variety of neurological disorders may be misdiagnosed as the chronic fatigue syndrome, that no relationship has been found between laboratory findings and clinical status, and that the most obvious risk factor for the disorder is previous psychiatric illness.

A further important chapter on Cerebral Gliomas is written by three contributors from the Neuro-oncology Section from the Institute of Neurology. As might be expected, it tends to emphasise the more aggressive approach adopted by such centres, though overall it is rather more realistic than I expected. It certainly states clearly that cure is rare and aggressive treatment at best prolongs survival. Management of cerebral gliomas is one of the rather more controversial areas in neurology and this review is up to date and provides useful information regarding the options for modern treatment.

The review on New Therapeutic Horizons in Epilepsy is not limited to therapy but gives information on mechanisms of seizures, new thoughts concerning old drugs, and information on the various new drugs that are at different stages of therapeutic trials. In the chapter on a Systematic Approach to Vertigo I was delighted to see that the authors understand how sphenoid spongolysis are used for dizziness induced by head movement in old people is most commonly a vestibular abnormality and due to cupulolithiasis of the posterior semi-circular canal. The other reviews on Neurofibrromatosis, Spinocerebellar Mechanisms and Neurological Rehabilitation are of a similar high standard.

All the chapters are well referenced and up to date. Having read them all I feel that I have learnt a lot and sufficient of the information will help me in my clinical practice to justify a recommendation that all my colleagues should do to the same. I warmly recommend this volume and look forward to next years' edition.

NEF CARTLIDGE

Surgery of the Cranial Base. By OSSAMA AL-MOFTY. (Pp 356; Price: £375.00; US$150.00; UK£ 93.00.) Kluwer Academic Publishers Group, 1989.

At a time of multiple author productions, it is refreshing to have the views and experience of one man, one surgeon, on the variety of pathology affecting the skull base from the orbit down to the foramen magnum. There is thus an continuity and interlinking of his ideas of surgery and management and this more makes for some lack of depth which might be provided by a chapter written by a world authority confining himself to one particular topic, such as the management of cavernous sinus. However, the author is a extremely energetic neurosurgeon from the University of Mississippi who is rapidly gaining international respect for his presentations on skull base surgery, and this book represents a review of his expertise in these most difficult tumours.

The book is divided into seven sections:

i) Instrumentation; ii) The Sella and Parasellar Areas; iii) The Orbit; iv) Craniofacial Surgery; v) The Posterior Cranial Base; vi) Surgical Reconstruction and Adjuncts to Cranial Base Surgery, in which anaesthesia, electrophysiological monitoring and vii) Instrumentation and Reconstruction of the skull base. The illustrations are generally of an extremely high quality and the CT scans and MRI images beautifully exemplify the pathology. The colour plates too are excellent and are very necessary additions to this type of book.

The black and white operative pictures with artistic "overlining" of various structures perhaps are not as successful as the author had hoped. It is notoriously difficult to demonstrate the relationship of one small vessel or simple line diagram and my own opinion would be that many of the operative photographs could have been deleted and replaced by simple line diagrams. Those who operate on the area will understand immediately the finer points, those who do not operate in the area will not be confused by the excess detail seen in black and white photographs.

The author has performed an extensive literature search which, at the time of going to press, is arguably the best rehearsed skull base review. For that, he is to be congratulated; this is an expertly written book and has been well performed. The weakest sections in the book are the first on Ergonomics and Power Equipment. One is not quite sure what is being prescribed. The other sections are clearly directed at those who have already have surgical experience, for few would tackle the lesions that he has demonstrated without previous experience. That being so, the comments on operating microscope, ultrasonic aspirators and bayonetted instrumentation would be superfluous. The deletion of this section, or a scientific review of the advantages and disadvantages of laser versus ultrasonic line diagrams might be considered in a second edition.

Taken as a whole, the book is a useful addition to the library of the skull base surgeon from whatever discipline.

ALAN CROCKARD


That neuropsychology is an expanding subject is demonstrated by the number of new books and journals devoted to it over the last decade and by the frequent need to update the former. This book is intended to replace one from 1983, which is barely two student generations ago, and most of the text collected are from the 1980s, which witnesses the wealth of publication that has occurred recently. For neuropsychologists and anyone acquainted with the area it provides a useful summary of recent work, with some thought-provoking discussion of current ideas concerning both hemispheric specialization and other neuropsychological issues.

The book covers most of the usual topics associated with hemisphericity such as language laterality, handedness, sex differences and "cognitive style", as well as straying into various peripheral areas and even into topics only distantly related to brain mechanisms, such as which way protons spin and whether
BOOK REVIEWS: Neurobiology of Disease.

LS Illis

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