Matters arising


Hughes et al reply:
We thank Professor Rajput for his interest and contribution to the discussion of our study.1 It is unclear why progressive supranuclear palsy (PSNP) comprised such a high percentage of patients clinically misdiagnosed as having Parkinson's disease (PD) in our study, while in his series all necropy proven cases of PSNP were recognised before death.2 A pure akineti-syndrome does not seem to be the common manifestation of PSNP.3 The UK Parkinson's Disease Society Brain Bank (PDSSB) receives donor tissue from Parkinsonian patients throughout the UK. Only those enrolled in the scheme, patients are examined annually by one of 70 neurologists and geriatricians associated with the Brain Bank and information is recorded according to a standard format. Despite the use of diagnostic criteria it is clearly impossible to completely standardise diagnostic practice across such a group of assessors. The stage of disease when patients are examined is clearly important in studies of this type. The clinical diagnoses used in our present study were all made within 12 months of death, at the time of the last assessment, and during or after 1986. All patients were considered specifically to have PD rather than a less well-defined Parkinsonian syndrome.

We agree that no diagnostic criteria for PD are fool-proof and have subsequently analysed the clinical features of our cases in terms of their diagnostic value.4 By using selected criteria (asymmetrical onset, no atypical features, and no possible aetiology for another Parkinsonian syndrome) the proportion of PD cases identified was increased to 93%, but at the expense of excluding 32% of pathologically confirmed cases. Twelve of 100 cases of histologically confirmed PD cases examined at the PDSSB had atypical clinical features according to Brain Bank diagnostic criteria for this disease,5 More than half of these cases had no other associated neuropathological findings that could account for the atypical features.

These findings suggest that studies based on consultant diagnosis of Parkinson's disease will include patients without the disease as well as excluding some who subsequently satisfy the histological criteria for the disease, thus results from clinical trials and epidemiological studies may be distorted.


BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom. For overseas customers the following is recommended: for the family, for future patients and families. Storage of DNA will provide a valuable resource for new and young PD patients. British Neurologists. A Clinico-pathological study of 100 cases of Parkinson's Disease. Arch Neurol 1993;50:140-8.


At the present time there is some fascinating progress in the definition and territorial demarcation of neurodegenerations causing dementia and for splitters, among clinicians and pathologists to identify new diseases such as corticobasal degeneration and the causes of frontal lobe degeneration, and primary progressive aphasia. In contrast, molecular genetics is tending to lump diverse phenotypes together in the prion disorders and familial Alzheimer's disease. Real progress in other areas, notably Pick's disease, is to some extent lacking. This condition is finding a more critical definition by the effect of sorpresion, because some examples of non-Pick body Pick's disease are undoubtedly other things, such as the prionopathy. The remaining chapters in the book cover the neuropathology of unusual dementias, dementia and motor neurone disease and Lewy body dementia: so not, as you might have thought, small print causes of dementia as implied by the title, nor a fully comprehensive account of these disorders, but mostly areas of real progress and new territorial.

The introduction provides a paragraph on the clinician's approach to a patient with dementia. This is a useful summary, to which collection, storage and analysis of genetic and pathological material could be added. Many of these diagnoses remain neuropathological ones, and their genetic implications are still uncertain. In addition, peculiar phenotypes of these neurodegenerations can mimic almost any other. Future developments should justify this encouragement to obtain a postmortem diagnosis, even if not for the family, for future patients and families. Storage of DNA will provide a valuable resource for new and young PD patients. British Neurologists. A Clinico-pathological study of 100 cases of Parkinson's Disease. Arch Neurol 1993;50:140-8.


It has been the experience of most neurologists on appointment to receive instructions from solicitors to examine and give an opinion by way of report on claimants undertaking civil action for injuries sustained in domestic, social or work situations. Unfortunately most young neurologists when first approached, have had neither advice nor instruction on the preparation of reports and the implications of a medicolegal assessment. Nor have they been informed of the pitfalls when preparing reports, between writing a report and submitting to a cross examination in the high court. Often the prospect is disturbing. With the increasing civil litigation in the western world and a specific increase in claims of medical negligence, most of us will be invited to undertake assessments of claims. In some departments of neurology, a feature of the post graduate training is an introduction to this aspect of the neurologists work, but these are few.

For many years there has been a need for published advice. This is now provided by David Bell's book. Medico-legal assessment of head injury. He addresses the duties of the medical expert and the court's expectations. If in a chapter almost certainly written for the lawyers, he describes the anatomy and pathology of the head injury and the major consequences of the brain damage. In his chapter on the syndromes of regional brain injury he finds space for a useful account of the effects of extension-flexion injury to the cervical spine and quotes some important figures and references to an acceptable estimation of prognosis. He assesses the literature on the prediction of post traumatic epilepsy both in the adult and child and considers the question of iatral violence and the attribution of serious crime to epileptic activity.

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and outcome of surgery. Therefore the National Institute of Health set up a Consensus Conference on Surgery for Epilepsy. The majority of contributors are from the USA or Canada. However, most of the activity and development in this field has taken place there in recent years, especially in the use of assessment techniques such as MRI which are expensive and technologically demanding. The papers represent up-to-date views, bearing in mind that the planning began in 1989 and the majority of the papers were written in 1990.

The chapters cover all the possible topics comprehensively, the first seven are devoted to the natural history of epilepsy. The next thirteen discuss various aspects of preoperative assessment. They are all written by experts who nevertheless give broad impartial accounts of the usefulness of the techniques and compare the proper place and usefulness of various structural and functional brain imaging techniques, both between themselves and with neurophysiological and neuropsychological techniques.

There remain six chapters devoted to the diagnostic and use of various surgical procedures including callosotomy and three devoted to the outcome of surgery and methods of assessing outcome.

Since the first Palm Desert Symposium in 1987 there has been a considerable increase in the interest in the surgical treatment of epilepsy and a corresponding increase in the number of publications. This book is a very concise well-argued account of the present position and a suitable introduction. Unfortunately it is relatively expensive at $200.

CE POLKEY


This volume is a synopsis of the presentations at the 10th International Symposium on Parkinson's disease held in Japan in 1991 hence the author list has a distinctly oriental flavour. It includes around 400 contributors to 126 chapters and consequently suffers from the repetition inherent in such works. Nevertheless, it presents a comprehensive survey of advances in Parkinson's disease research in recent years.

Most sections contain one or more reviews by leading authorities in the field followed by a selection of related papers. The latter at times fall short of an otherwise high standard. However, Wichmann and DeLong's contribution on the pathophysiology of parkinsonism is particularly valuable in view of the implications regarding excitatory amino acid antagonist therapy and subthalamic nucleus. Current hypotheses concerning neurotoxins are well covered, along with their relation to monoamine oxidase, iron, superoxide radicals, and mitochondria. The recent tole face concerning the genetic component to Parkinson's disease is well reviewed by Duvoisin.

Not surprisingly for a meeting in Japan, the relationship between dystonia and parkinsonism is debated at length, especially dopa-responsive dystonia or Segawa syndrome. The work performed by the UK Parkinson's Disease Society Brain Bank continues to raise doubts regarding our clinical ability to diagnose idiopathic Parkinson's disease, with profound implications for all aspects of research. The highlights of the drug therapy section are chapters on MAO and COMT inhibitors, along with the benefits of broad budesonide, the latest fad in Parkinson's disease treatment.

The most promising area of research concerns transplantation techniques which are well reviewed, if a little out of date. The final chapter reviews work with cell cultures transfected by viruses containing the tyrosine hydroxylase gene. Such cells release levodopa both in vitro and in vivo models of parkinsonism. Extrapolated to man, homologous skin fibroblasts could be cultured then transfected with tyrosine hydroxylase before autologous striatal transplantation, thereby obviating the present problems of using heterologous mesencephalon from aborted foetuses.

This is an authoritative and well indexed source of reference. It will prove to be a valuable tool for all those engaged in Parkinson's disease research and should find its way onto the shelves of most university libraries.

CE CLARKE

**SHORT NOTICES**


This second edition of a successful comprehensive text has been largely re-written. It now includes historical aspects, neuropsychology and imaging, depression in medical settings, and maintenance treatment. Anxiety disorders are discussed in relation to depression, and mania receives less space. This is a valuable text.

**NOTICE**

European Federation of Neurological Societies. The EFNS meeting will be held in Berlin, 8–11 December 1993. Further information from P & R Kongresse GmbH, Neptun Promenade 6, D-1020 Berlin, Germany.