Progressive supranuclear palsy: neuropathologically based diagnostic clinical criteria

In their excellent retrospective clinicopathological study of 12 cases of progressive supranuclear palsy (PSP), Collins *et al*¹ noted a variety of clinical signs and symptoms beyond those in the original description of this disorder. They proposed an algorithm for the clinical diagnosis of PSP, a definite diagnosis of which had been made during life only in eight of 12 of their patients. In principle, we agree with their clinical criteria and results, based on a retrospective clinicopathological study of 24 cases of PSP from the files of the Ludwig Boltzmann Institute of Clinical Neurobiology, Vienna, and the R Escourolle Neuropathology Laboratory, Paris.² In this material, a definite diagnosis had only been made in 12 of 24 of the cases. Our diagnostic criteria were as follows: (a) onset over age 40; (b) progressive course of a non-familial disease; (c) duration less than 10 years; (d) postural instability or falls without specific aetiology; (e) akinesia and rigidity; (f) supranuclear ophthalmoplegia including down gaze abnormalities; (g) dysarthria or pseudobulbar palsy; (h) frontal lobe-like symptoms; (i) lack of focal lesions on CT; (j) no appreciable improvement with levodopa treatment. Almost all of these diagnostic criteria are identical to those used by Collins *et al*, except for retrocollis or dystonic arm, sitting "en bloc", and Babinski's signs, which were not seen in most of our patients. Based on these diagnostic items, we concluded that a clinical diagnosis of PSP was probable when nine of 10 criteria were present, whereas in the absence of two signs or symptoms, the diagnosis was considered "possible". The retrospective evaluation of these criteria allowed identification of 88% of the cases; 18 being "probable", and three "possible". These data seem of interest, as in other recent postmortem series of PSP, only a small percentage fulfilled currently accepted clinical diagnostic criteria—for example, seven of 17 (41%)—whereas the remainder who lacked these criteria had alternative clinical diagnoses.³ In another recent series⁴ the clinical diagnosis of PSP had been made in eight of 13 histologically confirmed cases (61%), whereas 13 brains (54%) showed concomitant pathological changes of Alzheimer's or Parkinson's disease. In view of the clinical heterogeneity of PSP and some difficulties in the postmortem diagnosis of typical, atypical, and combined PSP—the last featured by the presence of typical neuropathological changes of PSP together with findings that are diagnostic of other neurological disorders⁵—an evaluation of the sensitivity and specificity of clinical and neuropathological criteria of PSP in a larger series of histopathologically confirmed cases seems mandatory.

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- 1 Collins SJ, Ahlskog JE, Parisi JE, Maraganore DM. Progressive supranuclear palsy: neuropathologically based diagnostic clinical criteria. *J Neurol Neurosurg Psychiatry* 1995; 58:167–73.

- 2 Verny M, Jellinger KA, Hauw JJ, Bancher C, Litvan I, Agid Y. Diagnostic criteria for progressive supranuclear palsy (PSP): a retrospective clinicopathological study of 24 cases. *Mov Disord* 1995 (in press).
- 3 Daniel SE, De Bruin VMS, Lees AJ. The clinical and pathological spectrum of Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). A reappraisal. *Brain* 1995 (in press).
- 4 Gearing M, Olson DA, Watts RL, Mirra SS. Progressive supranuclear palsy: neuropathologic and clinical heterogeneity. *Neurology* 1994;44:1015–24.
- 5 Hauw JJ, Daniel SE, Dickson D, Horoupian DS, Jellinger K, Lantos PL, *et al*. Preliminary NINDS neuropathological criteria for Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). *Neurology* 1994;44:2015–9.

Collins *et al* reply:

We appreciate the comments of Jellinger *et al* and echo their appeal for a more comprehensive clinicopathological study of cases of PSP. It is reassuring that our findings are similar to those of other recent series quoted.^{1–3} We were also impressed by the wide range of clinical and pathological features in these cases and sought to introduce some order into the classification of PSP by restricting our analysis to cases only with typical histopathological features at post-mortem. The resulting paradigm, however imperfect, provides a useful working framework in which to place patients with clinical features suggestive of PSP, until a more specific biological marker is available.

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- 1 Verny M, Jellinger KA, Hauw JJ, Bancher C, Litvan I, Agid Y. Diagnostic criteria for progressive supranuclear palsy (PSP): a retrospective clinicopathological study of 24 cases. *Mov Disord* 1995 (in press).
- 2 Daniel SE, De Bruin VMS, Lees AJ. The clinical and pathological spectrum of Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). A reappraisal. *Brain* 1995 (in press).
- 3 Gearing M, Olson DA, Watts RL, Mirra SS. Progressive supranuclear palsy: neuropathologic and clinical heterogeneity. *Neurology* 1994;44:1015–24.

NOTICE

Announcement from the British Neuropsychiatry Association

The 1995 Summer meeting—to include joint sessions with the British Association for Psychopharmacology—will be held on 15–17 July in Cambridge

On 16 July BNPA will hold a scientific meeting with the theme of "movement disorders" and its AGM. On 17 July BNPA/BAP will have a joint session on neuroimaging, psychiatry, and psychopharmacology. Short scientific papers and single case videos by members of both associations will also be presented. For further details please contact Ms Sue Garratt, 17 Clocktower Mews, London N1 7BB, UK.

For details of membership of the BNPA, which is open to medical practitioners in psychiatry, neurology, and related clinical neurosciences, please contact Sue Garratt at the address above, or Dr Jonathan Bird, Burden Neurological Hospital, Stoke Lane, Stapleton, Bristol BS16 1QT, UK.

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 and for members of the British Forces Overseas, but overseas customers should add £2 per item for postage and packing. Payment can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.

Vascular Dementia. Etiological, Pathogenetic, Clinical and Treatment Aspects. Edited by Lars A Carlson, Carl Gerhard Gottfries and Bengt Winblad. (Pp 86 \$89.75.) Published by Karger, Basel 1994. ISBN 3-8055-5984-4.

Most of the big names in vascular dementia came together at the second symposium on Age and Ageing Disorders which took place in August 1993 in Stockholm. It was organised by the Gamla Tjanarinnor Foundation and the presentations form the 17 chapters of this book which appears as a special issue of *Dementia*.

Vladimir Hachinski opens with a marvelous chapter. Having put vascular dementia on the map in the mid-1970s he now retreats and argues that the term has outlived its usefulness. Vascular is too generic and dementia too restrictive. The traditional concept of dementia is epitomized by SDAT; a disease of old age in which progressive memory failure is associated with global cognitive impairment. Furthermore, the condition is irreversible. Vascular disease rarely, if ever, gives rise to such a condition. The cognitive deficit is multi-focal and not necessarily progressive. The patients usually have deficits in motor function which are not seen in SDAT. For Hachinski the concept of vascular dementia is redundant but the range and variety of cognitive impairment due to various vascular causes demands further description and investigation. After all, vascular disease is potentially stoppable and to some extent reversible.

Erkinjuntti describes the 1993 NINDS-AIREN criteria for vascular dementia. Here dementia was defined as a cognitive decline from a previously higher level of functioning and manifested by impairment of memory in two or more other cognitive domains. The different types of ischaemic lesions which can cause dementia include multiple large vessel stroke, single strategically

placed infarcts, lacunar infarcts in the deep grey and white matter, extensive ischaemic white matter lesions, or a combination of these. The problem is, of course, that without a brain biopsy all these patients may have coincidental Alzheimer's disease.

White matter lesions receive a lot of attention. If the clinical picture resembles Alzheimer's then their contribution to the dementia is likely to be restricted. If, on the other hand, the patient has less profound impairments in episodic memory, more depressive symptomatology and a gait disturbance, then the white matter lesions (which may be due to vascular disease) are probably pertinent to the clinical presentation.

Binswanger's disease remains a mystery. If the ischaemic demyelination is due to hypoperfusion of the white matter why haven't we got eloquent PET data to show high OEF and low CMRO2?

Cummings has written a useful chapter on the clinical characteristics of patients with presumed vascular sub-cortical dementia. Subcortical damage arises from small vessel disease which may give rise to a lacunar state (with multiple subcortical lacuna infarctions) or Binswanger's disease (see above). Subcortical dementia is distinguishable from Alzheimer's by its clinical features. These patients demonstrate slow rather than impaired cognition, forgetfulness rather than amnesia, executive dysfunction and mood and personality alteration. These alterations include loss of initiative, diminished drive, apathy, poor insight and eventually profound abulia. Furthermore they develop a characteristic frontal gait disturbance often with focal signs such as extensor plantar responses.

Haan and coworkers from Leiden provide a succinct and useful review on cerebral amyloid angiopathy. This condition can present with a broad spectrum of clinical and radiological manifestations including dementia, cerebellar and cerebral haemorrhage, subarachnoid haemorrhage and white matter disease. The controversy concerning its role in Alzheimer's disease is reviewed and referenced.

The chapters are occasionally repetitious, the subject index is rudimentary and this special issue of *Dementia* never exceeds the sum of its parts. Its parts, however, are occasionally very good and the contributions from Hachinski, Cummings and Erkinjuntti are outstanding.

J P H WADE

Stroke in Children and Young Adults.

By JOSE BILLER, KATHERINE D MATHEWS and BETSY B LOVE. (Pp 259 £55.00.) Published by Butterworth Heinemann, Oxford 1994. ISBN 0-7506-9203-0.

Young people are not expected to have strokes and the admission of a young stroke creates more diagnostic activity and therapeutic interest than the usual mesiatric and geriatric patients with strokes. Young stroke victims do not have a different list of causes for their strokes from their seniors but the same ingredients in different proportion. Clinicians are disappointed in themselves when no cause is found in 20-30% of young stroke victims, though less concerned

by the similar number of middle to old aged stroke patients in whom no cause is apparent. In young patients without a cardiac cause or atherosclerosis, physicians are rightly concerned may be lurking various rare obscure thrombophilic disorders or non-atherosclerotic vessel disease of which they feel they know rather too little.

The main purpose of this slender but densely packed book is to relieve this diagnostic uncertainty. There are lots of facts here and a mean of 122 references per chapter (range 69-268). However, there could have been more attention paid to providing the information in a more practically useful form, so that the wood can be seen from the trees. At least Katherine Mathews tries to reassure her readers in the chapter overviewing stroke in children and neonates that many items on her "laundry list" of causes of ischaemic infarction in children "can easily be eliminated in a given patient". There are a lot of laundry lists in this book, some rather superfluous such as the names of all the cerebral "sinovenous structures" in the chapter on cerebral venous thrombosis where there is a useful review of the use of anticoagulants. Some of the chapters, such as that on neonatal intracerebral haemorrhage are of purely paediatric interest. Others such as that on subarachnoid haemorrhage cover ground as well or better covered in more general books on cerebrovascular disease.

The practical value of the book is not improved by an underusage of subheadings to break up large sections of text and the absence of any line diagrams (essential when explaining the thrombophilias), though there are well reproduced x ray and scan photographs. There are too many repetitions, for example haemostatic disorders get a chapter on their own but many are mentioned again in some detail in the chapter on rare genetic causes of stroke. Mitral valve prolapse is well covered in the chapter on cardiac disorders and stroke but again reviewed along with some other cardiac disorders in the genetic disorders chapter. Many neurologists will be surprised to see migraine listed as a rare genetic cause of stroke, whilst there is a separate sensible chapter devoted to migraine and cerebral infarction, which approaches this difficult topic with a lot of good sense. Notwithstanding these criticisms, this book should be helpful to those caring for young stroke victims, since most of the information required is here and one's reference manager can be loaded with the sources of any further details one may ever require.

CHRIS ALLEN

Images of Mind. By Michael I Posner and Marcus E Raichle. (Pp 257 Illustrated £19.95.) Publishers: W H Freeman and Co Ltd, Oxford 1994. ISBN 0-7167-5045-7.

Over the past 20 years radiological advances have transformed the everyday practice of neurology. The impact of functional imaging on neuropsychology, the subject of *Images of Mind*, promises to be just as profound.

Written by two of the doyens of the field, this book is an invigorating canter through the common ground between cognitive and

neuroscience. Cognitive science uses the methods of psychology to dissect mental operations, such as the act of reading, into their component processes. The use of such dissections to guide the design of studies in functional imaging creates a powerful tool to explore the neural basis of our mental life. Simple experiments have proved revealing: subtraction of the activity set up by looking at an unpronounceable consonant string from the response to a plausible "pseudo-word", for example, provides a wealth of intriguing data.

Posner and Raichle concentrate on the three areas in which the combination of psychological analysis and functional imaging has won its spurs: vision, language and attention. They accept that functional imaging—whether by PET or MRI—is frustratingly slow: it relies on secondary changes in blood flow in areas of active brain, which take seconds to develop, while complex psychological operations can be completed in under a second. An interesting chapter discusses the use of event related potentials, which can track neural activity as it evolves, to complement the results from imaging studies.

Written in a semi-popular style the book is accessible, and well illustrated, but best read an hour before bed: there are data here to tax your cingulate gyri. Its title is provocative but apt: Posner, Raichle and their colleagues world-wide have been remarkably successful in giving "to airy nothing/ a local habitation and a name".

ADAM ZEMAN

Handbook of Neurorehabilitation. Edited by David C Good and James R Couch. (Pp 684 \$185.00.) Published by Marcel Dekker Inc, New York 1994. ISBN 0-8247-8822-2.

Neurological Rehabilitation, Second Edition. Edited by L S Illis. (Pp 581 £69.50.) Published by Blackwell Scientific Publications, Oxford 1994. ISBN 0-632-03282-0.

These books join a growing stream of publications on neurological rehabilitation. They have different strengths but similar shortcomings. Neither book coherently describes the distinctive contribution made by physicians to rehabilitation assessment and therapy (which is ironic, since most of the contributors are medically qualified). Good and Couch begin promisingly with a chapter on clinical assessment but this proves to be a description of conventional neurological routine rather than a functionally-orientated account. Nevertheless Good and Couch's is the more pragmatic of the two books and can function as a brief reference text. It has a compendious index and has useful sections on many topics directly relevant to clinical practice.

Illis's book is less systematic and perhaps more interesting as a result. The book's first edition was a pioneer in the field and the new version is enlarged and enhanced. Its theoretical bias has produced good chapters on topics such as nervous system recovery, plasticity and spasticity but also some less satisfactory material. Rehabilitation comes across as a process which happens to nervous