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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 series[4] 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 disorders[5]—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 postmortem. 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).
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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

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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 marvellous chapter. Having put vascular dementia on the map in the mid-1970s he now retreats and argues that the term has outlived its usefullness. 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

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CONFIDENTIALITY Consent must be obtained from the patient (or if the patient has died, from relatives) to publish *any* information that might alone or in combination identify a patient, whether living or dead, adult or child. However, such details that are given should not be falsified.

ABBREVIATIONS Measurements should be expressed in SI units (see *BMJ* 1991;302:338–41. *SI unit conversion guide* 1992; Boston: New England Journal of Medicine). For recognised abbreviations see *Units, Symbols, and Abbreviations*, Fifth Edition 1994, edited by DN Baron, Royal Society of Medicine: London.

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Coers C, Woolf AL. *The innervation of muscle. A biopsy study*: Oxford: Blackwell, 1951:16-24.

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