Progressive supranuclear palsy: neuropathologically based diagnostic clinical criteria

In their recent retrospective clinicopathological study of 12 cases of progressive supranuclear palsy (PSP), Collins et al. 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 Escourroule Neuropathology Laboratory, Paris. 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 ataxiology; (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) slow demonstration of improvement with levodopa treatment. Almost all of these diagnostic criteria are identical to those used by Collins et al., except for retrocortical 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.1 In another recent series1 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—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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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 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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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 Tjänarinnor 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 essentially stoppage of and to some extent reversible.

Announcement from the British Neuro-psychiatry Association
The 1995 Summer meeting—to include joint sessions with the British Association for Psychopharmacology—will be held on 15–17 July at 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 at the address above, or Dr Jonathan Bird, Burden Neurological Hospital, Stoke Lane, Stapleton, Bristol BS16 1QT, UK.
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SUBMISSION Please submit articles on clinical neurology, neurosurgery or psychiatry (especially neuropsychiatry) to Professor B A C Hughes, Editor, Journal of Neurology, Neurosurgery, and Psychiatry, Medical School Building, UMDS, Guy's Hospital, St Thomas Street, London SE1 9RT, UK. Telephone: 0171-378 6758 and fax 0171-378 1221. Supply up to three key words or phrases suitable for use in an index. We need four copies of the manuscript and figures. Manuscripts should conform to the "Uniform requirements for manuscripts submitted to biomedical journals (BMJ 1991; 302:338-41)". Follow the format of articles in this issue and submit your text double-spaced, on one side of the paper. Receipt will be acknowledged. If the paper is rejected the manuscript will be shredded after three months. Original figures will be returned if requested when the paper is submitted. If requested, you should produce the data upon which the manuscript is based for examination by the Editor. The article must not duplicate material published or submitted elsewhere. The article should be accompanied by the following statement, signed by all the authors: "No work resembling the enclosed article has been or will be published anywhere than in the JNNP".

Full Papers must present important and substantial new material. Short Reports and Letters may also be submitted. Topics suitable for presentation for Short Reports include single case reports which illustrate important new phenomena, or reports of short, original research studies. Short reports should be restricted to about 1500 words with a minimum of references and no more than one figure and one table. Short case reports may be selected for a Lesson of the month series. Neurological Pictures occupying one journal page, following a format similar to that in this issue, and with a maximum of five authors, will be considered. Letters should be no longer than 1000 words, with a maximum of five references and no more than one illustration or table. Short letters concerning papers published in the journal will be printed under Matters Arising. Occasional Reviews, Clinicalopathological Case Conferences, and regular Editorial will be solicited by the Editor and are subjected to a review process. Authors wishing to submit an editorial, clinico-pathological case conference, or review should seek the advice of the Editor in advance.

AUTHORSHIP All authors must have participated sufficiently in the work to take public responsibility for the content (see BMJ 1991;302:309).

ETHICS Ethical considerations will be taken into account in the assessment of papers (see the Medical Research Council's publications on the ethics of human experimentation, and the World Medical Association's code of ethics, known as the Declaration of Helsinki (see BMJ 1964;2:1777)).

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