MATTERS ARISING

Neuropsychological assessment in patients with multiple sclerosis and mild functional impairment

The paper by Anzola *et al*¹ addressed the important issue of cognitive functioning in patients suffering from MS. We appreciate their study, and share to a large degree their conclusions concerning cognition in MS patients, and we can contribute some additional information.

A) The authors did not mention the exact number or the percentage of patients with deficits on neuropsychological assessment; apparently, number as well as degree of impairment allowed characterisation of the deficit as "very mild". B) The authors attributed their deviating results to their selection of ambulatory patients with relapsing-remitting course of MS. C) They considered the pattern of impairment (inferior performances in concept formation, non-verbal reasoning and verbal memory tests) as indicative of so-called subcortical disruption. We wish to confirm A, comment on B and query C.

A) For counselling and management it is important to know that MS is not a sufficient or necessary condition for suffering cognitive defects, let alone dementia. Findings that are at variance with current quite high estimations of cognitive defects in MS will ultimately add to revealing the as yet insufficiently known spectrum of severity in MS.² Our findings concur with those of Anzola *et al*. In a comprehensive neuropsychological study of 39 outpatients with relapsing-remitting ($n = 20$) and chronically progressive ($n = 19$) MS, who presumably were slightly more handicapped than the patients of their study (table), and all of whom were in quiescent disease stages, we also found evidence of generally adequate cognition. On a case by case basis we found signs of cognitive decline in 18% of the patients.³

B) The suggestion of mild physical handicap and relapsing-remitting course of MS explaining the absence of MS-related dementia cannot be endorsed by our findings. We studied the explanatory value of several ill-

ness variables, among which Kurtzke DSS, duration of illness, and course of MS (RR versus CP). However, using parametric and, when appropriate, nonparametric procedures, we failed to identify a significant influence of any of these variables in any of the behavioural measures (table). The critical illness variables, apart from extensive periventricular demyelination, as stressed by the authors, remain to be identified.

C) In our view, the presence of weak memory performance, poor concept formation and poor nonverbal problem solving is insufficient to result in subcortical dysfunction. The distinction between cortical and subcortical "dementia" rests on inferring the mental disorganisation underlying poor overt performances. Important variables underlying so-called cortical performance deficit should be disordered instruments of cognition. Key variables underlying so-called subcortical performance deficit should be apathy and slowness of information processing. The discrepancy between relatively adequate acquisition and poor retrieval should be taken as the distinguishing feature of so-called subcortical memory failure.⁵ The authors present no data that may help to decide for one or the other mental disorganisation. We may gain some additional information, by reporting that, given the weak acquisition in some of the patients, no clues for a specific retrieval deficit were present in our group. This would render the subcortical or white matter hypothesis of mental deficits questionable.

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- 1 Anzola GP, Bevilacqua L, Cappa SF, *et al*. Neuropsychological assessment in patients with relapsing-remitting multiple sclerosis and mild functional impairment: correlation with magnetic resonance imaging. *J Neurol Neurosurg Psychiatry* 1990;53:142-5.
- 2 Herndon RM. Cognitive deficits and emotional dysfunction in multiple sclerosis (Editorial). *Arch Neurol* 1990;47:18.
- 3 Jennekens-Schinkel A, Velde EA van der, Sanders EACM, Lanser JBK. Visuospatial problem solving, conceptual reasoning and sorting behaviour in multiple sclerosis outpatients. *J Neurol Sci* 1989;90:187-201.
- 4 Jennekens-Schinkel A, Velde EA van der, Sanders EACM, Lanser JBK. Memory and learning in outpatients with quiescent multiple sclerosis. *J Neurol Sci* 1990;(in press).
- 5 Cummings JL, Benson F. Subcortical dementia. Review of an emerging concept. *Arch Neurol* 1984;41:874-9.

Annola *et al* reply:

We completely agree with the remarks in A. Indeed, the computation of the percentage of impaired patients (considering as "impaired" a patient who had a score below the cut-off on at least two tests) yielded a surprisingly similar result (19 versus 18% reported by Jennekens-Schinkel *et al*). As for B) we agree about the lack of correlation of neuropsychological impairment with the Kurtzke score. The data reported in the table are indeed impressive on the lack of difference between chronic progressive and relapsing remitting patients, although we should point out that the findings of other investigators are not in agreement.¹ As for the last point C) the label of subcortical dementia was only meant to be clinical in nature, indicating a cognitive impairment without prominent language and visuo-spatial disorders. We are fully aware of the fact that the very concept of subcortical dementia is controversial, and that other findings may lead investigators to a thorough reappraisal of the concept.

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- 1 Beatty WM, Goodkin DE, Monson N, Beatty PA. Cognitive disturbances in patients with relapsing remitting multiple sclerosis. *Arch Neurol* 1989;46:1113-19.

Asymmetrical "temporal" Pick's disease?

In addition to the article by Scheltens *et al*¹ concerning a patient with a progressive aphasia based on a bilateral temporal Pick's disease we report a case of aphasia based on a unilateral temporal abnormality.

A 77 year old woman showed slowly progressive misunderstanding of words, starting at the age of 65 years. There was no precipitating factor. She was in good physical condition. Over the next couple of years she developed expressive and receptive aphasia. Except for her phasic disturbance no problems in social function or in cognition were established, until the last year. Most obvious were changes in personality. She became more and more inactive in house-keeping and more and more compulsive. Also a slight progressive disorientation to place occurred, followed by disorientation to place and persons. At admission a complete expressive and receptive aphasia was established, she could only sing and did this in a fanatical way. Reading and writing were completely disturbed. Some visual agnosia seemed present, combined with a slight apraxia. There were no deficits in attention or concentration. Activities of daily life were only possible when she was encouraged. She actively made contact with other patients. Internal and neurological examination showed no abnormalities.

Except for her son, who had multiple sclerosis for more than 20 years, there were no internal or neurological diseases in her family.

The EEG in the first year of her disease was normal and remained normal during the years thereafter. The CT scan revealed enlargement of the temporal horn of the left ventricle with temporal lobe atrophy on the same side (figure). Besides that there was a mild generalised cortical atrophy.

Table Comparison of patients with relapsing-remitting (RR) and chronically progressive (CP) MS. Demographic, clinical and cognitive data.

	RR ($n = 20$) Mean (Range)	CP ($n = 19$) Mean (Range)
Age (years)	38.9 (17-60)	49.9 (33-73)
Education (years)	12 (6-18)	11 (6-17)
Age at onset (years)	29.5 (15-45)	35.0 (19-50)
Duration of MS	9.5 (1-25)	15.0 (2-48)
Kurtzke Disability Score	2.8 (0-7)	4.2 (2-7)
Raven's Progressive Matrices*	112.6 (93-130)	109.6 (91-130)
Wechsler Memory Scale#	111.9 (92-143)	111.6 (89-135)
Short Tale	8.9 (2-13)	7.6 (3-11)
Digit Span	10.5 (7-15)	9.8 (6-15)
Block Span (Knox)	9.0 (5-14)	8.5 (3-13)
Pattern Learning (7/24)*	0.6 (0-2)	0.7 (0-2)
Word Fluency	50.5 (13-79)	41.6 (18-65)
Wisconsin modified Card Sorting*	4.1 (0-26)	5.4 (0-42)

*Standard Progressive Matrices: Intelligence Quotient.

#Memory quotient. *Errors.