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.

Figure CT scan revealing enlargement of the temporal horn of the left ventricle with temporal lobe atrophy on the same side.

The development of the disease in the patient resembles the description of the patient in the article by Scheltens *et al.*¹ Our patient, however, showed a unilateral lesion in the temporal region of the brain as the most striking feature. Thus conforming with Scheltens' case we assume our patient had a unilateral temporal form of Pick's disease. These temporal lobe variants are probably more frequent than had been suspected.

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- 1 Scheltens Ph, Hazenburg GJ, Lindeboom J, Valk J, Wolters EC. A case of progressive aphasia without dementia: "Temporal" Pick's disease? *J Neurol Neurosurg Psychiatry* 1990;53:79-80.

Scheltens *et al* reply:

We thank Dr Colon *et al* for their comment on our article.¹ Their clinical description of the patient indeed resembles the one given by us. We based our clinical diagnosis of a strictly temporal form of Pick's disease on the age of onset (50 years), the relative sparing of visuospatial abilities and memory function and the bilateral temporal atrophy seen on MRI.

In the case of Colon *et al* the age of onset was 65 years with a 12 year duration of slowly progressive aphasia. However, disorientation in time, place and persons was also prominent and together with the slight apraxia and agnosia, a diagnosis of Alzheimer's disease might be just as likely. In that respect it resembles the case of Pogacar and Williams.²

Unfortunately, in the case of Colon *et al* only CT was performed. MRI, with its higher sensitivity might have shown more lesions in the left hemisphere or lesions in the contralateral temporal lobe.

Since our paper was published we have had the opportunity to examine another patient, aged 63 years, with a severe slowly progressive receptive aphasia for six years, without any precipitating factor. An MRI scan showed extensive bilateral temporal damage.

We agree with Dr Colon and colleagues that temporal variants of primary degenerative cerebral disorders are probably more frequent than had been expected.

Improved neuro-imaging techniques have contributed significantly to this knowledge.

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- 1 Scheltens Ph, Hazenberg GJ, Lindeboom J, Valk J, Wolters E Ch. A case of progressive aphasia without dementia: "temporal" Pick's disease? *J Neurol Neurosurg Psychiatry* 1990;53:79-80.
- 2 Pogacar S, Williams RS. Alzheimer's disease presenting as slowly progressive aphasia. *RI Med J* 1984;67:181-5.

Complications of carotid angiography

We found the survey of complications of carotid angiography¹ interesting, but do not feel it illuminates the authors' goal in evaluating the risks of angiography vis à vis carotid surgery. Most of their patients (about 75%) could not be considered for carotid endarterectomy, so complications in the total cohort studied are largely irrelevant.

Many of the procedures are of historical interest only, since the series encompasses the period from 1977-86, during which time the approach, methodology and materials for angiography have undergone radical changes.

The conclusions do not seem to relate to the data. The authors state that angiographic risks will be reduced by selective, aortic arch procedures, yet in their series, only one patient developed a complication following direct carotid puncture, which argues just the opposite. There are no data to substantiate their statement that patients with least complications should be "younger, systemically well and neurologically stable". The mean age of the ten patients with neurological complications was 58.9/year, a little less than the mean age of the whole series.

Surely their data represent simply a personal audit of a whole spectrum of neurological disorders, in patients aged seven to 76 years, and it would be misleading to extrapolate these results to the special subset of patients being screened for carotid surgery. In a recent report of angiographic complications in 1002 consecutive procedures, permanent neurological sequelae were encountered in only 0.3% of cases, and in 0.7% of those undergoing angiography for cerebrovascular disease.²

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- 1 Complications of cerebral angiography for patients with mild carotid territory ischaemia being considered for carotid endarterectomy. *J Neurol Neurosurg Psychiatry* 1990;53:542-8.
- 2 Dion JE, Gates PC, Fox AJ, Barnett HJM, Blom RJ. Clinical events following neuroangiography: a prospective study. *Stroke* 1987;18:997-1004.

Hankey *et al* reply:

Drs Zhu and Norris have raised several points

about our paper to which we would like to reply. It is true that after cerebral angiography in 382 patients, 74% (282) were not considered to have an "operable" lesion at the carotid bifurcation but it was stated that "carotid ultrasound facilities were not available". If patients could have been screened by duplex carotid ultrasound, as we advocate in our conclusion, then perhaps many or even most of those 74% of patients who did not have an "operable" lesion would have been spared the unnecessary risks and costs of angiography. However, only two of the eight patients, who had a post-angiographic stroke, did not have an "operable" lesion (50-99% stenosis of the symptomatic internal carotid artery) and one of those two had an occluded ICA which can be difficult to diagnose with duplex alone. So, if duplex had been available ideally it could have reduced the number of angiograms being performed (by 74%) to 100 but it would have only reduced the absolute number of post-angiographic strokes from eight to seven with a significant increase in the relative risk of angiography from 2.1% (eight strokes out of 382 angiograms) to 7% (seven strokes out of 100 angiograms).

The fact that our series is of "historical interest only" applies to any series of patients. This is a general problem with reporting medical information and generalising the results to future practice. Unlike many other series, however, at least we described what type of investigations were done from which the reader can draw their own conclusion. Besides, 67% of patients were studied with selective carotid angiography, which is still current practice. The only real difference in the management of this cohort of patients from those of today is that they were not screened with duplex carotid ultrasound, as mentioned above.

In our concluding paragraph we stated that "it would seem logical to assume that the complications of cerebral angiography can be reduced if patients are selected carefully for the procedure" and that the selection criteria should include "physiologically younger patients who are systemically well, not uraemic and neurologically stable." This conclusion was prefaced as such to indicate that it was not derived from our data but from what data there are and what we believe is common sense. Apart from the lack of any statistical difference between the ages of the whole cohort and those with neurological complication (as inferred by Drs Zhu and Norris) we were referring to patients who were physiologically younger. We also suggested that "the risk of angiography will be reduced if arch and/or selective cerebral angiography is performed . . . using a transfemoral approach". It is not possible to prove or disprove this claim (which again was not based on our data but on common sense) because there have been no randomised studies comparing the complication rates of cerebral angiography via the transfemoral approach with direct carotid or brachial puncture. The few non-randomised comparisons, for what they are worth, yield no significant difference in permanent neurological complication rates.¹⁻³

The advantages of the transfemoral approach over direct carotid puncture are two fold: 1) it provides greater flexibility by allowing the radiologist to study different arteries without repeated arterial puncture, and 2) the consequences of local complications (such as haematoma, intimal dissection or fragmenta-