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CORRESPONDENCE

Painful stimulation

Once again, the meaningless term "response to pain" has been allowed to appear in *the Journal*.¹

In this particular instance, the patient was unconscious and the brain stem was separated from higher centres. The stimulus may have been *noxious*, but there is no evidence whatsoever that it was painful. Furthermore, as it is insisted, rightly, that other sensory stimuli be fully and accurately described, why is "pain" permitted without qualification? This often means pricking with a pin. Unless the pin or needle is driven in and twisted, the modality tested is in fact sharpness discrimination and not pain—the two are dissociated in some lesions. Mechanical noxious stimulation, which is interpreted as painful in the intact and fully conscious person, may be brought about by skinfold pinch or various other manoeuvres; thermal and chemical noxious stimulation also, of course, exist. But unless the nature of the stimulus is specified, the expression *noxious stimulation* (let *pain* and *painful stimulation* be forever abolished) is not only almost devoid of relevance, but misleading.

It also seems unfortunate that the recent *Journal of Neurology, Neurosurgery, and Psychiatry/Association of British Neurologists* supplement on stroke did not mention central poststroke pain. This difficult neurological sequel occurs in some 8%–10% of survivors of stroke^{2,3} (6000–7000 new cases/year in the United Kingdom). Its correct diagnosis, treatment, and possible prevention should be a matter of great concern to neurologists.

D BOWSHER

Pain Research Institute, University Hospital Aintree,
Lower Lane, Liverpool L9 7AL, UK

Correspondence to: pri@liv.ac.uk

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Is it really Alzheimer's disease?

We read the recent article of Cappa *et al* with great interest.¹ Using HMPAO SPECT, the authors studied 24 patients diagnosed as having (probable) Alzheimer's disease (AD) on the basis of DSM-III-R and the

NINCDS-ADRDA criteria. According to Butters *et al*, they distinguished, on the basis of neuropsychological tests, patients with a diffuse pattern of cognitive deficits (dAD) from those with focal temporal lobe dysfunction (FTLD).² Patients with FTLD are reported by both studies as having a better cognitive prognosis. Whereas the dAD group had diffuse perfusion deficits, the patients with FTLD showed a circumscribed reduction of tracer uptake in the left or right, or both temporal lobes.

We comment on the problem of suggesting AD in these patients with dementia. In the study of Butters *et al*, seven of the patients of the FTLD group underwent necropsy. Six had the typical histopathological signs of AD. One patient, however, showed the features of hippocampal sclerosis (HS) and not those of AD (Cappa *et al* wrongly state that all necropsied cases of Butters *et al* had AD). Two major groups of patients with the histopathological signs of HS have been described in the literature: (1) HS is the most common morphological substrate of medial temporal lobe epilepsy (MTLE). The affected patients are known to show characteristic memory impairment, which correlates well with the degree of pathological changes. Progress of memory impairment in MTLE is very slow and other cognitive functions are mostly preserved.³ Most (but not all) of them have an abnormal MRI signal and HMPAO SPECT usually shows temporal hypoperfusion.⁴ (2) Patients with HS have been described in necropsy series of patients with dementia.⁵ It is unknown whether these patients could have been identified by MRI, whether they had abnormal brain perfusion, and what the perfusion pattern would have been.

Cappa *et al* did not exclude patients with epilepsy, and it is not clear if their MRI procedure included high resolution T2/FLAIR sections angulated along or perpendicular to the hippocampus, which considerably increases the sensitivity for detection of MTLE-HS. They also had no histopathological evidence of AD. Thus, the authors may have included an unknown number of patients who actually did not have AD, but, for example, had temporal lobe epilepsy or dementia with the morphological substrate of HS. It is noteworthy that in a recent histopathological study on the causes of dementia (with further references)⁶ it was noted that fewer patients than clinically suspected fall into the histopathological category of AD but—among others—fall into the category of HS.

C G BIEN

C HELMSTAEDTER

C E ELGER

University of Bonn, Department of Epileptology,
Sigmund-Freud-Str. 25, 53105 Bonn, Germany

Correspondence to: Dr C G Bien
c.bien@uni-bonn.de

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Cappa, *et al* reply:

We are grateful to Bien, Helmstaedter, and Elger as they allow us to clarify a point of our study that was not explicitly discussed in the paper recently published in this *Journal*.¹

We obviously agree with them that, in the absence of histopathological confirmation, the diagnosis of "probable" Alzheimer's disease (AD) can be questioned on methodological grounds, particularly in patients with focal temporal lobe dysfunction (FTLD).

However, we think it rather implausible that some of our patients with FTLD were in reality affected by other non-AD diseases, in particular by those suggested by Bien *et al*: medial temporal lobe epilepsy (MTLE) or dementia sustained by hippocampal sclerosis (HS).

As for MTLE, which Bien *et al* consider a relevant source of possible errors in the diagnosis of patients with AD and FTLD,² we emphasise that none of our patients—with diffuse cognitive impairment (dAD) or with FTLD—was affected by epilepsy. In our paper, the exclusion of epileptic patients was implicit in the statement that all our patients met the NINCDS-ADRDA criteria for probable AD (according to these criteria, patients who have epilepsy could at the most attain a diagnosis of possible AD).

Apart from MTLE, we are certainly aware that HS is sometimes responsible for a dementia syndrome mimicking AD, as recently discussed in several neuropathological studies.³⁻⁶

On the other hand, a careful review of these studies leads to the following conclusions: (1) in a general demented population, pure HS (HS without Alzheimer-type or other well characterised degenerative or vascular pathological changes) seems to be a very rare cause of dementia. Ala *et al*³ found only 0.4% of pure HS in a retrospective study of 1771 unselected demented patients and Jellinger⁷ obtained similar results (0.53%) in a consecutive necropsy series of 746 demented subjects older than 55 years! (2) HS significantly contributes to dementia only in old or very old (≥ 80 years of age) demented patients with documented cardiovascular diseases (for example, ischaemic heart disease, arrhythmias, congestive heart failure in about 88% of cases) or with depression as another frequent (63%) clinical feature.^{4,6} (3) When HS is associated with other neuropathological changes, the precise boundaries with other forms of dementia are often less clear and the differential diagnosis may be controversial even at a pathological level. In the necropsy series of Corey-Bloom *et al*,⁴ for instance, more than 50% of patients with HS had enough amyloid plaques to meet NIA criteria⁸ for AD; and even using the more conservative criteria of CERAD,¹⁰ four patients out of eight could have been diagnosed as AD. (4) It is quite uncommon for demented patients with postmortem evidence of HS to have previously received a clinical diagnosis of probable AD (in most cases, patients with HS had been diagnosed as possible AD⁹).