A behavioural audiogram showed mild bilateral sensorineural hearing loss in the frequencies of 2000 Hz-8000 Hz with normal speech discrimination, interpreted as presbycusis. The brain stem auditory evoked potentials were normal.

Brain T1 and T2 weighted MRI with gadolinium enhancement disclosed low lying cerebellar tonsils with elongation of the medulla and pons, compatible with ACM I. A brain stem or cervical syrinx was not demonstrable (fig 1).

In view of the progressive symptomatology, posterior fossa decompression was considered, but postponed because of the cardiac situation of the patient. Clonazepam was offered to the patient for alleviation of the posturally evoked symptoms, but was refused because of possible sedation.

Central paroxysmal positioning vertigo occurs transiently on changing the head position, as opposed to central positional nystagmus, which persists as long as the head position is sustained.^{3,4} Central paroxysmal positioning vertigo can be differentiated from benign paroxysmal positioning vertigo by its shorter latency, longer duration of the attack, and a direction changing nystagmus which is not attributable to stimulation of a single canal.^{3,4} It usually indicates a lesion around the fourth ventricle, dorsal vermis, or diffuse cerebellar pathology.³ Central positional nystagmus is often bilateral, of low constant frequency, and occurs in association with lower brain stem or vestibulocerebellar lesions.³

In 1977 Drachman *et al* described the entity of PEV.⁵ In their two patients (one with a metastasis and the other with a glioblastoma of the cerebellar vermis) PEV was a prominent clinical sign, whereas vertigo and nystagmus diminished with the progression of the pathological process. Positional vomiting became so severe that the patients had to hold their head in a forced position to prevent vomiting. The symptoms cleared after treatment with vestibular suppressants. Drachman *et al* explained the condition of PEV by dissociation of the vomiting centres from oculomotor and receptive pathways of the complex vestibular syndrome by a brain stem infiltrating lesion.

Arbusow *et al* described a patient with amiodarone induced positional vomiting with mild vertigo, downbeat nystagmus, and limb ataxia, responsive to benzodiazepines. According to their concept, it is in keeping with that of Drachman *et al*, amiodarone disinhibits selectively the connections between the dorsal cerebellar vermis and the vomiting centres in the postremal area and the lateral

reticular formation, sparing relatively the vermis-ocular motor circuitry.⁶

Severe positioning nausea and vomiting with inconsistent dizziness and without nystagmus, was present in this patient with ACM I. Initially, nausea occurred only on head extension, but later on all head movements but anteflexion. An ENG showed only low speed positional—that is, sustained nystagmus in one head direction. On Hallpike positioning testing nystagmus was not recorded. Other ocular motor findings (saccadic pursuit and abnormal optokinetic nystagmus) were compatible with a cerebellar lesion.^{3,4}

In ACM, a normal vestibular input after changes in head position might be misprocessed by transient brain stem compression, leading to dissociation of the vomiting centres from other parts of the vestibular reflex. The relatively mild signs of positional vomiting in our patient might be due to the benign nature of the underlying disease, as opposed to the invasiveness of tumours or acute neurotoxicity, leading to severe PEV.^{5,6}

This patient with ACM seems to be unique in the literature, as PEV has not been reported in this clinical setting.

Further neurophysiological studies are needed to elucidate the pathogenesis of PEV.

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- 1 Stevens JM, Serva WAD, Kendall BE, *et al*. Chiari malformation in adults: relation of morphological aspects to clinical features and operative outcome. *J Neurol Neurosurg Psychiatry* 1993;56:1072-7.
- 2 Bronstein AM, Miller DH, Rudge P, *et al*. Down beating nystagmus: magnetic resonance imaging and neuro-otological findings. *J Neurol Sci* 1987;81:173-84.
- 3 Brandt T. *Vertigo. Its multisensory syndromes*, 2nd ed. London: Springer-Verlag 1999:199-213, 291-8.
- 4 Baloh RW, Halmaghi GM. *Disorders of the vestibular system*. New York: Oxford University Press, 1996:446-53, 488-93.
- 5 Drachman DA, Diamond ER, Hart CW. Posturally evoked vomiting: association with posterior fossa lesions. *Ann Otol Rhinol Laryngol* 1977;86:97-101.
- 6 Arbusow V, Strupp M, Brandt T. Amiodarone induced severe prolonged head positional vertigo and vomiting. *Neurology* 1998;51:917.

Lunchtime headache

Chronic primary unilateral headaches fall into one of five categories: chronic cluster headache, chronic paroxysmal hemicrania, hemicrania continua, cervicogenic headache, and SUNCT syndrome. Overlap between types is recognised. Although the differentiation of these is sometimes difficult, there are important therapeutic implications—for example, indomethacin has a dramatic effect on chronic paroxysmal hemicrania but is less effective in chronic cluster headache. Here, a patient with paroxysmal unilateral headaches, occurring precisely on the same day of the week and at the same time, is described.

A 57 year old man presented with episodic, right sided, moderate to severe headaches of 9 months' duration. He developed these always on a Monday at 1300 hours. The

headaches were sharp and throbbing with the maximum pain behind the right eye. The headache then radiated to the back of the head without crossing the midline. The pain was felt in paroxysms each lasting several minutes. He did not experience any visual symptoms, nausea, nasal congestion, lacrimation, or ptosis. Initially the total duration of the headaches was about 10 hours but it became progressively longer, remaining until Wednesday on some occasions. They were not related to the consumption of food and he did not go out drinking on Sunday nights.

At the age of 20, he had had four episodes of severe headache on the right on four consecutive days after breaking rest during a week of night shifts as a warehouseman. In between the Monday "lunchtime headaches", he remained well. He had no personal or family history of migraine. He was taking propranolol for mild hypertension. His examination was normal. There was mild cervical spondylosis on plain radiology. Brain CT showed minor atrophy.

A diagnosis of chronic paroxysmal hemicrania was made, and he was prescribed indomethacin (50 mg three times on Mondays). With this his headaches were delayed until Tuesday morning and the duration of the headaches was reduced to less than 24 hours. The headaches also became less predictable. A more protracted course of indomethacin rendered him headache free.

Review of this man's headache history suggests that the episode he had when aged 20 was probably an isolated "cluster" headache. The more recent, highly predictable, right sided headaches on Monday afternoons were unusual. The predictability of these was such that he could time it precisely to 1300 hours. Their characteristics were not classic for chronic paroxysmal hemicrania, in which attacks last between 20 and 30 minutes and are accompanied by ipsilateral nasal congestion and lacrimation.¹ Attacks of chronic paroxysmal hemicrania can, however, have clockwork regularity and are abolished by indomethacin.¹ Headaches in this case are also different from chronic cluster headache as the paroxysms occurred many times throughout the day (repetition rate 12-16/day) and the total duration of an "attack" could be up to 72 hours. Recently another variant of chronic paroxysmal hemicrania—hemicrania continua—has been described² and these headaches are non-paroxysmal and continuous. The degree of cervical spondylosis in this patient was mild, and furthermore the nature of the attacks was not that of cervicogenic headache. SUNCT syndrome is a type of paroxysmal unilateral headache consisting of short lasting, unilateral, neuralgiform headaches with conjunctival injection and tearing.^{3,4} Patients have up to 30 paroxysms/hour, occurring once or twice a day.³ This diagnosis is also highly unlikely in our patient, with his clinical presentation and therapeutic response. It was thought that this man's headaches were most probably due to chronic paroxysmal hemicrania evolving from a possible early "cluster" attack, supporting the suggestion that a common pathophysiology underlies these "trigeminal-autonomic cephalalgias".⁴ The precise predictability of the Monday afternoon headaches is fascinating, and unexplained.

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Figure 1 Brain MRI shows low lying cerebellar tonsils and elongation of the pons and medulla, compatible with ACM I.

- 1 Sjaastad O, Dale I. A new (?) clinical headache entity "chronic paroxysmal hemicrania". *Acta Neurol Scand* 1976;54:140-59.
- 2 Sjaastad O, Spierings ELH. Another headache absolutely responsive to indomethacin. *Cephalalgia* 1984;4:65-70.
- 3 Pareja JA, Sjaastad O. SUNCT syndrome, a clinical review. *Headache* 1997;37:195-202.
- 4 Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short lasting headaches with autonomic features including new cases. *Brain* 1997;120:193-209.

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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.

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- 1 Wijdicks EFM, Atkinson JLD, Okazaki H. Isolated medulla oblongata function after severe traumatic brain injury. *J Neurol Neurosurg Psychiatry* 2001;70:127-9.
- 2 Andersen G, Vestergaard K, Ingeman-Nielsen M, et al. Incidence of central post-stroke pain. *Pain* 1995;61:187-94.
- 3 Bowsher D. Stroke and central post-stroke pain in an elderly population. *Journal of Pain* 2001 (in press).

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.

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- 1 Cappa A, Calcagni ML, Villa G, et al. Brain perfusion abnormalities in Alzheimer's disease: comparison between patients with focal temporal lobe dysfunction and patients with diffuse cognitive impairment. *J Neurol Neurosurg Psychiatry* 2001;70:22-7.
- 2 Butters MA, Lopez OL, Becker JT. Focal temporal lobe dysfunction in probable Alzheimer's disease predicts a slow rate of cognitive decline. *Neurology* 1996;46:687-92.
- 3 Helmstaedter C, Elger CE. The phantom of progressive dementia in epilepsy. *Lancet* 1999;354:2133-4.
- 4 Menzel C, Hufnagel A, Grunwald F, et al. The relevance of interictal rCBF brain SPECT in temporal lobe epilepsy: diagnostic value and

effects of spatial resolution. *Ann Nucl Med* 1995;9:215-23.

- 5 Corey-Bloom J, Sabbagh MN, Bondi MW, et al. Hippocampal sclerosis contributes to dementia in the elderly. *Neurology* 1997;48:154-60.
- 6 Crystal HA, Dickson D, Davies P, et al. The relative frequency of "dementia of unknown etiology" increases with age and is nearly 50% in nonagenarians. *Arch Neurol* 2001;57:713-19.

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).