showed an extensive basal subarachnoid haemorrhage that had diffused into both sylvian fissures and into the sulci over the convexity of the cerebral hemispheres. In addition there was a rounded heterogeneous density mass in the suprasellar cistern (fig 1 A). After the administration of contrast material the lesion displayed heterogeneous central enhancement (fig 1 B). A ruptured anterior communicating aneurysm was suspected but was excluded by normal cerebral angiography.

Brain MRI confirmed haemorrhage within a pituitary tumour (fig 2 A). It could also be seen that the haemorrhage had ruptured through a defect in the tumour capsule into the subarachnoid space (fig 2 B). Despite the administration of 100 mg hydrocortisone intravenously every 6 hours and cardiopulmonary support her condition progressively worsened and she died 3 days after admission.

The clinical syndrome of pituitary apoplexy evolves within hours to days.² The symptoms vary from mild to severe and can progress rapidly to coma and death. Because many patients are unaware that they harbour a pituitary tumour, pituitary apoplexy is often unrecognised at presentation. In pituitary apoplexy, blood and necrotic tumour tissue are enclosed and compressed within the confined space of the sella turcica, a region that is in close anatomical proximity to the basal cisterns. When the pressure gradient within the sella exceeds the resistance of the surrounding structures, blood is expelled into the subarachnoid space producing a clinical picture that may be indistinguishable from aneurysmal SAH.³ Thus pituitary apoplexy must be included in the differential diagnosis of "angiographically negative" SAH.⁴ Brain CT is the modality of choice in the initial investigation of SAH. It will show the spread and severity of the haemorrhage within the

subarachnoid space, as well as any extension into the brain.¹ When pituitary apoplexy is the cause, CT will usually show the pituitary tumour as well as any recent haemorrhage. Brain MRI is superior to CT in identifying the tumour as well as the associated haemorrhage or infarction. In the case presented here, the enhanced MRI also displayed the defect in the tumour capsule which represented the site of rupture. The management of pituitary apoplexy includes the immediate administration of high dose corticosteroids to combat adrenal insufficiency. When there is rapid deterioration of vision or a progressively worsening level of consciousness, urgent surgery in the form of trans-sphenoidal decompression should be carried out.

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Figure 1 (A) Cranial CT without contrast, showing a heterogeneous but mostly high density mass in the suprasellar cistern (arrowheads). There is also extensive subarachnoid haemorrhage. (B) Cranial CT with contrast, showing central enhancement within a heterogenous high density mass (arrowheads).

Figure 2 (A) Coronal T1 weighted MRI, showing suprasellar extension of a sellar mass. The area of high intensity signal (arrow) represents methaemoglobin within clotted blood. Note also the hyperintense tumour rim. (B) Contrast enhanced sagittal MRI T1 weighted image, showing that the bulk of the mass is isointense with brain. There is a defect posteriorly in the superior aspect of the capsule representing the site of rupture (arrow).

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Early detection of non-compliance in Wilson's disease by consecutive copper determination in cerebrospinal fluid

A 41 year old technical employee was diagnosed with Wilson's disease in 1982, 2 years after onset of dysarthria, diplopia, visual deficits, ataxia, and concentration deficits. The patient improved rapidly with *D*-penicillamine and a copper free diet, and has since returned to normal neurological function. Penicillamine was stopped in 1989 and he changed his diet from copper free to a normal diet in 1987. In 1990 he was put on 800 mg zinc a day. During the past 17 years he underwent 20 determinations of copper concentration in CSF for follow up of treatment (figure) as copper concentrations in CSF can be used as an indicator of brain copper concentration in the cerebral manifestation of Wilson's disease.¹⁻⁵

Copper was measured by flameless atomic absorption (Perkin Elmer, HGA 500, Überlingen, Germany). The main resonance line was 324.7 nm with deuterium background compensation and argon as a protective gas; the sample volume was 20 µl using an autosampling system. The CSF was measured undiluted. The patient was now again routinely admitted for monitoring of the efficiency of the treatment. He was free of complaints and the neurological examination as well as a neuropsychological test battery and visually evoked potentials were normal. He explicitly denied any change in diet or drug therapy (zinc, 800 mg a day). We found, however, an almost threefold increase of his CSF copper concentration compared with the previous values (figure). Serum

Copper concentration in CSF during the course of treatment in a 41 year old patient with Wilson's disease. The arrow marks the time when the patient started to take his zinc in reduced doses and irregularly.

copper and ceruloplasmin were unchanged. Confronted with these results the patient admitted that he had been non-adherent to his zinc regimen for the past 2 years by reducing the daily zinc intake to less than 400 mg a day. One established parameter for the efficacy of treatment in Wilson's disease is the balancing of the intake and excretion of copper. This, however, was not feasible as the patient refused to take a standardised diet, rendering the daily copper intake unknown. As is displayed in the figure there had been a continuous decrease of the CSF copper concentrations over the past years, although the treatment consisted only of 800 mg zinc a day without a restriction in copper intake. The repeated determination of the CSF copper allowed for the demonstration of a sufficient treatment and for the detection of non-adherence to the treatment regimen before the reappearance of clinical symptoms. We conclude that the determination of CSF copper reliably detects copper accumulation in the CNS and may be used to monitor the treatment efficacy, particularly in patients with non-adherence to (copper free) diet and drug treatment.

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CORRESPONDENCE

Between will and action

I read with great interest the recent paper by Bundick and Spinella¹ and the related commentary by Goldberg.² These articles address the neurological substrates of volitional disturbance and in places they adopt the vocabulary of the philosophy of action—for example, Goldberg refers to the “will”. However, their uses of action related terms are mutually inconsistent and some clarification might assist in elucidating the functional anatomical relevance of the disorders described.

If we are to use the terms “will” and “action” then their use should be internally consistent. The “will” is that process that deliberates (consciously) on what is to be done, and the “action” is the performance, which follows on from that deliberation.³⁻⁴ (It should be noted that the temporal sequence of this philosophical model of volition is substantially undermined by the classic EEG experiments of Libet *et al* on “intention”.⁵⁻⁶) An “action” is consciously chosen; there is no such thing as an “involuntary action” (according to this model). Involuntary movements are movements not the intended actions of the agent (“the one who acts”).⁴ It follows that the movements initiated by an “alien hand” may seem purposeful, but they are not actions (chosen by the patient). They are failures of action in so far as the patient cannot make the limb “behave”. Hence, although Goldberg refers to the “will” being involved in action generation, his terminology is extrapolated inconsistently: he refers to alien hands performing “purposeful acts” and “involuntary actions”; the first is an attribute of an agent, the second is an oxymoron.⁴

Bundick and Spinella, by contrast, refer to “involuntary motor activity” and “non-purposeful” movements.¹ These terms are coherent within the context of the “will” vocabulary being used.

The above points are not merely pedantic, as a case can be made for “action” and “agency” implicating different brain systems,⁷ and thus the volitional deficit demonstrated in each form of the alien hand syndrome may have some cognitive-neurobiological relevance.

In the medial-prefrontal and callosal forms of alien hand syndrome, the patients, although they have a failure of motor control, and an inability to impose their “will” on the alien limb, do not generally attribute alien agency to that limb—that is, they do not experience it as belonging to an “other”. However, in the posterior alien hand syndrome, personal agency may be lost, and the limb experienced as belonging to someone else. Consider the case of Leiguarda *et al*,⁸ cited by neither Bundick and Spinella nor Goldberg.^{1,2} This alien limb (associated with ictal activity from a right parietal lesion) was experienced thus by the patient:

“Suddenly I had a strange feeling on my left side; later I could not recognize the left arm as my own; I felt it belonged to someone else and wanted to hurt me because it moved towards me.”⁸

This third form of alien hand syndrome has been referred to previously.⁷ The loss of

agency it comprises is consistent with that alienation noted in “somatoparaphrenia” by Critchley⁹ and is congruent with that attribution of agency to external forces so characteristic of schizophrenic “alien control”; itself associated with functional abnormality of the right parietal region.¹⁰

Hence, a consistent application of action terminology may help to elucidate the functional anatomical correlates of disorders of volition.⁷

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Progressive dementia and gait disorder in a 78 year old woman

Although the provisional diagnosis of gliomatosis cerebri in the *clinicopathological case conference* by Tagliati *et al* was the one eventually validated at necropsy,¹ the discussion should also have entertained the possibility that the occurrence of signal hyperintensity on MRI, in the context of dementia, ataxia, and Babinski's sign could also be consistent with the diagnosis of cerebral amyloid angiopathy (with giant cell inflammatory reaction to B4-amyloid and vasculitis), exemplified by a 63 year old man presenting with some of these stigmata.² In the classic triad, consisting of cognitive impairment, upper motor neuron signs, and lobar haemorrhage,³ the third might well be a criterion potentially interchangeable with, or antedated by amyloid related vasculitis and attendant stigmata such as focal non-specific hyperintensity on MRI.

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