Finally, we would like to add a word of caution and remind the authors that it would be wise to avoid deriving the measure of "objective duration" of sleep benefit from three scales filled in at home by patients without any further instructions than to fill them at waking, and during best and worst before drug intake.

As the only dopamine agonist mentioned in this study is bromocriptine, we would be grateful to know what year the study was conducted in.

In any case, although "little is known about sleep benefit" any study concerning this phenomenon should certainly attempt to increase knowledge and avoid a further increment of confusion.

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Bateman replies:

The difference in results is due to a different definition of sleep benefit. Sleep benefit as defined in our paper refers to mobility as good as "on" on waking, which wears off over a variable period. Högl *et al* define sleep benefit as "self perceived mobility in the morning before drug intake as better than during the rest of the day."

The purpose of the first part of our study was to verify the existence of sleep benefit as we had defined it, particularly in view of the findings of Högl *et al* that "patients with sleep benefit had a small improvement between night and morning" and "sleep benefit patients were clearly in the "off" state during baseline motor examination". Our inpatient study showed that six out of the 16 "who were studied from the moment of waking" performed as well as when they were "on" due to medication. Subsequently they spontaneously turned "off" to an identical state to "off" after medication. Four patients could not be studied from the moment of waking as they awoke before the investigator! We clearly found that sleep benefit is as good as "on" after medication and wears off, not an intermediate stable state between "on" and "off" as Högl *et al* have defined it, by subsequently giving these patients their normal medication on the same day and monitoring their response by half hourly UPDRS and ADL scores.

Aware of the patient's misperceptions about sleep benefit, we wished to confirm as objectively as possible the findings from our outpatient questionnaire by using the ADL rating scales. The inpatient study showed a correlation between motor UPDRS "on" score and ADL "on" score of $r=0.72$, $t=4.42$, $p<0.001$ and sleep benefit ratings $r=0.62$, $t=3.35$, $p<0.01$. The ADL maximum score is 125. It consists of 25 items that can be rated on a five point scale. Dr P Brown, originator of the scale, suggested that a change of 12 would

be sufficient to confirm sleep benefit. As we were aware that sleep benefit, confirmed by personal observation, can represent a substantial change in motor performance, this criterion seemed reasonable. The correlation between motor UPDRS and ADL scores in our study was good, showing that the ADL scores are generally a reliable measure, although there will inevitably be exceptions.

Our study showed that sleep benefit as we defined it was generally a feature of patients with young onset Parkinson's disease. A 73 year old patient described in their paper, with disease onset at 62, would be unlikely to have sleep benefit as we defined it. Their paper, as their figure 2 shows, refers to a different phenomenon.

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Intracranial dural fistula as a cause of diffuse MRI enhancement of the cervical spinal cord

We read the recent short report by Bousson *et al*¹ on spinal MR findings in a patient with progressive myelopathy and intracranial dural arteriovenous fistula with great interest.

We recently had a 42 year old man admitted as an emergency with a 3 week history of stepwise altered sensation in both lower limbs ascending to the torso which progressed to weakness involving his legs and hands. Two years before this he had an episode of severe backache associated with a tight band of pain around the waist and significant bilateral leg weakness. Resolution occurred only after 4 months, when he was able to walk normally. On the current admission examination showed a spastic tetraparesis; there was minimally increased tone in the upper limbs, mild weakness of the small muscles of both hands, and marked pyramidal weakness of the legs with extensor plantar responses. He was unable to support his weight and was in

urinary retention. He had a sensory level at T5 although dorsal column function was preserved.

Brain and spinal cord MRI showed increased signal in the medulla extending into the upper cervical cord down to C4 (figure). Slightly prominent vessels were seen overlying the right cerebellar hemisphere and a varix was visible close to the torcula. There was no enhancement in the cord or medulla and no abnormal flow voids in the spinal veins. The changes were thought to represent a spinal cord infarct and in view of the "stuttering" course in his history we proceeded to cerebral angiography. This showed an arteriovenous fistula supplied by the left middle and posterior meningeal artery and both occipital arteries. Venous drainage was into prominent varices lying just to the left of the midline and in front of the transverse sinus and then on the transverse sinus itself.

After an unsuccessful attempt at embolisation via the arterial route, the fistula was occluded by packing the varix with Guglielmi detachable coils.

He made an uneventful recovery; after 2 months of intensive neurorhabilitation he recovered full function in his upper limbs and now has sufficient power in his legs to be able to walk with the aid of crutches.

This case emphasises that an intracranial arteriovenous fistula should be included in the differential diagnosis of increased signal on MRI of the cervical cord, even when dilated veins are not, as in this case, very apparent. Prodromal symptoms can occur and a careful history in a patient with ascending paraparesis and tetraparesis is essential. Endovascular occlusion at these fistulae can lead to useful improvement in neurological function.

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Inverse relation between Braak stage and cerebrovascular pathology in Alzheimer predominant dementia

Goulding *et al*¹ carried out a preliminary retrospective postmortem analysis of 25 patients (13 men, 12 women, mean age 80.7 years) with the clinical diagnosis of Alzheimer-type dementia (only one with suspected multi-infarct dementia) and a 36.4% frequency of the ApoEε4 allele. Eighteen brains (89%) with neuritic Braak stage ≤4 had either additional cerebrovascular lesions (n=14), or Lewy bodies (n=1), or both (n=6), with a significant inverse correlation between cerebrovascular lesions and Braak stage. Forty eight per cent of the brains showed small focal infarcts, and only 20% disclosed "pure" Alzheimer's disease pathology. No association between the ε4 allele and any pathological variable was found. Based on these data, the authors emphasised the importance of screening for concomitant pathology in Alzheimer's disease, in which a cerebrovascular component has been suggested as an

MRI of lower brain and spinal cord.

additional pathogenic factor. These data can only in part be confirmed by personal experience in a cohort of 27 necropsy cases (13 men, 14 women) aged 77 to 91 (mean 85.9) years with the clinical diagnosis of degenerative dementia (possible or probable Alzheimer's disease) in 24, of vascular dementia in two, and Parkinson's+Alzheimer's disease in one, studied between 1989 and 1998. Mini mental stages (n=14) ranged from 0 to 15 (mean 8.0); ApoE genotyping performed from paraffin blocks using a polymerase chain reaction (PCR) method disclosed the $\epsilon 4$ allele in 33%. In addition to the neuropathological evaluation criteria by Goulding *et al*¹ the NIA² and CERAD criteria for Alzheimer's disease³ were used. The following data were obtained.

The NIA criteria for Alzheimer's disease were positive in all but three cases diagnosed as senile dementia of the tangle type,⁴ 44% staged CERAD A and 28% staged each CERAD B and C. Only 16/27 brains (59%) showed additional cerebrovascular lesions, either white matter changes alone (n=3) or in combination with lacunar state or small focal infarcts in the basal ganglia (n=13), or an old infarct in the area of the left posterior cerebral artery (n=1). Six of 27 (22%) showed additional subcortical (n=4) or both subcortical and cortical Lewy bodies, thus fitting the diagnostic criteria of dementia with Lewy bodies⁵ in two. Neuritic Braak stages 5 or 6 were present in 12 brains (44%), stage 4 in 14 (52%), and stage 2 in one (3%). Association with cerebrovascular lesions was seen in eight cases staged Braak 4 and 5 (or 5 and 6). When looking at the severity of the associated cerebrovascular lesions, severe ones (combined cerebrovascular disease II or III¹) were seen in seven brains with Braak stage 4 and in six staged Braak 5 (or 6), mild ones (cerebrovascular disease I) in one brain staged Braak 4, and in two with Braak stages 5 or 6. Carriers of the ApoE $\epsilon 4$ allele staged Braak 4 in five cases and Braak 5 in three, with additional cerebrovascular lesions in three brains each. Although in our small necropsy cohort, similar to that of Goulding *et al*,¹ "pure" Alzheimer's disease was seen only in 22% (three cases each with Braak stages 4 and 5), in view of our data, the inverse relation between cerebrovascular disease and Alzheimer's disease, suggested by these authors, requires further confirmation in larger and possibly prospective clinicopathological case series.

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Link between the CSF shunt and achievement in adults with spina bifida

We read with interest the results of Hunt *et al* in their long term follow up of spina bifida survivors with intraventricular shunts.¹ In 48 shunt treated adult patients, they find that 27.1% (13 patients) live independently, 35.4% (17 patients) drive a car and 25% (12 patients) are in open employment. They also report that those requiring shunt revision, particularly after 2 years of age, have lower levels of achievement as defined by these three criteria. Overall, 22.9% (11 patients) are "community walkers".

The clinic for adults with spina bifida and hydrocephalus (CASBAH) in Belfast receives referrals on a regional and non-selective basis from paediatric services after the age of 16 years. Of those currently attending the clinic, 95 are shunt treated survivors. There are 50 men and 45 women, average age 25.6 years (range 16-39 years). Fifty three patients (55.8%) are wheelchair dependent, 7 (7.4%) are largely wheelchair dependent but retain some ambulatory capacity, 18 (18.9%) are ambulatory with aid, and 17 (17.9%) are independently ambulatory. Overall 35/95 (36.8%) can be considered "community walkers".

Of 61 patients in whom complete data are available, 22 (36.1%) are in full or part-time employment, 10 (16.4%) are students (six of these in higher/further education), three (4.9%) are in fully fledged training schemes, two receive sheltered training, and 24 (39.3%) are unemployed. Twenty patients (32.7%) are regular drivers and a further two (3.3%) are actively learning to drive. Eight patients are married (13.1%) and three are parents (4.9%). In five patients (8.2%), epilepsy is an active problem.

Our data therefore suggest a rather more optimistic outcome in terms of mobility and employment for long term shunt treated survivors of spina bifida. Interestingly, and again by contrast with the data of Hunt *et al*, these figures differ little from those for our whole clinic population, shunt and non-shunt treated survivors, where 34% are in employment, 33% are unemployed, 32% are regular drivers, 17% are married or engaged, 8% are parents, and 8% have epilepsy.² Although it is possible that the more severely affected shunt treated patients simply did not survive into adulthood, thereby causing an intrinsic bias, these figures would indicate that the presence of a shunt and the potential for subsequent revision has rather less of an impact on prognosis than is suggested by their paper.

We would, however, agree with their conclusion that ease of access to a neurosurgical unit is essential for infants and children with spina bifida. We would further recommend that this continues in adulthood as seven of our patients have required surgical intervention for a Chiari/hydrosyringomyelia complex at that stage, another shunt related complication.³

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Crying spells as symptoms of a transient ischaemic attack

I would like to comment on both the temporal and neuroanatomical aspects of the case of crying spells as symptoms of a transient ischaemic attack (TIA), presumed to involve the right capsular-thalamic region, recently reported by Mendez and Bronstein.¹

As the authors point out, laughter preceding a cerebrovascular event involving the pontine, capsular-thalamic, or lenticular-caudate regions ("le fou rire prodromique") is a rare but well recognised phenomenon, first described almost a century ago. However, they do not mention previous reports of pathological crying heralding ischaemic vascular events,²⁻⁴ including one patient with TIA.³ The term "les folles larmes prodromiques" has been suggested for this phenomenon.⁴ In the series of Davison and Kelman,² case 36 was a hypertensive female patient who had two cerebrovascular events 3 years apart, in the second of which the "ictus was associated with frequent spontaneous crying spells"; there were left sided pyramidal signs, and a right capsular-thalamic insult was suspected.² Tatemichi *et al*³ reported seven patients with basilar ischaemia, in one of whom involuntary weeping was the first symptom; another patient had TIAs with subclavian steal in whom "explosive crying" was a conspicuous feature, along with diplopia, dysarthria, and gait instability. A more recent report presented a case of embolic occlusion of the top of the basilar artery heralded by pathological crying, with probable bilateral midbrain infarction (interpretation was complicated by acute intra-arterial thrombolytic therapy).⁴

As with laughter, the neuroanatomical substrates of crying are diffuse. Both are reflexive pontomedullary activities controlled by at least two supranuclear pathways: a volitional pathway running in the corticobulbar pathways of the posterior limb of the internal capsule; and a more anterior pathway, presumably with inputs from the limbic system, running rostral to the knee of the internal capsule carrying inhibitory and/or facilitatory fibres.⁵ The correlation of pathological crying occurring after strokes with damage to brain areas involved in serotonin-ergic neurotransmission,⁶ and the response of such crying to specific serotonin reuptake inhibitors,⁷ suggests the involvement of the brainstem raphe nuclei and their projections in the pathogenesis of pathological crying. As demonstrated by these cases,¹⁻⁴ both transient and focal brain insults, as well as chronic and diffuse brain disease, may cause crying spells, presumably by interrupting descending inhibitory input to a brainstem "crying centre".

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