LETTERS TO THE EDITOR

Febrile Hashimoto's encephalopathy

Hashimoto's encephalopathy is a rare association of Hashimoto's thyroiditis. The clinical course is often characterised by relapses.¹⁻² Persistent fever has not been previously recorded in the literature.

A 48 year old white woman was referred in April 1996 from a psychiatric clinic, where she had presented with psychotic episodes with gradual deterioration and been diagnosed as possibly having Creutzfeldt-Jakob disease. She was confused and had brisk tendon reflexes, an extensor left plantar response, bilateral moderate cogwheeling, action myoclonus, and nuchal rigidity. The gait was unstable and uncoordinated.

Puerperal psychosis occurred at the age of 22 and from the age of 30 she had periodic mild confusional states of about a week long. Hashimoto's thyroiditis was diagnosed 10 years before the neurological admission and since then the patient had been euthyroid on thyroxine replacement, although confusional states appeared periodically, roughly every 3-4 months.

After admission the clinical features changed, with variable stupor, generalised seizures and myoclonus, incontinence, and anorexia. There was a persistent fever which varied between 37.7 and 39°C, with little or no response to antipyretic drugs. Antiepileptic drugs achieved some control of the seizures.

Normal or negative laboratory tests included blood count, CSF analysis, erythrocyte sedimentation rate, liver and kidney function tests, coagulation screen, serum and CSF electrophoresis, immunoelectrophoresis, ANCA, anti-DNA, ASMA, antimitochondrial, antiphospholipid, and antinuclear antibodies, C reactive protein, tumour markers, serum copper, ceruloplasmin, serum electrolytes, vitamin B12 and folic acid, serological tests for HIV, HSV, HBV, CMV, syphilis, Widal, Wright, and PPD. Blood, CSF, and urine cultures were negative. The EEG showed diffuse slow activity. Brain CT and MRI were normal. T3, T4, TSH, FT3, and FT4, were normal but the titre of autoantibodies to thyroid peroxidase (Anti-TPO) was increased at 7000 mIU/l (normal value <60 mIU/l).

Treatment with prednisolone (70 mg/day) led to gradual improvement and the fever ceased promptly. Within 2 months the patient had fully recovered with only a residual external (left) plantar response. Decrease of the Anti-TPO (1000 mIU/l) titre paralleled the clinical improvement. The patient was discharged after 4 months having made a good recovery on 40 mg/day prednisolone and had no relapses during 3 subsequent years (maintenance prednisolone dose 15 mg every other day, Anti-TPO titre within normal range).

The clinical features in this case are compatible with a diagnosis of Hashimoto's encephalopathy with an associated fever. Both fever and Anti-TPO concentrations were suppressed with prednisolone treatment. Increase of TNF- α concentration is seen in

the serum of patients with autoimmune thyroiditis³ and this is known to act as an endogenous pyrogen.⁴ This mechanism may explain the persistent fever as a prominent symptom in our case.

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Devic's neuromyelitis optica and HIV-1 infection

Neuromyelitis optica (Devic's syndrome) can be defined as a severe transverse myelitis, an acute unilateral or bilateral optic neuropathy, no clinical involvement beyond the spinal cord or optic nerves, and a monophasic or, rarely, a multiphasic illness.¹ Optic neuritis precedes transverse myelitis in 80% of cases, by less than 3 months in most cases. Neuromyelitis optica may occur as a result of demyelination from multiple sclerosis, but many differences have to be noted between neuromyelitis optica and multiple sclerosis. Neuromyelitis optica is not predominantly seen in the white ethnic group. Severe deficits after the acute episode are more frequent in neuromyelitis optica. Multiple sclerosis presenting as transverse myelitis is rare.² Oligoclonal bands in the CSF and white matter lesions on brain MRI are rare in neuromyelitis optica compared with over 90% of patients with definite multiple sclerosis. These two abnormalities can resolve in neuromyelitis optica but very rarely in multiple sclerosis. The high CSF protein concentration and pleiocytosis seen in 60% of cases are often more important in neuromyelitis optica. Swelling and signal change on MRI extending beyond a single segment are more frequent in neuromyelitis optica and cavitation can be seen. Most authors consider that neuromyelitis optica is often a variant of postviral acute disseminated encephalomyelitis mostly due to varicella zoster virus, but other causes have been identified—for example, systemic lupus erythematosus, antiphospholipid syndrome, and pulmonary tuberculosis. To the best of our knowledge, only one case of neuromyelitis optica (followed by a bilateral acute retinal necrosis) due to varicella zoster virus has been described in a patient with AIDS.³ We report the case of an HIV infected African woman developing neuromyelitis optica.

A 41 year old woman from the Congo Democratic Republic was admitted to hospital in March 1999 because of a 10 day progressive blindness of the left eye with optic disc oedema. There were left leg dysaesthe-

sias of 1 month duration and a T6 left sensory level was noted within the next few days. Progressive ascending paralysis of both legs without bowel or bladder dysfunction developed. HIV infection had been diagnosed 11 years earlier. Her CD4 lymphocyte count was 499/mm³ and her plasma viral load was 3.06 log (1150 RNA/ml). She received no treatment. Examination of CSF showed 57 white blood cells/mm³ (94% lymphocytes). Total protein concentration was 48 mg/dl. No oligoclonal bands were detected. Serology for syphilis and *Borrelia burgdorferi* and *Cryptococcus neoformans* antigen were negative in blood and CSF. Search for herpes simplex virus type 1 and 2, varicella zoster virus, cytomegalovirus, and Epstein-Barr virus by polymerase chain reaction remained negative in CSF. Serology for HTLV1 was negative. Interferon and angiotensin converting enzyme concentrations were normal in blood and CSF. Cultures of CSF were negative. Antinuclear and anticardiolipin antibodies were not detected. Brain MRI with and without gadolinium DPTA was normal. Spinal cord MRI disclosed four high signal intensity lesions on the T2 weighted sequence on T6, T7, T10, and T12 levels. Enhancement over the four segments was seen after intravenous gadolinium DPTA. The patient received 0.5 g/day intravenous methylprednisolone for 3 days. Subsequently, she was maintained on oral prednisone (80 mg/day) for 12 days and treatment with lamivudine (300 mg/day), zidovudine (600 mg/day), and nelfinavir (2250 mg/day) was started. Symptoms dramatically improved. The patient was able to walk alone without help after 10 days. Visual acuity returned to normal. Dysaesthesias were absent 1 month later. Three months later, plasmatic viral load was under 50 RNA/ml. Examination of CSF and MRI of the spinal cord gave normal results. There was no recurrence after a 5 month follow up.

Optic neuritis occurring in patients infected with HIV⁴ is usually due to syphilis or to opportunistic organisms such as varicella zoster virus¹, cytomegalovirus, *Cryptococcus*, *Histoplasma*, *Bartonella*, *Toxoplasma*, or *Mycobacterium tuberculosis*. The role of HIV itself is now well established. Intramedullary involvement in the course of HIV infection may result from HIV itself (vacuolar myelopathy), coinfection with HTLV1 in endemic areas, or transverse myelitis mostly due to varicella zoster virus, cytomegalovirus, or syphilis. Such infections were excluded in our patients. There are only few reports of multiple sclerosis-like illness occurring with HIV infection.⁵ Cases of sarcoidosis associated with HIV infection remain exceptional even when highly active antiretroviral therapy (HAART) is used⁶ (optic neuritis is rare in sarcoidosis and no case of neurosarcoidosis has been described with AIDS). Primary CNS lymphoma associated with HIV is associated with Epstein-Barr virus and may cause optic neuritis or spinal cord involvement. We think that multiple sclerosis, sarcoidosis, and lymphoma were excluded in our patient. Neuromyelitis optica without an identified cause can be seen in the course of HIV infection, even at an early stage of the disease, before immunodepression occurs.

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Hemiageusia from an ipsilateral multiple sclerosis plaque at the midpontine tegmentum

The exact location of the pontine gustatory pathway has not yet been clarified, probably because there are so few studies of taste in patients with well localised brainstem lesions.¹⁻³ Here we report on a patient with isolated hemiageusia and trigeminal sensory neuropathy from a single small pontine lesion.

A 46 year old woman experienced a burning sensation on the left side of the tongue. The next day she discovered a loss of taste on the entire left half of her tongue and numbness on the left side of her face. Neurological examination was normal except for hypaesthesia to pain, touch, and temperature sensation in all three divisions of the left trigeminal nerve, with decreased left corneal reflex and no weakness of masseters. Taste sensation was tested using separate solutions of 1.2 M NaCl, 0.47 M glucose, 0.17 M citric acid, and 2.52 mM quinine HCl. Marked disturbance on the left side involving the anterior two thirds and posterior one third of the tongue was noted with all four substances. Routine laboratory tests were normal. Analysis of CSF showed 8 lymphocytes/mm³, 46 mg/dl protein, increased quantitative intrathecal IgG, and oligoclonal bands. Visual evoked potentials, brainstem auditory evoked potentials, and somatosensory evoked potentials after both median and posterior tibial nerve stimulation were normal. The blink reflex was normal on stimulation on the right,

and when stimulated on the left, there was no R1 component and R2 responses were elicited normally. Masseter reflex was absent on the left side. Brain MRI demonstrated multiple bilateral hyperintense white matter signals in periventricular distribution on T2 weighted images, and a hyperintense small lesion in the lateral part of the left midpontine tegmentum that showed enhancement after gadolinium injection (figure A and B). After intravenous high dose methylprednisolone therapy there was no immediate improvement of the patient's neurological symptoms. In the follow up MRI after 3 months, the gadolinium enhanced pontine image had disappeared, the blink and masseter reflexes were normal, and trigeminal neuropathy and the taste disturbance had gradually reduced.

The present case supports the finding that unilateral pontine lesions result in ipsilateral gustatory deficits, suggesting that gustatory fibres ascend from the solitary nucleus in the medulla up to the homolateral pontine tegmentum without decussating.¹⁻³ Lower midbrain level decussation is supported by a recent report.³ Topography of gustatory pontine fibres has been discussed in cases with taste disturbance,^{1,3} and, as found here, the absence of limb sensory involvement with normal somatosensory evoked responses contradicts the widely accepted notion that the pontine medial lemniscus conveys taste. On the other hand, Uesaka *et al*² described a patient who presented with ageusia and ipsilateral truncal ataxia presumably due to brachium conjunctivum involvement, and, therefore, it was suggested that the adjacent parabrachial nucleus might constitute a pontine taste area. Our patient developed left sided hemiageusia and trigeminal sensory disturbance, and electrical stimulation on the left elicited no R1 response and absence of masseter reflex. These electrophysiological abnormalities imply ipsilateral brainstem lesions at the trigeminal principal sensory and motor nuclei, respectively.⁴ Moreover, MRI confirmed the existence of a new gadolinium-enhanced demyelinating lesion in the left midpontine tegmentum. The precise correlation between our patient's symptoms and the electrophysiological and MRI abnormalities indicates that the involvement of the central tegmental tract, which is the anatomical structure adjacent to the sensory and motor trigeminal nuclei at the midpontine level (figure C), is probably important in causing taste disturbance. According to this hypothesis, Norgren⁵ showed in primates that axons of neurons located in the solitary nucleus ascend in the central tegmental tract to the ventroposteromedial nucleus of the thalamus

without terminating first in the pontine parabrachial nucleus.

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Use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis

We report the use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis. Paroxysmal cases of kinesigenic choreoathetosis were first described in 1962 by Lishman *et al*¹ and the term "paroxysmal kinesigenic choreoathetosis" was first coined by Kertesz² in 1967. Recently, a more universal and potentially useful classification of these disorders has been proposed amending the terminology to "paroxysmal kinesigenic dyskinesia".³ However, we have used the more familiar terminology for our patient as it is more precise. The clinical features of paroxysmal kinesigenic choreoathetosis have been reviewed by Marsden and Luders.⁴

There is a male preponderance of the condition with about 50% of cases inherited as an autosomal dominant trait, the rest being sporadic. Attacks are precipitated by sudden movements or startle and may be unilateral, bilateral, or affect alternate sides. Often the cause is idiopathic but a few cases have been attributed to multiple sclerosis. There has been no clear evidence for seizure activity in this disorder even though the condition is very responsive to antiepileptic drugs such as phenytoin and carbamazepine.⁴

A 13 year old boy of unrelated parents and with no family history of neurological disorder presented with a 6 month history of muscle spasms affecting the left side of his body. Close questioning indicated that his symptoms were brought on during the initiation of sudden movements (such as starting

Gadolinium enhanced T1 weighted MRI of coronal (A) and axial (B) sections showing a high intensity lesion in the lateral portion of the left midpontine tegmentum. Diagram (C) showing the area clinically affected (shaded area). ML=medial lemniscus; CTT=central tegmental tract; Vs=trigeminal principal sensory nucleus; Vm=trigeminal motor nucleus.

to run) and manifested as spasm of the left side of the face and flexor spasm of the left arm and left leg. There were no premonitory symptoms and consciousness was normal during the attack. Attacks lasted for a few seconds only and he felt perfectly well afterwards. Neurological examination was normal. Brain MRI was normal and three EEGs performed over the course of the next year were all normal. There were no biochemical or haematological abnormalities; copper studies were normal. A diagnosis of paroxysmal kinesigenic choreoathetosis was made. He was allergic to carbamazepine and did not respond to sodium valproate.

Lamotrigine (3,5-diamino-6-[2,3-dichlorophenyl]-1,2,4-triazine) is a use dependent blocker of voltage gated sodium channels and has a similar anticonvulsant profile to phenytoin and carbamazepine in animal models.⁵ Lamotrigine at a dose of 50 mg twice daily, completely abolished the involuntary movements. On two occasions, these returned on stopping lamotrigine and were abolished by reinstituting treatment. Our patient has taken this for over 2 years without ill effect. He now only experiences symptoms of paroxysmal kinesigenic choreoathetosis if he omits his medication. The response to lamotrigine may provide insight into the pathogenesis of the disease, suggesting that it may be caused by an ion channel defect; these are known to be responsible for some paroxysmal neurological conditions.⁶ As far as we are aware, this is the first report of the successful use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis.

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Chronic inflammatory demyelinating polyneuropathy as a complication of cat scratch disease

Cat scratch disease (CSD) was first described in 1950 as a benign regional lymphadenitis. This infection is caused by *Bartonella henselae*. The clinical range of CSD has expanded beyond the classic presentation. In 5%–20% of the infected patients the disease may spread to other organs. However, neurologi-

cal complications associated with CSD are rare, with encephalopathy being by far the most common form (90%) of nervous system involvement. Encephalopathy occurs in 2%–3% of patients and is more common in adults than in children with the onset varying from a few days to months after diagnosis of CSD.¹ Other known neurological manifestations, often in combination with encephalopathy, are neuroretinitis, oculoglandular disease of Parinaud, myelopathy, radiculopathy or abducens nerve, and facial nerve paresis. We report on a 3 year old boy who developed chronic inflammatory demyelinating polyneuropathy (CIDP) 6 weeks after identification of CSD.

A previously healthy 3 year old boy presented in a paediatric clinic with regional lymphadenitis of the submandibular and suboccipital glands. He was afebrile and did not have any other symptoms. There was no hepatosplenomegaly. No neurological signs were present on physical examination. As the boy might have been scratched by one of his pet cats, in accordance with a small cut on his scalp, CSD was suspected. It was serologically confirmed (enzyme linked immunosorbent assay (ELISA) *B. henselae* IgG > 850 U/l; IgM > 250 U/l) and he was treated with clarithromycin.

Six weeks after the onset of CSD, he showed difficulty in walking, inability to run or climb stairs, and frequent falls. Slowly progressive pain and a gait disorder were noted over a course of a further 8 weeks. On admission to our department there were no general signs and only a small submandibular lymph node remained. Mental state and cranial nerve examination were normal. He showed a symmetric distal muscle weakness in all limbs. A marked sensory ataxia was noted. We found mild root pain on a straight leg raising test. Deep tendon reflexes were very low to absent, especially in the legs. Plantar responses were flexor.

Serum concentrations of liver enzymes and glucose, and a protein spectrum were normal. Laboratory tests for adeno, RS, corona, influenza 1–2–3, parainfluenza 1–2–3, sendai, mumps, measles, herpes simplex and varicella zoster viruses, *Mycoplasma pneumoniae*, *Chlamydia psittaci*, *Coxiella burnetii*, *Mycobacterium tuberculosis* and atypical mycobacteria were negative. Antinuclear factor and ANCA were negative. Levels of complement factor 3 and 4 were normal. ELISA *B. henselae* IgG in serum, 3.5 months after outbreak of lymphadenitis, was still >850 U/l. IgM specific

antibodies now were <200 U/l. Examination of CSF showed 13 mononuclear leucocytes/ μ l, no polynuclear leucocytes, raised protein concentration (558 mg/l, normally 160–310 mg/l), three oligoclonal bands in CSF on isoelectric focusing, and a slight intrathecal IgG synthesis (51 mg/l, normally 3.2–15 mg/l). IgG and IgM specific antibodies against *B. henselae* were negative in two CSF samples. Polymerase chain reaction (PCR) on *B. henselae* in CSF and in serum was negative. Enolase, myelin basic protein, S100, lactate and glucose concentrations in CSF were normal.

Electroencephalography, cranial CT, and gadolinium enhanced MRI of the thoracolumbar region were normal. Nerve conduction studies showed a marked decrease of motor nerve conduction velocities in all limbs (median nerve 20 m/s). In sensory conduction studies no response could be elicited. The EMG was normal.

The sural nerve biopsy showed many demyelinated axons and signs of early remyelination, but no onion bulbs (figure A). The density of myelinated fibres was slightly below normal, which could be attributed partly to intrafascicular oedema. Myelinated fibre size histogram was bimodal. There were no endoneurial infiltrates, or signs of vasculitis. Electron microscopy showed macrophage induced demyelination (figure B). Polymerase chain reaction (PCR) on *B. henselae* in sural nerve fragments was negative.

The history and clinical presentation, combined with CSF findings and the results of EMG and light and electron microscopy studies, are compatible with the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

He was treated with prednisone (15 mg every other day for 1 month) and the dose was reduced slowly in the course of 4 months. His motor and sensory function recovered completely and deep tendon reflexes reappeared. Nerve conduction studies 1 year after the onset of CSD were normal.

Neurological complications of CSD are rare and predominantly of the CNS. Myelitis has been described in combination with encephalopathy² and radiculopathy was reported only in combination with encephalomyelitis. Ophthalmological problems are oculoglandular disease of Parinaud³ and neuroretinitis.⁴ In a study by Carithers and Margileth of 76 patients with CSD and neurological complications, 15 patients showed signs of dysfunction of cranial or

(A) Sural nerve. Low power electron micrograph. Demyelinated fibres (arrows) and thinly remyelinated fibres. Bar=2.0 μ m. (B) Sural nerve; detail of myelinated fibre. Macrophage has invaded the myelinated fibre; process of macrophage is extending between axon and myelin sheath (arrow head). The damaged inner major dense line is terminating as a small dark swelling (arrow). Bar=0.2 μ m.

peripheral nerves. Ten patients had neuroretinitis, two children had paresis of the facial nerve, and three adult women complained of neuralgia.⁷ One case study presented a peripheral facial nerve paralysis as a complication of CSD.⁸

Up until now CIDP has never been reported as a neurological complication of CSD.

Given the history and clinical course, the electrophysiological and nerve biopsy findings, coupled with the strongly positive serology to *B henselae*, we think that the CIDP in this patient is a direct complication of CSD. CIDP is an autoimmune process in which both humoral and cellular factors are thought to participate in the pathogenesis. Wheeler *et al* also suggested an immune response as a pathophysiological mechanism responsible for CSD encephalopathy.¹ In our patient a delayed myelin destruction is induced by sensitised macrophages, originally activated by the Bartonella infection. Therefore, we hypothesise that the pathophysiology of both central and peripheral nervous system complications after a CSD infection shares a similar immunological mechanism.

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CORRESPONDENCE

"Can't you use another vaccine"? Postrabies vaccination encephalitis

I read with interest the letter of Chau *et al* related to iatrogenic disseminated encephalomyelitis, in a man bitten "by his own apparently normal dog".¹ Whereas I appreciate the novelty of documenting such entities with brain MRI, I wish to question the reasoning behind the practice of antirabies vaccination in similar doubtful circumstances. I have commented on that issue in another occasion.² I have also documented instances of excessive enthusiasm, often encountered in situations such as those under discussion, in a research project conducted at the Pasteur Institute in Tehran, Iran.³ There is no justification for vaccinating a person

simply because of fear when the remedy itself is to be feared even more, as documented by Chua *et al*.

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The authors reply:

We thank Derakhshan for his comments on our case report of iatrogenic disseminated encephalomyelitis after use of the suckling mouse brain postrabies exposure vaccination. We were not responsible for the postexposure vaccination, which was administered in a provincial hospital in central VietNam. However, in the circumstances (and in the absence of the human diploid cell tissue culture vaccine) we think that it was appropriate to use the vaccine in this case. The dog had previously been well behaved and it was highly uncharacteristic for it to bite its owner. After the event the dog disappeared into the forest and was not seen again. Hence, it was not possible to retrieve the brain for analysis, as is usual in most cases.

The mortality from rabies is essentially 100%, a figure that can be reduced dramatically by the expeditious use of the suckling mouse brain vaccine after exposure. We agree with Derakhshan's comments on excessive enthusiasm for any medication, and obviously the relative risks and potential benefits must always be balanced. In a disease with a 100% mortality, where a potentially effective treatment is associated with a severe side effect in only 1:27 000 cases it would seem reasonable to use the treatment. At this centre we vaccinate 2000 people every year after a dog bite, we see about 50 people a year die of rabies. We would therefore anticipate seeing a case of iatrogenic disseminated encephalomyelitis after use of the suckling mouse brain postrabies exposure vaccination once every 13.5 years. In the same period we would see 675 people dying from the disease.

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Sleep benefit in Parkinson's disease

We read with great interest the report by Bateman *et al* on sleep benefit in Parkinson's disease.¹ The authors state that no objective study has been undertaken about sleep benefit. We must object to that, as our group has recently published an extensive objective study about sleep benefit, which includes objective motor examinations, levodopa plasma concentration determinations, and polysomnographies.² Moreover, we would like to briefly discuss some of the authors' findings in the light of our own results.

The authors studied 20 patients with the motor part of the UPDRS, at waking (apparently twice), and after medication during "on" and "off". At each rating, the patients

completed an "activities of daily living" (ADL) questionnaire. The authors also administered ADL questionnaires to heterogeneous groups of outpatients.

In their results, the authors comment on six out of 16 patients of the first series: "When they awoke these patients performed as well as when they were 'on' due to medication". Unfortunately, they give no data at all, such as mean UPDRS scores and ranges, for baseline state and "on" time schedules and hours of the ratings. Furthermore, no information is provided to account for the remaining four patients who were included in the first series but do not appear in the results.

The findings by Bateman *et al* contrast with our own results, in that patients with sleep benefit performed only slightly better in the morning compared with those without. A clear "on" compared with baseline was found in our study both in patients with and without sleep benefit after intake of their regular medication. We concluded from our data that sleep benefit was much smaller than expected. A morning baseline function as good as a drug induced "on", as described by Bateman *et al*, would be indeed a sleep benefit of considerable magnitude. On the other hand, a drug induced "on" similar to the morning baseline state could theoretically also point to an insufficiently drug treated patient group. In any case, as no data are given to allow comparisons, one is confined to speculate why sleep benefit could be so much greater in British patients than the Argentinian population, where sleep benefit, although objectively existing, was quite a subtle phenomenon.

The authors further state that a "strong correlation" was found between ADL and UPDRS, and conclude that ADL may serve as a "more objective" instrument to measure sleep benefit. Unfortunately they do not indicate if the correlation was found at any point in time or if all evaluations were lumped together, as no correlation index or graph is given.

In the second sample of the study, 113 patients completed an ADL questionnaire at three points in time (at waking, best, and worst) before any drug intake. This was done at home. The authors determined that sleep benefit was present when the mean ADL score difference between best and worst was more than 12—that is, when strong variations occurred in baseline score before medication. The validity of this arbitrary definition deserves some discussion. Firstly, to take this variation as a criterion for sleep benefit may lead to a confusion with motor fluctuations. As the ADL score has a maximum of 52 points, an absolute score difference of 12 as a prerequisite for sleep benefit will lead to the exclusion of patients with smaller fluctuations irrespective of sleep benefit. So their own definition could have biased the authors' finding that patients with sleep benefit had a younger disease onset, longer disease duration, and more frequent use of bromocriptine. All this might also occur in a fluctuating subgroup of patients and an association of sleep benefit with fluctuations has been previously described.³

Secondly, it is necessary to be cautious in considering ADL questionnaires as an objective measure to determine the presence of sleep benefit. The patient rating is based on how well he thinks he could perform at a given moment, and as we pointed out in our study, a large difference between self perceived motor function and objective motor function may occur in sleep benefit.

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

MRI of lower brain and spinal cord.

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

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 clinico-pathological 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 serotoninergic 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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in the coming years and this is to be welcomed.

Finally, it is worth pointing out that the computer system requirements are quite high to run the video adequately. Preferably a modern Pentium machine is required with at least 16 Mb of Ram and obviously a multimedia configuration with a high quality sound card.

Overall, a novel approach to an important topic and one to be recommended.

MICHAEL P BARNES

McAlpine's Multiple Sclerosis. Third edition. By ALISTAIR COMPSTON, GEORGE EBERS, HANS LASSMANN, IAN MACDONALD, BRYAN MATTHEWS, and HARTMUT WEKERLE. (Pp 562, £99.00.) Published by Churchill Livingstone, London, 1996. ISBN 0443-050082

Those familiar with previous incarnations of *McAlpine's Multiple Sclerosis* will know that authors have changed from one edition to the next and with that the emphasis has changed as well. This third edition is no exception and it is almost entirely a new book. It has six outstanding authors, each bringing formidable authority to bear on their own and their colleagues' contributions. The book is big and aims at a more comprehensive coverage of the subject than previous editions. It is, however, no mere indiscriminate assemblage of the facts, but very much reflects the views, indeed the vision (individual and collective), which the authors have of the pathogenesis and natural history and epidemiology of multiple sclerosis.

Professor Alastair Compston has the lion's share of the writing, and is in his element on epidemiology and genetics and the cellular biology of neuronal and glial cells. He starts the book with an absorbing and scholarly historical piece.

Far from resting on his laurels Professor Bryan Matthews has extensively rewritten and updated his chapters on symptoms and signs and differential diagnosis.

Magisterial is a word Compston is fond of using to describe the achievements of luminaries from the past and it could reasonably be applied to Professor Ian McDonald's professional lifetime of laboratory and clinical research which is reflected in his chapters on pathophysiology and diagnostic methods and investigations.

There are stimulating and provocative chapters on the natural history and on neuropathology. Arguably we all need to try to understand the immunology of multiple sclerosis, particularly in the current era of therapeutic strategies aimed at modifying the course of the disease. Clinicians may think that a chapter on animal models of multiple sclerosis will not be of immediate relevance to their day to day work, and may think that more space could have been apportioned to some aspects of symptomatic treatment and the management and rehabilitation of patients with moderate or severe disability.

This is a very fine book; instructive, edifying, and enjoyable, chiefly because of the writing but also because of the quality of the print and the abundance of excellently reproduced illustrations.

RODNEY WALKER

MR Imaging of the Brachial Plexus. By H W VAN ES. (Pp 131.) Published by

Febodruk B V, Utrecht, The Netherlands. ISBN 90-393-1429-2.

This book is a good description of the normal anatomy and imaging findings of some of the most common clinical conditions affecting the brachial plexus. I am sure that it will be useful for neuroradiologists and general radiologists confronted with clinical questions about brachial plexus pathology.

The book summarises the author's own experience during 5 years and includes about 230 MRI studies performed in patients with suspected brachial plexus pathology. The book is overall well written, in simple and concise language. Tables and good quality figures are well explained, and contain interesting information. The number and quality of the references are adequate. The book is divided into six chapters. This division results in a well balanced book in general, although some chapters may be too extensive when one considers the more limited author's own experience. The anatomical description of the brachial plexus in chapter 2 is one of the best depicted parts of the book, and I am sure that it will be very useful for radiologists. On the contrary, the section dealing with imaging techniques is somewhat long and confusing. Some studies comparing the efficacy of different techniques are missing, something which I am sure would have enhanced the work significantly, and at the end of this section, the reader does not really know which imaging techniques to choose for a simple examination of the brachial plexus. I was more than pleased that in the chapter on trauma and the thoracic outlet syndrome that the author's experience agrees with my own of MRI not being very helpful here. There is a chapter on radiation induced brachial plexopathy, not an infrequent disorder in hospitals with large oncology units, but only three cases are presented. The last chapter deals with inflammatory conditions such as multifocal motor neuropathy and chronic inflammatory demyelinating polyradiculoneuropathy, and the ability of MRI to differentiate them from others having a similar clinical presentation (lower motor neuron disease). In my opinion, this is probably the most interesting chapter of the book, as it covers a novel subject.

BEATRIZ GOMEZ-ANSON

A Textbook of SPECT in Neurology and Psychiatry. Edited by PETER P DE DEYN, R A DIERCKX, A ALAVI, AND B A PICKUT. (Pp 580, £95.00.) Published by John Libbey and Co, London, 1997. ISBN 0-86196-542-6.

The editors claim to provide a "state of the art review of the role of SPECT in neurology and psychiatry". The 64 chapters are divided into 11 sections, covering dementia, neuropsychology, psychiatry, movement disorders, epilepsy, paediatrics, cerebrovascular disease, tumours, trauma, specific applications, and physical techniques. Although the emphasis is on SPECT, the role of PET is also mentioned. Most chapters represent the results of the authors' research and experience; unfortunately this piecemeal approach does not lend itself to a cohesive text. Although most sections commence with a brief review it would have been conducive for the reader if these reviews were more comprehensive, this could then have provided the entree for the subsequent detailed studies.

BOOK REVIEWS

Botulinum Toxin in the Treatment of Cerebral Palsy. Edited by WERNER POEWE and JÖRG WISSELL. (CD Rom) Published by Blackwell Science, Austria, 1997. ISBN 3-89412-364-8.

It was a pleasure to review this CD Rom. It is certainly a sign of changing times that the book review section now also contains reviews of CD Rom material. This is basically a review of the use of botulinum toxin in the treatment of cerebral palsy built around a meeting held in the Department of Neurology in Innsbruck in May 1997. The CD Rom contains the talks of eight speakers recorded at the time. While listening to the speaker's voice the slides come on and off the screen at the right moment. Thus, it is simply like being at the talk itself in Innsbruck. If this idea catches on then we may begin to see the end of international medical travel! The final section contains two videos regarding botulinum treatment of ankle clonus and dynamic equinus foot deformity as well as treatment of an unusual self mutilating condition. However, there is no explanation or sound with these videos and this section is thus rather a waste of time. I suspect this is a technical fault as the quality of the video pictures was very good but obviously needed some verbal context to make any sense.

The content of the previous sections mainly focuses on the use of botulinum toxin in cerebral palsy both in the treatment of upper and lower limb problems. Whilst the talks contained nothing new (and were now about 2 years out of date) they were certainly a comprehensive summary of the state of knowledge at that time. My main objection is that the talks were not edited and thus each talk averaged around 15 minutes; this amounted to considerable time sitting and listening. A certain amount of judicious editing and streamlining of the CD Rom would have been useful. Frankly it may have been better for the talks and the slides to be specifically written with a CD Rom in mind. However, this is a rather minor point and overall I found the CD Rom easy to use, instructive, and certainly a novel way of imparting information. I suspect that we shall see a lot more of this methodology

It is unfortunate that the first section on dementia, most probably the major indication for SPECT at present, lacks an overview. Furthermore, there is little mention of the OPTIMA project (Oxford prospective investigation into memory and ageing)—the largest neuroimaging study in the world with histological confirmation. Presumably, this is partially a reflection of the inherent delay in the production of such a wide ranging text.

In essence this is a reference text, and despite the foregoing criticisms, this book does fulfil a niche in the literature. Neuropsychiatrists, neurosurgeons, psychologists, and other relevant clinical specialists will find their appropriate sections to be extremely useful in demonstrating the contribution of PET/SPECT investigations and also showing the new areas of development. Trainees in nuclear medicine and neuroradiology would benefit by studying the review chapters. Certainly, for the nuclear medicine specialists with an interest in neuroimaging, this text is an essential acquisition.

PAUL KEMP

Challenge Epilepsy—New Antiepileptic Drugs. Edited by STEFAN, KRAMER AND MAMOLI. (Pp258 £39.40.) Published by Blackwell Science, Oxford 1998. ISBN 3894123850.

This multiauthor review is based on a symposium on new antiepileptic drugs (AEDs). The book suffers from the usual difficulties with symposia publications including too many chapters which are often too short to usefully summarise the topic in detail but with much repetition and a lack of consistency and style. Nevertheless, excellent practical guidance is given in several chapters, including those on monotherapy, combination therapy, and the management of infants and children. For most patients with epilepsy none of the new AEDs are more effective than standard current first line treatment, with which we have many patient-years of experience. Marketing of the new drugs against these has leant heavily on negative campaigning (teratogenicity and female hormonal problems with valproate). Attempts to measure various aspects of quality of life have become important and these (particularly cognitive side effects) are considered in chapters including those by Chadwick and Aldenkamp. It comes as no surprise to learn

that the single factor which most improves quality of life for patients is stopping seizures and most experience with these drugs has been obtained in that 20% of the epilepsy population with intractable seizure disorders. Treatment of this group of patients has been most improved after assessment by those with a special interest in epilepsy and identification of pseudoseizures, correct diagnosis of epilepsy syndromes requiring specific treatment, and identification of cases suitable for surgery. It is difficult to determine which of the new drugs are most beneficial in this treatment resistant group and as practical experience with the drugs in clinical practice is obtained it would be appropriate to design a randomised study comparing several of the new drugs against each other as add on or replacement therapy.

Further toxic effects of the new drugs are almost certain to be discovered. For example, at the time of the symposium the effect on visual fields of treatment with vigabatrin was not widely recognised and this is not covered in the text.

STEVE WROE

Advances in Multiple Sclerosis—Clinical Research and Therapy. Edited by STEN FREDRIKSON and HANS LINK. (Pp 232, £65.00.) Published by Martin Dunitz, London, 1999. ISBN 1-85317-871-3.

This is an interesting, well written, and useful book—but will not sell many copies. Regrettably it seems pitched at various different audiences and thus I suspect it will not greatly appeal to any particular group. Nevertheless I would thoroughly recommend this book. It contains some excellent descriptions of complex subjects and should be of considerable interest to all practitioners working within the field of multiple sclerosis.

The first part provides an overview of genetics. The first chapter is entitled "What the specialist in multiple sclerosis needs to know about genetics". It is an excellent overview and easily understandable by the non-specialist. The second part of the book covers the field of immunology. Some of the chapters in this section are rather short and of somewhat patchy quality but nevertheless still provide the non-specialist with a good summary of our present state of knowledge. The chapter, for example, by Neil Scolding on oligodendrocyte injury and the role of

complement provides a good update on the subject. The next chapter on cytokines is a little less clear to the non-specialist but nevertheless is useful. The following chapters in this section then become rather too specific to be of use to the non-specialist and probably too superficial to provide any new information to the specialist in the field. The third section covers MRI and once again the same pattern predominates: good updates in the subject but I am uncertain as to whether they are directed to the specialist or non-specialist. The chapter on diffusion MRI is an exception and is beautifully written. The fourth section discusses therapy. John Noseworthy provides his usual excellent contribution on emerging therapeutic options in multiple sclerosis but the chapters on gene therapy and the rationale for antiviral therapies are of somewhat less interest to the clinician.

In the final part(5) the book broadens out even further and discusses various aspects of the organisation of multiple sclerosis care. Personally I found this section of considerable interest but I suspect that those at the more scientific end of the multiple sclerosis spectrum will not find this section very enthralling. However, more scientific colleagues should certainly read the two excellent chapters by Alan Thompson and Jeremy Hobart on advances in multiple sclerosis rehabilitation and an update on outcome measurements.

Overall this book tries to be everything to everybody and thus probably fails in the market place. However, it would be a pity if some of the excellent chapters were lost to a wider audience—so buy it anyway.

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CORRECTION

Thwaites G, Chau TTH, Mai NTH, Drobniowski Fi, McAdam K, Farrar J. Tuberculous meningitis. *J Neurol Neurosurg Psychiatry* 2000;68:289-99.

Page 292, para 2. During the editorial process, the sentence "The limit of detection on microscopy is 100 mycobacteria/ml" was altered. It should read "The limit of detection on microscopy is 10 000 mycobacteria/ml".