LETTERS TO
THE EDITOR

Creutzfeldt-Jakob disease presenting as complex partial status epilepticus: a report of two cases

Creutzfeldt-Jakob disease is a transmissible human spongiform encephalopathy which may be familial, iatrogenic, or sporadic. The clinical features include a rapidly progressive dementia with the patient retaining clear consciousness until the terminal stages of the disease. We report on two patients presenting with a rapidly declining level of consciousness, in whom the clinical picture and EEG were suggestive of complex partial status epilepticus.

The first patient was a 58 year old woman who was admitted to a psychiatric unit with a short history of mood disturbance, confusion, and unsteadiness. A provisional diagnosis of agitated depression was made and she was started on lorazepam. She then became unsteady on her feet and required support when walking. She had had occasional complex partial seizures for 30 years but at presentation was not taking any anticonvulsant drugs.

On examination, she appeared perplexed, tearful, and agitated, and was unable to give a coherent history. She was intermittently confused and her gait was ataxic. There were no other cerebellar signs. The rest of the neurological examination was unremarkable although limited by poor cooperation.

She became more withdrawn and uncommunicative with incontinence of urine. She would occasionally jump when sitting in a chair.

Brain CT and MRI were normal, as was her CSF. An EEG showed frequent, almost continuous variable amplitude sharp waves in all areas, although with a right sided emphasis, with a repetitive appearance up to 2 per second (figure). The record was thought to be in keeping with partial status epilepticus.

Her level of consciousness deteriorated despite intravenous valproate and phenytoin and she was referred to the intensive care unit for continuous EEG monitoring. On arrival, she was deeply unconscious and despite aggressive management of her presumed complex partial status she died 3 weeks later. Histology of the brain was diagnostic of the sporadic form of Creutzfeldt-Jakob disease.

The second patient was a 68 year old man who was admitted with a short history of confusion and inappropriate behaviour. He appeared not to recognise his family. Initially he was dysphasic and obtunded. His conscious level then deteriorated and he became mute with evidence of right sided weakness.

All investigations including contrast enhanced brain CT and CSF examination were normal. An EEG was reported as showing frequent bilateral epileptiform activity, but there was no improvement in his clinical state after a loading dose of intravenous phenytoin. A repeat EEG was dominated by periodic lateralised epileptiform discharges (PLEDs), more marked on the left side. A third EEG 3 weeks later again showed frequent predominantly left sided epileptiform discharges, which were attenuated by a bolus of intravenous diazepam but without any improvement in his clinical condition.

He was transferred to this hospital for artificial ventilation because of the concern that he was in complex partial status. On admission he was mute, his eyes were closed, and he flexed to pain on the left side only. Intermittent twitching of both sides at a rate of between 1 Hz–2 Hz was seen. Reflexes were brisk and symmetric. His right plantar response was extensor, his left flexor.

A further EEG 5 days later showed generalised bitemporal continuous periodic sharp waves occurring at a frequency of 1.3 Hz, at times in the form of biphasic or triphasic complexes. Myoclonic jerks occurred during the recording.

It was considered that overall these features were consistent with a diagnosis of Creutzfeldt-Jakob disease. His condition continued to deteriorate and he died 2 weeks later. A request for a postmortem examination was refused.

These two cases illustrate a previously unrecognised presentation of Creutzfeldt-Jakob disease, namely presumed complex partial status.

In the first case, the interpretation of the EEG findings was made more difficult by the patient's depressed conscious level and the previous history of complex partial seizures, albeit mild. The initial psychiatric presentation, with mood and behaviour disturbance, as well as fluctuating confusion, was compatible with complex partial status. The initial EEG report, suggestive of partial status epilepticus, prompted treatment, unsuccessfully, with anticonvulsant drugs and subsequent transfer for continuous EEG monitoring. This disclosed marked fluctuations, including discrete runs of rhythmic sharp waves that were considered to be electrographic seizures. Even after sustained burst suppression, the recording fluctuated between generalised periodic discharges and periods of relative inactivity within a matter of seconds.

In the second case, the patient developed focal seizures and PLEDs on the EEG. The initial recordings were suggestive of complex partial status, with asymmetric discharges abolished by diazepam but without any observable clinical change. Subsequent recordings were more characteristic of Creutzfeldt-Jakob disease, particularly as the patient had developed myoclonus. Although the electrographic changes were abolished by diazepam, suggesting seizure activity, the modification of both clinical and EEG activity in Creutzfeldt-Jakob disease by benzodiazepines has been reported giving rise to further confusion with epileptiform sharp wave activity. The focal nature of the patient's signs and the lateralisation on the EEG is well recognised in Creutzfeldt-Jakob disease as are...
periodic PLEDS, which are often associated with contralateral myoclonic jerks. 1,2

The two cases described here illustrate that a diagnosis of Creutzfeldt-Jakob disease should be considered where a rapid decrease in consciousness is accompanied by EEG changes apparently compatible with complex partial status. When there is a clinical suspicion of Creutzfeldt-Jakob disease, the ideal method of monitoring such patients is with continuous EEG recording, allowing documentation of rapid fluctuations. The present cases are atypical in that the progression from presentation to death was rapid, but they underline the fact that minute to minute changes in EEG rhythm, asymmetry, and electrographic responsiveness to benzodiazepines can all be seen in Creutzfeldt-Jakob disease.

Brain MRI of patient 1 at the age of 14.5 years. The T2 weighted image (TR 2000/TE 50) shows a high intensity signal in the periventricular white matter, involving the right centrum semiovale, with mass effect.

Childhood demyelinating diseases with a prolonged remitting course and their relation to Schilder’s disease: report of two cases

Schilder’s disease or myelinoclastic diffuse sclerosis is a rare acute or subacute demyelinating disorder which primarily affects children and young adults.1,2 We report the clinical and neuroradiological follow up of two boys affected by a demyelinating disease with a prolonged relapsing-remitting course, response to corticosteroids, and relatively good long-term prognosis.

The first patient presented at the age of 12 with a 2 month history of repeated episodes of headache and blurred vision followed by weakness of the left leg, lasting a few hours. Head CT and bilateral carotid angiography were normal. Two weeks later the left hemiparesis and headache recurred. T2 weighted images on brain MRI disclosed a hypersignal intensity in the right parieto-occipital white matter, involving the centrum semiovale, without a mass effect. Flash visual evoked stimuli elicited a decreased potential on the left side. Motor and sensory nerve conduction were normal. Corticosteroid treatment (prednisone (1mg/kg/day)) reversed the clinical symptoms. At the age of 14, the boy began to have daily headaches. Treatment with carbamazepine and cyclophosphamide improved the hemiparesis and controlled the seizures. Brain MRI showed that the mass effect had disappeared, leaving a right parietal, temporal, and occipital lesion. Follow up at the age of 19 showed that, except for a left visual field defect, the patient had a normal neurological and mental status. He is now receiving azaithiaprine (75 mg/day).

The second patient was admitted at the age of 4 because of the sudden onset of headache and vomiting with ataxia and drowsiness followed by generalised clonic seizures. Clinical examination on admission showed left hemiparesis, anisocoria (left>right), and dysarthria. Ocular fundoscopy was normal. Head CT disclosed a reduced right lateral ventricle and subarachnoid spaces and, 1 week later, a small hypodense area in the right periventricular white matter. A carotid angiogram was normal at age 5. The child had a second episode characterised by high fever, vomiting, sixth nerve bilateral paresis, dysarthria, trapezial ataxia, and stupor. Treatment with corticosterphin (35 units daily for 5 days, then every 48 hours for 20 days) induced a rapid clinical improvement. From the ages of 5 to 14 the child had yearly relapses characterised by the sudden onset of left hemiparesis with variable involvement of the cranial nerves and impairment of consciousness associated with inconsistent alteration of white matter on brain CT (widespread hypodensity in the right centrum semiovale with a mass effect on the right ventricle).

These attacks regressed spontaneously or after corticosteroid treatment. The last episode, at the age of 14, consisted of right sided paraparesis of the face and hand, right hemiparesis, dysarthria, and drowsiness. T2 weighted sequences on MRI disclosed multiple focal abnormalities in supratentorial white matter. Corticosteroid treatment induced marked clinical improvement. Follow up at the age of 18 detected only a bilateral parietocentral atrophy and hypometabolic saccades. Mental development was normal. Brain MRI showed that the white matter lesions had partly regressed.

In both patients the following investigations during and between attacks yielded normal findings: CSF examination (absence of oligoclonal bands), CSF lactate and pyruvate; extensive serological and CSF immune screen; extensive viral, bacterial, and parasitic serological and CSF tests; blood gases, electrolytes, blood ammonia, amino acids in plasma, urine and CSF, gas chromatography-mass spectrometer analysis of organic acid in urine, adrenal function, plasma C26/C22 fatty acid ratio, serum copper, plasma ceruloplasmin, pyruvate dehydrogenase complex and cytochrome c oxidase activity in skin fibroblasts; arylsulphatase A, β-galactosidase, β-glucosidase, and galactocerebroside-β-galactosidase activities in leucocytes.

Both the overwarchs also raised the question of myelinoclastic diffuse sclerosis.1,2 A prerequisite for the diagnosis is a normal very long chain fatty acid plasma concentration. Clinical signs include an intracranial hypertension syndrome, mental deterioration, hemiplegia, and visual field defects. The disease has either a monophasic course, rarely rapid and fatal, or a relapsing-remitting course. Most patients have neurological sequelae during follow up, few patients fully recover.1 Histological studies typically show a demyelinating process similar to that of multiple sclerosis, with an inflammatory perivascular infiltrate, and in severe cases, cystic lesions. Neuroimaging findings tend to parallel the clinical course. Corticosteroids may improve the outcome of the single relapse and possibly of the disease, as they did in our patients. Some patients respond to immunosuppressive therapy.1,2 In both the patients described the association of headache, signs of diffuse and focal brain dysfunction, a relapsing course, and the response to corticosteroids also raise the possibility of an isolated CNS angiitis, a condition primarily affecting middle aged and elderly people. But neither cerebral angiography nor histological examination disclosed a primary vascular disorder. In addition the early onset and the spontaneous occurrence of the disorder rule out another recently described vasculopathy often associated with familial hemiplegic migraine.3

In conclusion, although demyelinating diseases that do not fulfill the classical definition of multiple sclerosis or encephalomyelitis remain difficult to label in children, the two cases we report here seem to fit Schilder’s description of myelinoclastic diffuse sclerosis. Owing to the current lack of knowledge on the causes of this disease strict diagnostic criteria cannot be applied. Some presentations may warrant brain biopsy. The differing clinical and neuroradiological features seen in these patients may help in delineating Schilder’s disease subtypes.

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Guillain-Barré syndrome after heat stroke

Heat stroke is usually not listed among the events triggering Guillain-Barré syndrome. Two cases of a Guillain-Barré syndrome-like polyneuropathy after heat stroke are on record, although without reference to polyneuropathy after heatstroke.

We report on a patient, who developed Guillain-Barré syndrome 10 days after severe heat stroke. He had electrophysiological evidence of demyelination, increased CSF protein, and high anti-GM1 antibodies. Heat stroke activates the immune system by cytokine release, opens the blood-nerve barrier, and exposes peripheral nerve antigens and thus may induce Guillain-Barré syndrome, as suggested by results from our patients.

A 28 year old drug addict was using anticholinergic drugs against sweating during levomethadon withdrawal. He was found unresponsive in a public garden on a hot summer day (ambient temperature 32 °C) after ingesting cocaine. His core temperature was 42.5 °C at admission. He was in deep coma with unreactive pupils and without corneal and pharyngeal reflexes. Tachypnoea had induced hypocapnia. Blood pressure was 85/50 mm Hg and heart rate was 165/min. He developed disseminated intravascular coagulation, thrombopenia below 40 000 mmol/l, and metabolic acidosis. Creatine kinase rose from 128 to 751 U/l. Leucocytes and C reactive protein remained normal. After 4 days of coma he was transferred to a closed psychiatry ward because of agitation and frightening hallucinations. After 5 more days he complained of fatigue, myalgia, and arthralgia and, 3 days later, developed fever (38.9 °C), tetrapilegia, and dysphagia. He required intensive care within hours. Vital capacity was 1.5 l. Proximal arm muscles had MRC grades between 2 and 3. All other limb muscles had grades 1 or 2. Facial muscle weakness increased for 2 more days. There was no ophthalmoplegia, but areflexia and stockling glove hypalgesia for vibratory and cold stimuli. He did not respond to early intravenous immunoglobulin treatment and underwent plasma exchange from day 14 to 18. Bulbar muscles improved on day 15. Head movements improved 1 week later. Minimal hand functions recurred after 4 more weeks. He still required help with dressing and was unable to stand 14 months after disease onset.

Protein concentration in CSF was 480 mg/l 2 days after onset of tetraparesis, and 7300 mg/l 3 weeks later. Cell count was 6 cell/mm³. He had high IgM (500 U/l), enzyme linked immunosorbent assay (ELISA) normal below 120 U/l but only moderately increased IgA antibodies against GM1. Only one of 20 patients with Guillain-Barré syndrome examined in the same laboratory had higher anti-GM1 IgM antibodies. The anti-GM1 antibodies were normal 8 weeks after plasmapheresis.

Compound motor action potentials were <0.7 mV in all tested nerves from day 2 to day 95. Distal latencies were more than 150% above the upper limit of normal in the left peroneal nerve. Conduction velocities were below 70% of the lower limit of normal in the left peroneal and the left median nerve. F latency was above 150% of the upper limit of normal in the peroneal nerve. F responses were missing in both median nerves and in the right peroneal nerve. A conduction block was present along the right ulnar nerve (wrist stimulation 0.69 mV; plexus stimulation 0.37 mV). Abnormal temporal dispersion and possible conduction block was present in the left ulnar nerve (wrist stimulation amplitude 0.36 mV; duration 8.6 ms; elbow stimulation amplitude 0.19 mV; duration 10.2 ms). Median sensory nerve conduction was normal and sural nerve conduction was moderately slowed (36 m/s) at day 2. Needle EMG disclosed abundant fibrillations and positive sharp waves in proximal and distal limb muscles at day 95.

Decreased sweating due to anticholinergic medication, cocaine induced increased heat production, and high ambient temperature precipitated heat stroke in our patient. Ten days afterwards he developed an acute neuropathy that met clinical and neurophysiological criteria for Guillain-Barré syndrome. Similar time delays have been seen in two other patients with Guillain-Barré syndrome-like neuropathies after heatstroke and in the second of two patients reported as critical illness neuropathy after hyperpyrexia. This patient had increased CSF protein and fasciculations which are unusual in critical illness neuropathy. He may have had Guillain-Barré syndrome as well. Weakness evolved with delay in these four patients with Guillain-Barré syndrome-like neuropathies, whereas it was present immediately after hyperpyrexia in five more patients, who probably did not have Guillain-Barré syndrome. One patient with heat stroke was tetraparetic when he regained consciousness. He had pyramidal and cerebellar signs and persistent atrophic weakness due to axonal or motor neuron loss and no neurophysiological evidence for demyelination. Four of 14 patients with cancer exposed to whole body hyperthermia and chemotherapy complained of weakness immediately after hyperthermia. Their nerve conduction abnormalities are reported as “compatible with scattered demyelination”. Our patient had chronic HCV infection which may be associated with vasculitic neuropathy and cryoglobulinaemia, both absent in our patient. A connection between Guillain-Barré syndrome and non-A non-B hepatitis is suggested, but the close temporal relation makes heat stroke a more probable cause of the disease in our patient. His high anti-GM1 antibodies suggested immune mediation. Anti-GM1 IgA is increased after Campylobacter jejuni infection. Heat stroke disrupts the gastrointestinal mucosal wall. Endotoxins enter circulation and stimulate macrophages, which release TNF-α, IL-1, IL-6, and IFN-γ. All these cytokines are raised after heat stroke and open the blood-nerve barrier. This may have exposed the GM1 epitope in our patient. IFN-γ induces Schwann cells to express MHC class II gene product, inviting T cell attack. TNF-α is proinflammatory and toxic, and increased in Guillain-Barré syndrome.

Guillain-Barré syndrome-like neuropathies have been reported from heatstroke (17), where heat stroke is common, but they were not noted in connection with epidemic heat stroke in North America. Our patient had all features associated with fatal heat stroke: long lasting coma, shock requiring vasoactive catecholamines, metabolic acidosis, and disseminated intravascular coagulation. Guillain-Barré syndrome may occur more often after heat stroke, if more patients survive hyperthermia thanks to intensive care.
also reduce clinical complications related to overdrainage in the upright body position. It prevents excessive CSF drainage by instantaneously increasing its hydrodynamic resistance when the drainage rate rises. The new Codman SiphonGuard device is intended to reduce the drainage rate when the flow dramatically increases during transition from a horizontal to vertical body position. It consists of two passages for the CSF drainage. In the central, wide channel a ball is inserted. The valve, unlike in all hydrocephalus shunts, is normally open and closes when the flow rate exceeds the specific threshold level. Then the drainage of CSF is diverted to a much thinner channel, which constitutes a high hydrodynamic resistance. This action may help to prevent posture related overdrainage.

We tested a sample of three SiphonGuards (kindly provided by Johnson and Johnson) in the United Kingdom Shunt Evaluation Laboratory to characterise the hydrodynamic performance of the device and its ability to reduce posture related overdrainage.

The pressure flow performance curve consisted of two straight lines of different slopes, both crossing the origin. They represent the two possible states of the SiphonGuard—low resistance (mean of 1.5 mm Hg/ml/min) and high resistance (mean of 42 mm Hg/ml/min, figure A). The differential pressures resulting from the above values, providing the CSF flow is on average 0.3 ml/min in the horizontal body position, would be 0.45 mm Hg and 12.6 mm Hg respectively.

Switching between low and high resistance was initiated by a flow rate, the threshold of which varied between 0.7 and 1.8 ml/min (figure B).

Switching from the high to low resistance was initiated by the differential pressure decreasing below the threshold from 4 to 6 mm Hg.

Overall, the mechanism of the SiphonGuard seemed to work according to the designers’ intention. It is supported by the concept that, during rapid transition from horizontal to vertical body position, initial flow rate increases above 2–3 ml/min. This is enough to switch the valve to the high resistance state, limiting overdrainage. However, in practice, it may not always be the case. In patients with small or slit ventricles previously having overdrainage, CSF may not be available to produce the flow at such a high rate. Moreover, because reliable switching occurs above 1.8 ml/min, in shorter persons or in patients resting persistently in a semisitting position (for example, elderly patients watching TV or reading books) the drainage may increase and decreasing perfusion rate (lower plot in µl/min) controlled by computer controlled infusion pump. Switching point may be demonstrated by an abrupt increase in the pressure measured across the device (upper plot). The same device changed the resistance state at variable flow rate from 0.7 to 1.5 ml/min.

(A) Pressure-flow performance curves for the SiphonGuard for the low and high resistance states. (B) Switching between low and high resistance states was monitored by a repetition of triangularly increasing and decreasing perfusion rate (inner plot in µl/min) controlled by computer controlled infusion pump. Switching point may be demonstrated by an abrupt increase in the pressure measured across the device (upper plot). The same device changed the resistance state at variable flow rate from 0.7 to 1.5 ml/min.

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Severe toxic neuropathy due to fibrates

The main adverse effects of lipid lowering agents in the fibrate family involve the gut, the skin, the liver, the blood, and the muscular system. Some of these complications are more frequent when renal failure exists. Here we report a case of neuropathy secondary to long term treatment with fenofibrate in a patient without renal failure taking recommended doses.

A 60 year old man was seen in September 1996 complaining of leg pain for 6 months. His relevant medical history included coronary artery disease treated for 10 years with 60 mg molsidomine/ day and 60 mg isosorbide dinitrate/ day, high blood pressure and hyperlipidaemia treated respectively with 100 mg atenolol/ day and 100 mg fenofibrate/day for the past 5 years. He complained of parasthesias along the posterior aspect of both thighs, later complicated by progressive muscle weakness.

The physical examination disclosed a patient incapable of standing on his toes or heels. No proximal muscle weakness was present. The deep tendon reflexes were reduced in all limbs. There was no sensory loss to light touch, vibrational sense, pain perception, and joint position sense. There was no disturbance of sphincter control or postural fainting and no impairment of potency to suggest dysautonomia. The rest of the physical examination was within normal limits. The EMG suggested an axonal sensorimotor neuropathy with reduced amplitude and conduction limits. The EMG suggested an axonal sensorimotor neuropathy with reduced amplitude and conduction limits. The EMG suggested an axonal sensorimotor neuropathy with reduced amplitude and conduction limits. The EMG suggested an axonal sensorimotor neuropathy with reduced amplitude and conduction limits.

The patient indicated an improvement in the past 5 years. He complained of paresthesiae of the skin, the liver, the blood, and the muscular system. Some of these complications are more frequent when renal failure exists. Here we report a case of neuropathy secondary to long term treatment with fenofibrate in a patient without renal failure taking recommended doses.

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High incidence and prevalence of multiple sclerosis in south east Scotland: evidence of a genetic predisposition

We read with interest the results of Rothwell and Charlton regarding the incidence and prevalence of multiple sclerosis in south east Scotland. They have identified standardised multiple sclerosis prevalence rates for the Lothian and Border Regions of 219 and 209 per 100 000 respectively, the results challenging the theory that the high prevalence rates in the British Isles confirm the existence of a step in multiple sclerosis prevalence in the British Isles and therefore, its results are definitely flawed. Additionally, a basic experimental study design requires at least a minimisation regarding inclusion and exclusion criteria, randomisation, and definitions of response or outcome variables. This information is not provided by Del Brutto’s report; its design fails to protect against potential bias in patient selection or evaluation of outcome. The definition of subarachnoid cystersci used by Del Brutto was based on “appearance on CT of hypodense cystic lesions located over the cerebral hemispheres, the Sylvian fissure, or the CSF cisterns at the base of the brain”. It is well known that there are many other diagnostic possibilities to be considered in the differential diagnosis of subarachnoid hypodense lesions.1,2 Besides, CT is not a reliable procedure for diagnosing subarachnoid cystersci, as is MRI. In fact, we cannot be completely sure, for example, that the CT images shown in the report correspond to subarachnoid cystersci. If we were to use MRI on this patient, they might correspond to a parenchymal cyst which resolved as a reflection of the natural history of the condition. There is a report that objectively confirms or rejects this assertion.

Del Brutto’s report1 maintains that evaluation of the therapeutic response to albendazole included comparison of the size of the cysts, as well as clinical evaluation before and after treatment. To consider the size of cysts as a response variable is certainly useless because of the obvious difficulties in measuring cyst size in the subsequent follow up CT. It is also widely accepted that the clinical manifestations of neurocysticercosis are polymorphic, and their clinical course is unpredictable; therefore, the clinical manifestations as an outcome variable is entirely biased. Another personal appreciation of Del Brutto’s assertion is similarly flawed in that it does not include a control group against which the intervention group is compared; therefore, its results are definitely flawed. Additionally, a basic experimental study design requires at least a minimisation regarding inclusion and exclusion criteria, randomisation, and definitions of response or outcome variables. This information is not provided by Del Brutto’s report; its design fails to protect against potential bias in patient selection or evaluation of outcome. The definition of subarachnoid cystersci used by Del Brutto was based on “appearance on CT of hypodense cystic lesions located over the cerebral hemispheres, the Sylvian fissure, or the CSF cisterns at the base of the brain”. It is well known that there are many other diagnostic possibilities to be considered in the differential diagnosis of subarachnoid hypodense lesions.1,2 Besides, CT is not a reliable procedure for diagnosing subarachnoid cystersci, as is MRI. In fact, we cannot be completely sure, for example, that the CT images shown in the report correspond to subarachnoid cystersci. If we were to use MRI on this patient, they might correspond to a parenchymal cyst which resolved as a reflection of the natural history of the condition. There is a report that objectively confirms or rejects this assertion.

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out the need to conduct a long term, placebo controlled trial with precise end points, proper randomisation, sample size calculations, and predetermined statistical calculations, to evaluate properly the effectiveness and determine the indications of aetiological treatment for neurocysticercosis. In the era of evidence-based medicine, we neurologists and general practitioners should be demanding regarding use of sound scientific information with methodological rigour for improving our clinical decision making. Medical information from reports that do not conform to the minimal requirements of a clinical trial should be avoided.

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The author's reply:
I celebrate the rigid academic standards of Carpio's medical practice, but wish there were matched with knowledge on the available literature on albenzadole therapy for neurocysticercosis. That albenzadole actually has a cysticidal effect is beyond all doubt.1 The drug has been used to treat patients with neurocysticercosis since 1987, and physicians who are familiar with the disease know that it is effective. Moreover, the single study in which albenzadole has not been useful for adult parenchymal brain cysticerci—published by Carpio and associates—has been questioned due to inaccurate recollection of data.2 In our study, we did not attempt to verify the cysticidal effect of the drug (it has already been demonstrated) but to document if albenzadole could also be useful in a severe form of neurocysticercosis that has been associated with a grim prognosis.3 Under these circumstances, it is not ethical to deprive a group of patients of a safe and inexpensive treatment just for the sake of science. In addition, Carpio's concerns about the criteria we used for the diagnosis of subarachnoidal cysticerci are typical of those who are not familiar with the disease. The problem with CT is that this imaging method may not conform to the minimal requirements of a clinical trial should be avoided.

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BOOK REVIEWS


Ever since the landmark overview “Do stroke units save lives?” the momentum behind the organisation of stroke sevices has inexorably increased. Within this concise paperback of seven chapters and 112 pages, written in the main by Langhorne and Dennis on behalf of the Stroke Unit Trialists Collaboration, lies a detailed, evidence-based, and critical summary of the arguments for stroke units. The authors lead the reader in a logical fashion through the steps necessary to critically appraise and assimilate available evidence into a systematic review. Basic but often overlooked general principles such as sources of bias are discussed in detail as well as matters relating more specifically to stroke such as outcome measurement.

Particularly helpful are the chapters on the economics of stroke units and the implications for service planning in which the available evidence has been used to suggest how and at what cost (in fact an overall saving) stroke units might be developed. This comprehensively referenced book will be read by members of all disciplines involved in stroke care. If I was about to ask for funding for a stroke unit I would certainly have it in my pocket!

Peter Martin


There are two parallel strands to the development of our understanding of immune mediated disorders of peripheral nerve. The first grew from the demonstration in the 1950s, by Waksman and Adams, that rabbits immunised with homologous sciatic nerve and adjuvant developed an inflammatory demyelinating neuropathy. In this model, experimental allergic neuritis, the CSF characteristically shows a raised protein concentration and a paucity of cells. These findings replicated those of Guillain-Bare and Strohl on the CSF abnormalities of Landry’s disease and so spawned the notion that Guillain- Bare syndrome was immunologically mediated. Accordingly, over the past 20 years patients with Guillain-Bare syndrome and chronic inflammatory demyelinating polyneuropathy have been exposed to immunosuppressive regimes borrowed from other inflammatory disorders.

The second important development has been the growing understanding of the relation between plasma cell dyscrasias and peripheral neuropathies. The association of peripheral neuropathy and myeloma was noted in the 1930s and that with an IgM monoclonal gamopathy was reported in the 1960s. Twenty years later IgM antibodies were found that were directed against myelin associated glycoprotein. This work has gathered pace and over the past 10 years, peripheral neuropathies have been described in association with specific antiganglioside and antiphosphatidyl antibodies.

Alongside this expansion of interest in the immunology of peripheral nerve disorders, new infective neuropathies have emerged such as those due to HIV and Lyme disease, first recognised in 1993. It is an appropriate time then for this authoritative text on immune mediated neuropathies. The scope of the book is wide, including scientific overviews of immune interactions in the peripheral nervous system as well as pragmatic accounts of the use of immunosuppressant drugs and the management of neuropathic pain. The inflammatory demyelinating neuropathies and antibody associated neuropathies are comprehensively surveyed, as well as more difficult entities such as the post-polio syndrome and the rare toxic inflammatory neuropathies. The dry review of silicone neurorotocinity by Rosenberg is a special treat. British readers may be surprised to find only one United Kingdom contributor to this American-Dutch edited text, whereas 18 authors are American, 11 are from The Netherlands, two each from Italy, Japan, and Israel, and one each from Canada, Nepal, and Switzerland. It is not cheap, but it has no equal as a comprehensive, accessible, and useful resource for the practising neurologist.

Alasdair Coles

Tethered spinal cord comprises a group of dysraphic conditions in which the conus medullaris is located in an abnormally low position. Tethered cord syndrome is stretch induced symptoms manifested by motor and sensory deficits in the lower limbs and incon tinence, and is often associated with musculoskeletal deformities. This book draws together all aspects of the embryology, pathophysiology, diagnosis, and treatment of this rare but important condition in a detailed but readable format. Inevitably there is repetition but this is unavoidable in a multiauthour text in which each chapter can be read as a whole. The text is well illustrated with diagrams, clear radiological images, and well chosen clinical photographs.

The chapter on pathophysiology of the tethered spinal cord is a fascinating summary of the various experimental studies that have been undertaken in this condition. Although the relevance of some of the models to the condition may be questioned one cannot help but admire the ingenuity and inventiveness of the investigators. Acute traction on the spinal cord has been shown to be associated with impairment of evoked potentials, reduction in spinal cord blood flow, and changes in glucose metabolism. Chronic experiments have shown recovery in neurological deficits after 9 months.

There are well written chapters on diagnosis, investigation, and surgical treatment with plenty of intraoperative detail. The final chapter considers controversies associated with the treatment of the tethered cord syndrome. Most neurosurgeons would now agree that surgical treatment is definitely indicated in patients with progressive neurological deficits and most are increasingly prepared to consider prophylactic surgery in patients with tethering but in whom neurological deficits are absent or established. Although urodynamical testing helps to identify patients with neurogenic bladders, urinary dysfunction may be intermittent and symptomatic tethering may not always be disclosed. A tethered spinal cord is prone to produce problems during periods of rapid growth in childhood, but even when growth is complete patients with an undiagnosed tethered cord may undergo serious deterioration if subjected to sudden flexion movements associated with trauma. A selection process is outlined to help decide treatment in four main categories of presentation. Unfortunately the algorithm can be difficult to interpret—for example, “fluctuating signs and symptoms noted in a patient with stable neurological deficits—patient must be followed closely for detection of minor progression”

A useful book which I would recommend to all doctors who treat patients with spinal dysraphism and I suspect that many will wish to have a copy in their personal collection.

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