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 classical 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 lofepramine. 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, fearful, 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 She became more withdrawn and uncommunicative with incontinence of urine.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 transferred 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 virtually continuous semirepetitive sharp waves with some right sided predominance. Although seizure-like evolution of discharges was not seen, the electrographic picture was considered to be in keeping with complex partial status.

Following this, the administration of diazepam was tried. On questioning, the patient had developed myoclonus. Although initially the myoclonus was not abolished by 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 synchronous 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 laterisation on the EEG is well recognised in Creutzfeldt-Jakob disease as are
periodic PLEDs, which are often associated with contralateral myoclonic jerks.  

The two cases described here illustrate that a diagnosis of Creutzfeldt-Jakob disease should be considered where a rapid decrease of 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.

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1 Schilder P. Zur Kenntnis der sogenannnten diffusen Sklerose. Zeitschrift für die Gesamte Psychologie und Physiologie 1912; 65: 60.


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 heat stroke are on days afterwards he developed an acute polyneuropathy that met clinical and neurophysiological criteria for Guillain-Barré syndrome. Their nerve conduction abnormalities are reported as “compatible with scattered demyelination”.

Our patient had chronic HCV infection by the Anti-Siphon Device (ASD, Heyer Scientific, Inc.) 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, neurotoxic, and increased in Guillain-Barré syn-

Guillain-Barré syndrome-like neuropathies have been reported from south Asia,1 whereas heat stroke is common, but they were not noted in connection with epidemic heat stroke in North America.1 Our patient had all features associated with fatal heat stroke: long lasting coma, shock requiring aggressive catecholamines, metabolic acidosis, and disseminated intravascular coagulation.1 Guillain-Barré syndrome may occur more often after heat stroke, if more patients survive the hyperthermia thanks to intensive care.

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7 Hart GR, Anderson RJ, Crumpler CF, et al. Epidemic classical heat stroke: pathogenesis, pathophysiol-

ergies such as Elekta-Cordis Horizontal ER, SiphonGuard, Vertical Valve, Chhabra Valve, Fuji Valve, or Miehite Dual-Switch Valve are widely used.1 Their main drawback is susceptibility to malfunction when the shunt becomes displaced from its vertical axis after implantation and unpredictable operation during persistent bodily movements. The membrane devices: the Anti-Siphon Device (ASD, Heyer Schulte) or Siphon Control Device (SCD), Medtronic Inc., are also not proved clinically effective, but in several cases these devices may obstruct the CSF drainage when the subcutaneous pressure increases or the scar tissue isolates the device from atmospheric pressure. The flow regulat-

Hydrodynamic performance of a new siphon preventing device: the SiphonGuard

Around 10% to 30% of shunt revisions may be attributed to posture related overdrainage. Of the various siphon preventing devices available at present, two construction types are the most prominent: those using a gravitational mechanism and those using a subcutaneous membrane. Gravitational devices such as Elekta-Cordis Horizontal ER, SiphonGuard, Vertical Valve, Chhabra Valve, Fuji Valve, or Miehite Dual-Switch Valve are widely used. Their main drawback is susceptibility to malfunction when the shunt becomes displaced from its vertical axis after implantation and unpredictable operation during persistent bodily movements. The membrane devices: the Anti-Siphon Device (ASD, Heyer Schulte) or Siphon Control Device (SCD), Medtronic Inc., are also not proved clinically effective, but in several cases these devices may obstruct the CSF drainage when the subcutaneous pressure increases or the scar tissue isolates the device from atmospheric pressure. The flow regulat-
ing Orbis-Sigma Valve (Elekta-Cordis) may 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 valve 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 on spring valve 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 rates increase 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 rate of 1 ml-1.5 ml may cause clinical deterioration without initiating the antisiphon action of the SiphonGuard. Another possibility concerns the reverse change—that is, switching back from high to low resistance, to be expected when a patient moves from a vertical to a horizontal position. The device may not return to its state of low resistance. If the resistance switching mechanism is indeed triggered by a differential pressure (with a threshold of around 5 mm Hg) the SiphonGuard may stay in the high resistance state, causing underdrainage in the horizontal body position.

In vivo, the device may contribute to the significant fluctuations of pressure resulting from the difference between the operating pressures for low and high resistance—similar to that described for the Orbis-Sigma Valve. Moreover, it may not prevent the overdrainage related to nocturnal vasomotor pressure waves, as often reported in paediatric cases. These reservations, based on our short laboratory study, should be taken into consideration both by neurosurgeons and the manufacturer. Whether they cause system malfunction under specific clinical conditions remains to be shown. We advocate a well controlled multicentre study on this new and interesting device together with in vivo measurements of shunt function using a CSF infusion test during tilting.

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Convulsions induced by donepezil

Donepezil, a centrally acting acetylcholinesterase inhibitor, has been recently introduced for the symptomatic relief of cognitive impairment in patients with mild to moderate Alzheimer’s disease. Several adverse events thought to be related to donepezil have been reported so far, the most common ones being gastrointestinal disturbances due to cholinergic effects of donepezil.1 Convulsions have not been reported for donepezil to date. We report on a patient with mild Alzheimer’s disease who presented with convulsions during treatment with donepezil.

The patient was a highly educated, ApoE4 homozygous, 72 year old man, who was diagnosed with dementia of probable Alzheimer’s type (NINCDS-ADRND criteria) 14 months previously. His medical history, with the exception of non-familial dementia, was unremarkable and his only medication was 100 mg aspirin daily. His mini mental state examination score was 22 points. He was treated with 5 mg donepezil once daily for 2 weeks, then 10 mg a day for 23 days when he was admitted due to convulsions. The patient was unconscious for 40 minutes with urinary incontinence and bitten tongue. Blood analyses were normal. A contrast brain CT showed a mild degree of cortical atrophy with no structural lesions. EEG showed mild and diffuse neuronal dysfunction with the absence of grafoelements indicative of epileptic character. As donepezil was discontinued and no other therapy was instituted. Six weeks later 5 mg donepezil once daily was restarted. On day 52 of donepezil treatment the patient’s caregiver had reported loss of consciousness and convulsions in our patient. The donepezil was discontinued and 100 mg indomethacin a day was prescribed. For the subsequent 8 months the patient has been convulsion free and his current mini mental state examination score is 19.

Convulsions in Alzheimer’s disease are rare until late in the illness, when up to 5% of patients reportedly have infrequent seizures.2 We think that convulsions reported in our patient could be due to donepezil. It has already reported that some centrally acting cholinesterase inhibitors—that is, tacrine, varenclam, and physostigmine—might induce convulsions in patients with Alzheimer’s disease. The mechanism of convulsive action of acetylcholinesterase inhibitors is not clear. As donepezil seems a useful drug in some of the carefully selected patients with mild to moderate dementia of Alzheimer’s type we think that this report will extend our knowledge of donepezil’s safety profile.

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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.3,4 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 6 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 paresthesias 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, vibration 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 of nerve action potentials without any significant slowing of conduction velocity. There were spontaneous fibrillations in the right tibial anterior muscle. The complete blood picture was normal. There was a toxic cause was considered.


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The role of fenofibrate was confirmed by regression of the symptoms after discontinuation of this drug without the addition of any other treatment. There are no previous reports of histological findings in neuropathy due to fibrates. The delay between initial treatment with fenofibrate and the appearance of the symptoms as well as the time required for them to regress, suggest a cumulative toxic effect but no other predisposing risk factor such as high dosage or renal failure was present.

In conclusion, fibrates can be responsible for neuropathies even when given in approved doses and in the absence of renal failure.

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CORRESPONDENCE

Macs with multiple sclerosis

Rothwell and Charlton1 have suggested that Scottish ancestry is associated with an increased susceptibility to multiple sclerosis. They make the novel observation that a higher than expected proportion of patients with multiple sclerosis had Scottish surnames as defined by the prefix Mc or Mac. They quote that the prevalence of Scottish surnames in the Highlands and Islands with a surname of Mc or Mac is 22.6%. They then suggest that this is the percentage for Mc or Mac in Orkney and Shetland but these islands are not part of the Highlands and Islands. In Orkney and Shetland, in fact, only 3.5% of the population have a surname beginning with Mc or Mac, which is much lower than the percentage in north east Scotland—namely 7.5%.

Rothwell and Charlton do make the point, however, that an increase in the proportion of surnames prefixed with Mc or Mac with latitude within Scotland is not associated with an increase in the prevalence of multiple sclero-
Scotland is at least partly a function of the Mc or Mac.

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 203 and 219 per 100 000 respectively, the results challenging the theory that the high prevalence rates previously reported in Scotland are peculiar to the north east and its offshore islands. The authors postulate that the apparent step in prevalence rates between England and Scotland may be due to the distinctive Celtic ancestry of the Scottish population as can be crudely measured by surnames prefixed with Mc or Mac.

In Northern Ireland we have also identified a much higher prevalence rate for the disease than exists in England and Wales and have speculated that the similar rate to that in Scotland is at least partly a function of the common ethnic origins of the two populations. The contiguous region of Coleraine, Moyle, Ballymena, and Ballymoney lies less than 20 miles from Scotland at its closest point and has a standardised prevalence rate for all multiple sclerosis, based on the 1961 census population for Northern Ireland, of 224 (95% confidence interval (95% CI) 216–232) per 100 000. Using a similar method to Rothwell and Charlton (British Telecom phone book of the area), 17% of the study population had a surname prefixed with Mc or Mac and it is of note that 22.9% of prevalent cases had such a surname prefix (odds ratio = 1.46, 95% CI 1.09–1.93, \( \chi^2 = 6.82, p = 0.009 \)).

Our results support the conclusion of Rothwell and Charlton that Celtic ancestry is a risk factor for multiple sclerosis and confirm the existence of a step in multiple sclerosis prevalence in the British Isles between England/Wales and Scotland/Northern Ireland.

Abendazole therapy for subarachnoid cystercerci: clinical and neuroimaging analysis of 17 patients

By contrast with the weaknesses of anecdotal observations from case series, the power of randomised clinical trials for deciding the benefit of therapy has become increasingly evident and indisputable world wide. Nowadays, to argue against the validity of this assertion may seem superfluous; however, a recent paper reported by Del Brutto\(^1\) regarding treatment in neurocysticercosis ignores basic procedures for well performed clinical trials by using inappropriate and misleading terms as an outcome variable is entirely unpredictable; therefore, the clinical manifestations as an outcome variable is entirely biased. Another personal appreciation of Del Brutto is that albenzol reaches high concentrations in CSF, and has been used with success in some patients with subarachnoid cysts; nevertheless, studies used as support of this therapy in some patients might sometimes be harmful, particularly in the subarachnoid localisation, because some patients have developed arteritis and hydrocephalus after the administration of antihelminthic drugs.\(^2\)

According to these authors a parasite may be easily removed surgically before an inflammatory reaction develops.\(^3\) A randomised clinical trial of treatment of neurocysticercosis\(^4\) considers the question of to what extent and the which patients treatment with either praziquantel or albenzol is effective. The improvement attributed to these drugs in several studies may be related to the lack of appropriate controls and is likely to be a reflection of the natural history of the condition. The authors point
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 albendazole therapy for neurocysticercosis. That albendazole 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 albendazole has not been useful for adult parasite cerebral parenchymal brain cysticerci—published by Carpio et al—has been questioned due to inaccurate recollection of data.1 In our study, we did not attempt to verify the cysticidal effect of the drug (it has already been demonstrated) but to document if albendazole could also be useful in a severe form of neurocysticercosis that has been associated with a grim prognosis.2 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 subarachnoid cysticerci are typical of those who are not familiar with the disease. The problem with CT is that this imaging method may misdiagnose some subarachnoid cysts as parenchymal cysts, but the opposite is not true.

As a physician interested in the advancement of science, I applaud Carpio's interest in evidence-based medicine but I completely disagree with him in that information from reports other than clinical trials should be avoided. He must remember that outstanding contributions to medical knowledge have been made through single case reports, small clinical series, and open trials. On the contrary, the “third-hand” has been the shield of major medical frauds.3 Medicine is art and science, and wise physicians know that information from clinical findings actually have a “significant” impact on everyday clinical practice.

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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: a critical 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. This finding replaced those of Guillain-Barré and Strohl on the CSF abnormalities of Landry’s disease and so spawned the notion that Guillain-Barré syndrome was immunologically mediated. Accordingly, over the past 20 years patients with Guillain-Barré 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 gammapathy 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 antiphosphate 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 neurorototoxicity 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.

This is a handsome and literally illustrated guide to the success and frozen section diagnosis in neuropathology. This aspect of practice remains a central part of a clinical neuropathologist's role and this book can be recommended to trainees and practitioners for its wealth of illus, drawn and practically oriented text. It is particularly useful to see a wide range of appearances for each tumour illustrated—for example, 20 figures illustrating metastatic tumours, 13 illustrating pituitary adenomas, and 38 illustrating various grades of astrocytic tumours. This enables the less readily diagnosed examples to be considered as well as more typical varieties. Typical varieties tend to be the only ones illustrated in a less specialised text. There are 18 chapters that cover each of the main types of tumour encountered as well as providing advice on making and interpreting smears and dealing with lesions that do not smear well. The emphasis is on using smears as standard preparations with frozen sections as back up when required—a procedure that is probably adopted in most neuropathology departments.

The success of a book like this depends crucially on the quality of the photographs. These are, appropriately, all in colour. Many are of excellent quality. Some are intentionally obscure—for example, to make the point that dermatofibrosarcoma protuberans may be too tough to examine in smears (fig 18.1). A few have rather poorly defined features and these tend to be illustrations of frozen sections which inevitably lack the crispness of smears. However, even these illustrate the points intended. The legends to the figures are full enough to avoid the need for arrows that might otherwise have obscured the images. There is a useful index, but the book would have benefited from more references—I found only six.

Smear diagnosis is best learnt by doing it, with a sympathetic, experienced colleague at one's elbow. Often this condition cannot be fulfilled and I would strongly recommend this book as a very valuable alternative or adjunct.

MARGARET ESIRI


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 incontinence, 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 multiauthor 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.

R LAING


Do you find conference dinners a dull ordeal? Fear no longer! You can sparkle with amusing tales of seizure induced religious conversion or tales of primitive brain surgery. Often this condition cannot be remedied 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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