Pseudotumour after arteriovenous malformation embolisation

The association between venous outflow obstruction and the development of pseudotumour syndrome is well known, although the mechanism by which the rise in CSF pressure is brought about is less certain. Although there is much evidence that the manifestations are a result of a disturbance of CSF dynamics, previous reports have focused solely on a disturbance to absorption. We present a case in which it is proposed that alterations in CSF formation, and to a lesser extent absorption, are responsible for the development of the syndrome.

At 2 years of age, and being aware of a distinct abnormal growth, a female infant underwent cerebral CT. This showed an unexpected arteriovenous malformation involving the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was undertaken at the age of 5 years when vertigo and intermittent numbness of the left arm and leg had been present for about 12 months.

Angiography showed a deep right temporal lobe arteriovenous malformation consisting of three separate fistulae supplied by the right posterior cerebral and posterior communicant arteries. These drained into a large venous varix which subsequently drained into the Galenic venous system. A cerebral blood flow study showed a steal syndrome affecting the frontotemporal area, and a decision was made to attempt embolisation. Complete occlusion of the fistulae was achieved by transarterial platinum coil embolisation.

The patient complained of right sided headache for 24 hours after the procedure, resolving with minor analgesia. Brain CT the next day was reported as normal. A full ophthalmological review was undertaken before the next day was reported as normal. A full ophthalmological review was undertaken before the next day was reported as normal. A full ophthalmological review was undertaken before the next day was reported as normal.

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It is well known that obstruction to a major portion of the cranial venous outflow can produce intracranial hypertension, presumably by impairing CSF absorption across the arachnoid villi. In the present case it would seem that sluggish flow in the venous varix after embolisation has resulted in thrombosis, which has propagated to the vein of Galen. As all investigations seem to have the thrombus confined to this region, a region of relative paucity of arachnoid granulations, and the major outflow tracts seem normal, it is difficult to accept that impairment of absorption is the mechanism responsible in the current case. An alternative mechanism must be considered.

It is held that one of the determinants of the rate of CSF production is the pressure gradient across the choroid plexus capillaries. Reduction in this pressure has been shown to decrease the rate of CSF formation, and it is possible that increases in the transcapillary pressure will, in all other parts of the body, result in increased transudation from the capillaries, leading to increased CSF formation. The malformation in the present case, haemodynamically important enough to result in symptoms of steal, and present since birth, may have resulted in a subnormal transcapillary gradient, and hence a possibly decreased CSF production. If this were the case, with decreased production serving to retard the normal development of absorptive capacity, then the increase in the pressure in the choroid plexus capillaries brought about by both the closure of the fistulae and the subsequent venous thrombosis may have resulted in a rate of CSF production greater than could be handled by the absorptive system. Resolution of the thrombus, recruitment of venous collaterals, and possibly an increase in absorptive capacity would have resulted in the resolution of the syndrome.

Brain CT at level of vein of Galen demonstrating thrombus.

A cerebral angiographic at 3 months confirmed obliteration of the fistulae and vein of Galen and poor filling of the straight sinus with no evidence of obstruction to major venous outflow pathways. At this time CSF pleocytosis, via lumbar puncture, was normal, work has been done in the area of the relation between CSF formation and venous occlusion.

Although the above report is somewhat speculative, it could serve to explain the facts which at this stage of our understanding of CSF dynamics cannot be wholly accounted for. A case of pseudotumor developing in the setting of minimal venous thrombosis, particularly in part of the venous system not thought to play a major part in the absorption of CSF, must force us to reconsider our opinions as to the relation between venous obstruction and CSF dynamics.

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False negative polymerase chain reaction on cerebrospinal fluid samples in tuberculous meningitis established by culture

The polymerase chain reaction (PCR) has been reported to be of diagnostic value when performed on CSF samples in tuberculous meningitis. Examination of CSF disclosed a lymphocytic exudate. Repeated samples were sent to a British referral laboratory where CSF PCR for M tuberculosis was reported negative. Despite this, antituberculous treatment was continued for 12 months and both patients responded clinically. Several weeks after the negative PCR result, M tuberculosis was cultured on Lowenstein-Jensen slopes from CSF taken from both patients. False negative PCR in tuberculous meningitis established by culture has rarely been reported. The two patients are described to emphasise the dangers of overreliance on PCR in cases of suspected tuberculous meningitis. Premature cessation of treatment would have had tragic consequences for the two patients concerned.

The first patient was a 28 year old Asian man, last in India 8 years previously. He was sent from a clinic to hospital for incision and drainage of two deep seated Staphylococcus.
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False negative polymerase chain reaction on cerebrospinal fluid samples in tuberculosis meningitis

There have been few studies in the literature concerned solely with the use of the polymerase chain reaction (PCR) to identify Mycobacterium tuberculosis DNA directly from cerebrospinal fluid (CSF). These studies suggest that in some cases, PCR may be more sensitive than culture; however, in the largest study, performed by Nguyen et al.,

been recognised in Dejerine-Sottas disease, peripheral neuropathy with an early onset in childhood, and a more severe phenotype than CMT1. CMT1 and Dejerine-Sottas disease are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina, are characterised by thickening of peripheral nerves, and thickening of the cauda equina.

A 15 year old Japanese girl presented with CMT disease. She showed delayed motor development. Although she became ambulant at 1 year and 8 months of age, she was never able to run. She was referred to our hospital due to progression of her gait abnormality. Her mentality and higher brain function were normal. Neurological examination disclosed weakness in both proximal and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, hyperreflexia, and areflexia. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hands and deafness. Optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed abnormal prolongation of the I-III interpeak (2.81 ms on the right side, 2.88 ms on the left side). Brain MRI (figure) showed significant thickening of bilateral trigeminal nerves (7 mm) compared with that of controls (3.15 ± 1.62 mm [mean ± 2 SD], n=20). However, other cranial, spinal nerves and roots were not thick on physical examination or MRI study. Sural nerve biopsy was not performed.

Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A134C) at codon 81 resulting in a substitution of arginine for histidine only in the patient. A DNA duplication in chromosome 1p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. The patient’s mother did not show any mutations in the P0 gene.

CMT type 1 is caused by abnormalities in myelin protein of Schwann cells. Repeated demyelinating and remyelinating responses in the peripheral nerve produce diffusely enlarged diameters of nerves in CMT type 1, and thickening of the cauda equina, nerve roots, and ganglia has also been found. Although blepharoptosis, ophthalmoplegia, facial weakness, deafness, dysphagia, and dysphoria in CMT have been previously reported, clinical involvement in the cranial nerves is rare and thickening of cranial nerves has not been reported except for the acoustic or optic nerves in some cases.

In the present study, our patient showed severe clinical manifestations of early onset and undetectable conduction velocities. Therefore, this patient was considered to have a severe variant of CMT1 or Dejerine-Sottas disease. Although her facial sensation, mastication power, and hearing acuity were normal, the thickness of bilateral trigeminal nerves on MRI and prolongation of the I-III interpeak interval in auditory brain stem response were found. The I-III interpeak interval represents the conduction time from the eighth nerve to the pontomedullary interval and is related to the sensory and motor functions of the peripheral nerve.

Trigeminal neuropathy with CMT has been reported. In these rare cases, trigeminal neuromyalgia was inherited, suggesting a partial symptom of CMT. Although some patients were surgically treated, it was not clear whether a thickened trigeminal nerve was present. Moreover, on electrophysiological studies of facial and trigeminal nerves in CMT, Kimura et al. reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the facial and trigeminal nerves. However, the MRI study of our patient suggested that the fifth cranial nerves were subjected to the same pathological process that affects other peripheral nerves.

Our patient showed no DNA duplication on chromosome 1p11.2 and we found a novel mutation (A134C) representing an Arg134 to His substitution in the P0 gene. Histidine 81 is conserved among many other species, including cows, rats, chickens, and sharks. This mutant allele was absent in the DNA from 100 controls. Therefore we identified this mutation as pathogenic. Arg134His was located in exon 3, which codes for the extracellular domain of the P0, which is highly glycosylated, and plays a part in myelin compaction by homophilic interaction and many mutations in this area have been reported. Although the phenotypic variability is related to the position and nature of the P0 mutation, patients with cranial nerve involvement are rare in CMT with a P0 mutation. Therefore, the unique thickening of trigeminal nerves and the clinical severity in this patient may be related to this novel missense mutation. A careful comparison of the clinical, electrophysiological, and histopathological data between patients with CMT should be conducted.

We are indebted to the families studied for their cooperation and support. This work was partly supported by Ushara Memorial Foundation, the Sasaki Health Science Foundation, the Primary Amyloidosis Research Committee, and the Ministry of Education, Science and Culture of Japan 10528020, 18832993.
The patient made an uneventful recovery and was referred for staging investigations and consideration of postoperative therapy. An LDH estimation was within normal limits and HIV serology was negative. Whole body CT including repeat CT of the brain did not show any evidence of lymphadenopathy or lymphomatous deposit. Bone marrow examination was declined. Postoperative adjuvant whole brain or localised radiotherapy was discussed with the patient, however, she declined any further intervention. She has been closely reviewed in the follow up clinic and after 6 months there has been no clinical or radiological change.

Primary intracerebral lymphomas represent about 2% of intracranial neoplasms and 2% of all lymphomas. They occur most commonly in the 6th decade of life with a female to male ratio of 2:1. The association between primary intracranial lymphoma and immunodeficiency has long been established, and it is not surprising, therefore, that the incidence has increased 10-fold over the past 3 decades with the onset of transplant surgery and, particularly, the AIDS epidemic. In postmortem studies, these neoplasms are found, on average, in 5.5% of AIDS cases, and malignant cerebral lymphoma is the most common diagnosis of a focal intracranial lesion in patients with AIDS. Malignant primary lymphoma can occur throughout the CNS and they often have a periventricular distribution. Multifocality seems to be more common in patients with AIDS. The CT scan usually shows hyperdense masses with peritumoral oedema and 92% enhance after administration of contrast medium.

Leptomeningeal lymphoma is usually encountered as a late complication of systemic non-Hodgkin’s lymphoma, although primary leptomeningeal lymphoma is occasionally seen. The prognosis for these tumours is poor. Diffuse primary cerebral lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellar pontine angle mimicking acoustic neuromela or meningoia has been reported; Vigusin et al reported a patient with a calcified temporaloparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular lymphoma with clinical features similar to our case. Rubinstein described a case of follicular lymphoma metastasis found in the dura of a 61 year old man at necropsy.

We found no report of a primary follicular extracerebral lymphoma. Similar radiological and intraoperative appearances of the tumour in our case to splenoid wing meningioma suggest that this entity should be considered as a rare differential diagnosis.

Determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentration can serve as an indicator of brain copper metabolism. However, the complex mechanisms which by copper crosses the blood-brain barrier and the factors determining the CSF copper concentration in humans are largely obscure. Copper can pass into and out of the CSF by various mechanisms. For example, active transport through the blood-brain barrier or the blood-CSF barrier, or passive diffusion of the free or the bound fraction (bound to albumin or coeruloplasmin) through the blood-CSF barrier. We studied the factors influencing CSF copper concentration using a stepwise multiple linear regression model. The independent variables were age, plasma coeruloplasmin, CSF/serum albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum coeruloplasmin and total serum copper concentration). The CSF copper concentration was treated as a dependent continuous type. We investigated lumbar CSF samples from 113 patients. These patients had dementia, extrapyramidal, or tremor symptoms; lumbar puncture was performed to exclude Wilson’s disease, and none of the patients had the disease. Copper was measured by flameless atomic absorption (Perkin Elmer, HGA 500, Ueberlingen, Germany). Coeruloplasmin was determined nephelometrically (Beckman Instruments, Brea, CA, USA). The age of the patients was 50.0 (SD15.5) years; 50 were women and 63 were men. Mean serum coeruloplasmin concentrations were 394 (SD 79) mg/l. Mean serum copper concentrations were 1194 (SD 335) µg/l. Mean calculated free copper concentrations in serum were 78.5 (SD 1285) µg/l. Mean CSF copper concentrations were 14.16 (SD 6.60) µg/l. The mean albumin ratio (AR) was 6.63x10^-3. The mean ratio of calculated free copper concentration to total serum copper was 6.6%; the ratio of CSF copper to serum copper was 1.2%; and the ratio of free serum copper to CSF copper was 18%.
stepwise linear regression model (F to enter 4.0, F to remove: 3.996), significant positive predictive value of the CSF copper concentration were found to be AR (p=0.0001) and serum copperolaumlin (p=0.0057). The other independent variables mentioned above showed no statistically significant relation with CSF copper concentration. The figure shows the simple linear regression between CSF/serum albumin ratio and serum copperolaumlin concentration (on logarithmic axes; R²=0.46, p=0.001). The formula for the CSF copper concentration, derived from the multiple linear regression model, is: copper (µg/l) = 3.32 + 0.653 x CSF/serum albumin ratio (x10−3) + 0.012 x serum copperolaumlin (mg/l). According to this analysis, CSF/serum albumin ratio and serum copperolaumlin together determine 25.3% of the variation in CSF copper concentration (adjusted R²=0.253), implying that other (unknown) factors determine the remaining 74.7% of the variation. We have been able to demonstrate here that the CSF copper concentration is determined in a highly significant manner by disturbances in the blood-CSF barrier and by the serum copperolaumlin concentration. It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum copperolaumlin concentration, 29.7% of the measured CSF copper originated from the blood, the CSF by passive diffusion bound to copperolaumlin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20x10³, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to copperolaumlin). A variable fraction of the CSF copper concentration, depending on the degree of damage to the blood-CSF barrier, therefore crosses from the blood into the CSF and can be measured there. Our formula would therefore predict, in patients with Wilson’s disease with intact blood-CSF barrier (assuming a CSF/serum albumin ratio of 6.5x10³), that the CSF copper concentration is actually reduced by 27.4%, when the serum copperolaumlin concentration falls from its normal value of 394 µg/ml to 66 µg/ml. In consequence, CSF copper in patients with Wilson’s disease is evidently substantially free, implying that a larger fraction than previously assumed of the raised CSF copper in patients with untreated Wilson’s disease originally entered the CSF by passive diffusion (bound to copperolaumlin) tends towards zero. It can be concluded from this that, when the aim of therapy is considered in terms of the total CSF copper concentration, a region around 30% lower than the upper limit of the normal range should be aimed for. This is supported by the clinical findings that patients report feeling better when the CSF copper concentration is below this value. This analysis also shows that the raised copper concentration in the CSF can only originate from the brain. In particular, it is not associated with free serum copper, but evidently only via storage in the brain. The investigation here also shows that, after determining the CSF copper concentration, the copperolaumlin-bound fraction originating from the plasma should be subtracted according to the formula we have given, or better, all measured copper concentrations in the CSF should be adjusted using the CSF/serum albumin ratio and serum copperolaumlin concentration. A statistical relation with a low correlation (p=0.05) between CSF protein content and CSF copper was already shown in 1989 in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum copperolaumlin (therefore of bound serum copper). Furthermore, we have been able to determine statistically the correlation of CSF copper which enters the CSF across the blood-CSF barrier.

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Solitary intracranial myofibroma in a child

A rare case of solitary interhemispheric myofibroma with excellent outcome in a 20 month old boy is described. The clinicopathological features of this unusual condition are reviewed with emphasis on the CNS manifestations.

A case of congenital fibrosarcoma was first diagnosed by Williams and Schrum and was subsequently renamed congenital generalised fibromatosis by Stout in 1954 as a distinct form of juvenile fibromatosis characterised by tumour-like nodules involving the skin, soft tissues, bones, and viscera. Based on the ultrastructural and immunohistochemical features of the cell of origin and the occurrence of this condition in infants, as well as congenitally, it was renamed infantile myofibromatosis by Chung and Enzinger in 1981. This disorder is considered to represent a hamartomatic myofibroblastic prolif-
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofibrils.

Solitary lesions of infantile myofibromatosis are more common than multiple lesions, with twice as many males as females being affected, and generally involve the skin and soft tissues, especially of the head and neck. Solitary lesions are less commonly found in viscera or bones. Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported along with few cases of CNS involvement in the generalised form of infantile myofibromatosis. The prognosis is best for cases with solitary masses and less favourable for multicentric cases, particularly where visceral lesions are present, in which morbidity and mortality derive predominantly from pulmonary involvement or mass effect.

The differential diagnosis for this lesion included meningioma, schwannoma, and haemangiopericytoma. Regionally, the histology was reminiscent of the rare microcystic haemangiopericytoma. Regionally, the histology included meningioma, schwannoma, and extradural masses in another patient. Involvement of the CNS is well known in VTEC infection, with one patient and a single previouusal lesion. Involvement of the CNS is exceedingly rare in this age group, this lesion was not meningeal based and such lesions are usually reactive for epithelial membrane antigen unlike this tumour. This lesion, unlike some pleomorphic lesions, showed no immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

Solitary intracranial involvement by myofibromatosis includes patients with widespread systemic involvement and multiple leptomeningeal nodules in one patient and extradural masses in another, both of which were fatal at the age of 10 days, a non-fatal extradural mass in one patient, and a patient with systemic involvement, in which there was recurrence of orbital and temporal lesions 2 years after operation. A single previous case of solitary intracranial myofibroma has been reported in which the patient died within 24 hours of surgery, secondary to cardiorespiratory arrest.

We present a patient with a solitary intracranial myofibroma with an excellent postoperative outcome. Although rare, infantile myofibroma should be included in the differential diagnosis of intracranial neoplasms in children.

We acknowledge the expert assistance of Des Lucy, Roarte and Dr Louis Dehner in diagnosing this case.

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Axonal polyneuropathy and encephalopathy in a patient with verotoxin producing Escherichia coli (VTEC) infection

Escherichia coli serotype O157:H7 causes serious food poisoning worldwide, especially in children and elderly people. It is also called verotoxin producing E coli (VTEC), which produces a potent Shiga-like toxin. Gastrointestinal, haemorrhagic, and urometric effects are well known in VTEC infection, and neurological problems are likely to be more frequent than is generally recognised. Here we describe axonal polyneuropathy and encephalopathy in a young female patient associated with haemolytic-uraemic syndrome caused by VTEC infection.

A 26 year old woman began to have abdominal pain and haemorrhagic diarrhoea. She was admitted to an emergency hospital and diagnosed as having haemorrhagic colitis due to probable food poisoning. Then her urinary volume was increased and creatinine serum concentration increased, and she was transferred to our hospital. On the 9th day she had a high fever of 39.7°C with increased C reactive protein to 7.6 mg/ml and a leukocyto-ysis of 17 800/mm³. She was in a state of anuria and her blood analysis showed severe kidney dysfunction (increased serum creatin-ine of 6.76 mg/l). She had severe anaemia (haemoglobin 6.0 g/dl), fragmentation, and tear drop deformation of red blood cells in the blood smear and increased lactate dehydrogenase concentration of 4095 IU (normal range 250–460 IU), suggestive of haemolytic anaemia. Her platelets were also decreased to 21 000/mm³. The culture of her stool showed the growth of E coli O157:H7 and analysis of the bacterial toxins showed the presence of verotoxin, which confirmed the diagnosis of VTEC infection. In her plasma exchange, continuous haemodialysis, and antibiotics (4 g/day fosfomycin, 600 mg/day levofloxicin, and 2 g/day cefoperazon/sulbacam). Her general status was unchanged for 2 weeks after admission and she was in a delirious state with visual hallucinations and tonic convulsion., indicative of encephalopathy. Brain CT disclosed mild brain swelling and there was diffuse slow waves in the frontal and occipital areas. She was given 250 mg/day diphenylhydantoin. During the next two weeks her kidney function, haemolytic anaemia, and encephalopathy gradually improved.

After recovery of consciousness she began to complain of numbness of the limbs, manifested by soars feet. She developed feeling like frost bite when she was lying on the bed, and this gradually exacerbated to be a burning pain. On examination she was alert and cooperative. Her cranial nerves were normal. Muscle strength was normal and coordination was intact. Deep tendon re- flexes were decreased in the four limbs. Sensation for vibration was impaired in the lower legs, but preserved for pin prick, light touch, and joint position. Routine laboratory data including haematological studies, serum chemistry, urinalysis, and CSF analysis were normal. Serum concentrations of vitamin B1, B6, and B12 were normal. Nerve conduction studies were carried out on her right limbs, and showed normal findings in the distal latencies, motor conduction velocities, and F wave latencies of the median, ulnar, and tibial nerves, and no evidence of conduction block. However, there were markedly decreased amplitudes of the sensory nerve action potentials (0.2 μV) and sural (0.98 μV) nerves. These findings and the clinical features confirmed the diagnosis of sensory dominant, axonal polyneuropathy. She was given 300 mg/day salbutam (an anti-inflammatory agent) and 1500 μg/day meclocholamin (vitamin B12) without effect. Two weeks after administration of 300 mg/day oral mexiletin, her numbness and pain gradually disappeared.

The patient was diagnosed as having VTEC infection, because of a typical history of an acute haemorrhagic colitis, the cultured growth of enterohaemorrhagic E coli O157:H7, and the detection of verotoxin in her stool. She had haemolytic-uraemic syndrome (haemolytic anaemia, thrombocytope-enia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, by exposure to different doses of verotoxin through the process of apoptosis. Verotoxin shows similar cytotoxicity on human glomerular mesangial vascular cells via induction of apoptotic pathways such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis. Our patient was treated with antibiotics, plasma exchange, and continuous haemodialysis, with benefit.

During the course of the disease, our patient was in a delirious state with visual hallucinations and tonic convulsion. She showed mild brain swelling on CT and diffuse slow waves in the frontal area on EEG, evidence of encephalopathy. Previous reports have shown that the incidence of encephalopathy in haemolytic-uraemic syndrome (most of VTEC infection) is 1%, including seizures in 17%–44%, altered consciousness in 7%–40%, and paralysis in 1%–16. Many of the patients, including ours, seemed to have metabolic encephalopathy, but some developed encephalopathy without metabolic abnormalities. There is experimental evidence that verotoxin has direct virulence to both endothelial cells and neurons in the nervous system, and its initial lesion is in the hypothalamic areas, then
Crying spells as symptoms of a transient ischaemic attack

In the absence of depression, crying spells associated with neurologic disease usually result from pseudobulbar palsy or, more rarely, from crying seizures. To our knowledge, there are no prior reports of crying spells heralding or signifying a transient ischaemic attack. We report on a patient with prominent cerebrovascular risk factors who had a transient episode of intractable crying and focal neurological findings.

The patient was a 55 year old right handed man who presented with acute, uncontrolled crying spells following by left sided parasthesias. Around 6:00 am he woke with a severe pressure headache and suddenly started crying for no apparent reason. There was no accompanying feeling of sadness. This crying, which involved lacrimation and “sobbing,” abruptly ceased after 5 minutes.

Within 30 minutes of his initial crying spell, his headache had resolved but he became aware of numbness over his left face and numbness and pain in his left neck and arm. The numbness was not progressive, and the patient did not complain of paraesthesias in his trunk or legs. He had photophobia, nausea or vomiting, blurred vision, visual obscurations, difficulty swallowing, dysarthria, or focal weakness. Over the next 2 to 3 hours, he had five more crying spells, each lasting 5 to 10 minutes, occurring out of context, without precipitating factors or sadness, with an acute onset and offset, and without alteration of consciousness. The patient’s left face and arm numbness persisted during and between the episodes and abruptly resolved shortly after his last crying spell. This patient had hypertension, diabetes mellitus, coronary artery disease, an old myocardial infarction, raised cholesterol concentrations, and a history of heavy smoking.

On examination between recurrent crying spells, his blood pressure was 143/92 with a regular pulse of 62, and there were no carotid bruits. His mental status was normal. Cranial nerves examination disclosed a flattening of the left nasolabial fold and decreased pinprick sensation over his left face and arms. His reflexes were +2 and symmetric with downgoing toes.

The patient lacked prior depression, new depressuronic spells, or prior crying spells as an adult except for a single episode during dental anaesthesia. At the time of his admission, he had not had any recent adverse events in his life, and was totally surprised by his reaction.

The patient’s crying spells, paraesthesias, and neurological findings entirely resolved within about 3 hours. Routine laboratory tests, ECG, and CT were normal. Two days after admission, MRI disclosed a mild degree of white matter changes over the right frontal horn, and an EEG showed a frontal intermit-}

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Crying or dacrystic seizures also occur but are rare. These seizures are part of the range of complex partial seizures and usually emanate from the right temporolimbic system.1 Crying seizures may result from prior cerebrovascular infarctions.3 Although our patient had mild twitching of his left face, he had no other evidence suggesting definite seizure activity.

It is likely that this patient had a single transient ischaemic attack with multiple crying spells. The localisation of his attack is unclear; involvement of the right thalamus or neighbouring internal capsule is a possibility. Similar to spells of laughter, spells of crying may occur in relation to unilateral cerebrovascular events. Although most reports of crying after unilateral strokes have reported left hemispheric lesions, crying also may result from right hemispheric strokes.4 Even more similar to our patient, sudden laughing spells, “le fou rire prodromique,” rarely precede strokes involving the left capsular-thalamic, lenticular-caudate, or pontine regions.5 Our patient may have had a comparable phenomenon from the right hemisphere, and the unusual symptom of crying as a pseudobulbar palsy may include dysarthria, or a brisk jaw jerk. The rest of the neurological examination showed mild weakness in his left upper arm, and decreased pinprick and temperature sensation over the left half of his body. Other reflexes were +2 and symmetric with downgoing toes.

The patient lacked prior depression, new depressuronic spells, or prior crying spells as an adult except for a single episode during dental anaesthesia. At the time of his admission, he had not had any recent adverse events in his life, and was totally surprised by his reaction.

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Continuous drop type of orthostatic hypotension

Orthostatic hypotension has usually been evaluated for 2–10 minutes after standing.1 Multiple system atrophy (MSA: Shy-Drager syndrome) is one of the neurodegenerative diseases which show marked orthostatic hypotension. We studied changes of blood pressure for more than 20 minutes after standing in 30 patients with MSA.

The patients lay on their back, on a tilting table, and an intravenous cannula was introduced into the cubital vein more than 30 minutes before the 25 minute test of 60° head up tilt. Blood pressure and heart rate were recorded every minute with an automatic sphygmomanometer. Patients could clearly be classified into two groups in terms of the time taken to reach the minimum blood pressure. In 12 patients systolic blood pressure fell rapidly, reached a minimum within 5 minutes, and then remained stable or partially recovered (early drop type); whereas, in 13 patients blood pressure fell immediately after tilting but kept decreasing by more than 8 mm Hg from that at 5 minutes (mean 12.9 mm Hg;
Continuous drop type of orthostatic hypotension during 25 minute tilt up in a patient with MSA. SBP=systolic blood pressure; HR=heart rate; CO=cardiac output; SVR=systemic vascular resistance; NA=plasma noradrenaline concentration.

maximum 74 mm Hg), taking more than 10 minutes to reach the minimum (continuous drop type) (figure). The other five patients could not remain standing for more than 5 minutes because of symptoms of orthostatic hypotension. Most patients showed the sudden drop in blood pressure and heart rate seen in vasovagal syncope. In the continuous drop type, there were no decreases between 5 and 20 minutes in heart rate (+2.3 bpm) and the noradrenaline (norepinephrine) level (+0.05 ng/ml) during the decrease in blood pressure. A slight increase in packed cell volume (mean=1.4%) was noted between 5 and 20 minutes because of symptoms of orthostatic hypotension. No patient showed the sudden drop type (figure). The other five patients could not remain standing for more than 5 minutes because of symptoms of orthostatic hypotension. Most patients showed the sudden drop in blood pressure and heart rate seen in vasovagal syncope. In the continuous drop type, there were no decreases between 5 and 20 minutes in heart rate (+2.3 bpm) and the noradrenaline (norepinephrine) level (+0.05 ng/ml) during the decrease in blood pressure. A slight increase in packed cell volume (mean=1.4%) was noted between 5 and 20 minutes because of symptoms of orthostatic hypotension.

Our results suggest that in many patients with MSA the blood pressure drops continuously on standing. The continuous blood pressure drop is caused by continuous reduction of cardiac output. A part of the mechanism for continuous reduction of cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline which are caused by interruption of the baroreflex arc, as is known in MSA.

Most patients with continuous drop type orthostatic hypotension reported reduced endurance for more than 10 minutes of exercise (easy fatigability). Two experienced syncope more than 20 minutes after standing.

We used a Swan-Ganz catheter to investigate the haemodynamics in three patients with orthostatic hypotension of the continuous drop type. To prevent the concentration of plasma, saline of calculated volume was infused during tilting. During the continuous decrease in blood pressure, cardiac output proportionally decreased but systemic vascular resistance did not change (figure). Our results suggest that in many patients with MSA the blood pressure drops continuously on standing. The continuous blood pressure drop is caused by continuous reduction of cardiac output. A part of the mechanism for continuous reduction of cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline which are caused by interruption of the baroreflex arc, as is known in MSA.

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We thank Dr Jolobe for his interest in our previous report.1 Although we disagreed with the contribution of Pellecchia et al., we were surprised at the high prevalence of antigliadin antibodies in patients with idiopathic cerebellar ataxia.2 In our study, 20 out of 24 patients with idiopathic cerebellar ataxia had a late onset, and all three of them were affected by celiac disease.3 Furthermore, in our series we found a high prevalence (12.5%) of celiac disease among all ataxic patients.4 This is by contrast with the previous report.1

We agree that some discrepancies arise comparing our study with that of Hadjivassiliou et al.1 Firstly, only six out of their 28 patients had evidence of cerebellar atrophy on MRI, whereas all of our patients had cerebellar atrophy. Secondly, many of their patients had a peripheral neuropathy in the absence of cerebellar atrophy.3 This finding could explain the relatively mild upper limb signs.5 Furthermore, two of our three celiac patients had a clinically silent peripheral neuropathy, which we think that their ataxia was explained by cerebellar atrophy. Thirdly, we found a high prevalence (12.5%) of celiac disease on duodenal biopsy among patients with idiopathic cerebellar ataxia,6 whereas none of the six patients with cerebellar atrophy described by Hadjivassiliou et al.1 showed histological features of celiac disease.7 Finally, we do not think that cerebellar ataxia associated with celiac disease is diagnostically distinctive.8

We understand that some discrepancies might have been more appropriate.9 We were surprised at the high specificity and sensitivity of increased antigliadin antibody titres in their hands. We found both IgA and IgG antigliadin antibodies to be invaluable screening tools in patients with ataxia, only 11 of our 28 patients with increased antigliadin antibodies had histology of overt coeliac disease on duodenal biopsy, the remainder having normal or non-specific inflammatory changes but with an HLA genotype in keeping with gluten sensitivity. It is interesting to note that despite the often quoted high sensitivity for coeliac disease of increased antiendomysium antibody titres, such was found in only one of three patients of Pellecchia et al.1 with coeliac disease.10 This concurs with our impression of very modest sensitivity of antiendomysium antibodies in gluten ataxia.

Gluten sensitivity is common in patients with ataxia, and can be identified by increased antigliadin antibody titres in the presence of appropriate histocompatibility antigens10.11 Although the clinical features of gluten ataxia are not entirely specific, they are distinctive.

**Procarainamide for faecal incontinence in myotonic dystrophy**

We read with interest the article by Abercrombie et al which describes the pathophysiology and surgical management of faecal incontinence in two siblings with severe myotonic dystrophy.1

In the authors’ experience, long term results of both medical and surgical management of the faecal incontinence in myotonic dystrophy were unsatisfactory. Adequate treatment of faecal incontinence and the associated motor disorders is difficult to achieve by any currently available treatment other than colostomy.2 It should be noted, however, that the medical treatment used is not specified in the text.

Our experience with medical treatment using procainamide in a patient with severe myotonic dystrophy and faecal incontinence is less disappointing.3 The patient—a 19 year old man—had had his illness diagnosed 4 years earlier on clinical grounds and electro-physiological and genetic tests. Early symptoms of sphincteric impairment developed soon after, including mild stress urinary incontinence and minor episodes of poor control of loose stool.

A complete diagnostic investigation, including physical examination, defecography, and electromyographical tests of pelvic floor musculature, was performed. At physical examination, digital anorectal evaluation showed low squeeze pressures. A reduced rectal diameter (4.5 cm), anal gaping, and barium loss at rest were found in defecography. Motor evoked potentials elicited by cortical and lumbar magnetic stimulation and recorded from the external anal sphincter showed a normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed a normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed, in the first patient of Abercrombie et al, a decreased number of motor units and multiple myotonic discharges. Few motor unit potentials presented polysympathetic waves and decreased duration and amplitude.

A regular treatment with procainamide (300 mg twice a day) led to a dramatic improvement of both systemic myotonia and faecal incontinence. A 13 month follow up assessment has shown a stable clinical improvement. Repeated electromyographical investigation showed disappearance of myotonic discharges at the external anal sphincter, whereas defecography disclosed an improved rectal compliance (5.2 cm in diameter) at capacity and no more than a barium leak on straining.

The pathophysiology of motor disorders of the gastrointestinal tract in myotonic dystrophy is still debated and controversial. Histological study of the external anal sphincter and...
the EMG pattern in patients with myotonic dystrophy show a multitude of defects including expression of myotonia, myopathy, muscular atrophy, and neural abnormalities. 1,2 

The possible management of myotonia and some of its clinical manifestations, such as dysfunction, 1,2 in myotonic dystrophy (dystrophy-poly-pamida and procarinamide), justifies the use of the same pharmacological approach in anal sphincter dysfunction manifested in a few cases of myotonic dystrophy.

We conclude that treatment of faecal incontinence with procainamide should always be attempted before any surgical option in patients with myotonic dystrophy.

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4 Eckardt VF, Nix W. The anal sphincter in dystrophies of polyneuromyogenic origin, and called it chronic anterior polyomielitis. The patient showed symptoms of proximal amyotrophy, and signs of denervation and upper motor neuron involvement. Since then, in those countries and other countries under their influence, 1,2 we have come to use the eponym of Vulpian-Bernhardt’s syndrome to describe those forms of amyotrophic lateral sclerosis with more or less symmetric involvement of the proximal muscles of the upper limbs at the clinical onset. A certain enigma exists surrounding the characteristic distribution of weakness and muscle atrophy. The reason for the preva-

lence in the proximal muscles of the upper limbs is unknown. We can furnish little more information in this respect. However, in the 1960s, in the differential diagnosis of this syndrome, it was established that the muscles predominantly affected in Vulpian-Bernhardt’s form were the deltoideus, the infraspinatus, the supraspinatus, the sternocleidomastoideus, and the teres minor. The predominant affection of the elbow flexors in these mus-

cles permitted its distinction from that previously called Erb’s dystrophy. 3 As a consequence of the atrophy of these muscles, the upper limbs adopt a characteris-
tic position, with the shoulders slumped, and the arms, forearms, and hands in pronation.

As the illness progresses, the hand muscles are affected, with atrophy of the following muscles: opponens pollicis, flexor brevis, abductor pollicis brevis, adductor pollicis, interossi, and lumbricales, which leads to the formation of the characteristic Aran-Duchene hand.

Obiously, signs of cortisocinal involve-

ment with hyperreflexia in the lower limbs and Babinski’s sign both appear. In the initial stages of the illness, there is no effect on the dia-

phragm. The presence of signs of involve-

ment of the upper motor neuron, its different clinical evolution, and the data supplied by genetic molecular investigation allow us to distinguish the syndrome previously known as Vulpian-Bernhardt’s as also a variant of torticollis as flail arm syndrome from other motor neuron syndromes such as of the spinal muscular atrophies, Kennedy’s disease, multifocal motor neuropathy, and monomelic amyo-
trophy.

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Pain after whiplash

This latest study from Lithuania is an answer to many questions—namely, that the previous difficulties that these researchers had with identifying the late whiplash syndrome in Lithuania is that they were not looking “in the right place”. As it turns out, the problem is that Lithuanians simply are not behaving the way many in western coun-

tries, underlies whiplash associated disorders. Just like there. There are some methodological issues which can be considered, as below, but the lesson of discarding “unsightly” data because it is too disturbing to one’s personal view and vested interest in the whiplash controversy has already been taught elsewhere. Suffice it to say that the truth has been laid bare and we (those of us struggling with epidemic proportions of the late whiplash syndrome in our own countries) now need to enlighten ourselves and put this data to practical use in helping whiplash patients rather than resisting the inevitable.

After completion of the first historical cohort study, this more recent study selects an entirely separate, distinct sample of these “misbehaving” Lithuanians, but in a more intriguing fashion. This is the first true inception cohort study with people who have not been preselected by their attendance at emergency departments, or contaminated by therapists or lawyers, can be studied to appreciate the natural evolution of the injury which, underlies whiplash associated disorders grades 1 and 2. This is the study’s greatest strength. The study has, however, its limitations.

The first consideration is that there were 98 accident victims who reported acute symptoms, and thus were at risk for the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome.

The Swiss study may be useful for compari-

son because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by the selection atrocity of advertising with subjects, and has a host of other reportedly fatal faults, and giving some benefit of the doubt, the study is said to be an accurate representation of the state of affairs in Swit-
zerland at that time, in Switzerland, not even 60% manage to recover fully by 3 months and many of these were reporting total disability during that time, whereas the Lithuanians fully recover in 4 weeks or less, with little or no therapy, and no disability.

A few countries) now need to enlighten

studies in other western countries disclose an even greater contrast, with 50%–70% of patients reporting pain even after 3–6 months, despite the fact that all these studies are examining the same diseases (1 and 2) of whiplash associated disorders. 

Thus, while the sample size is small in this Lithuanian study, it is comparable with others reporting the prognosis of whiplash, and yet gives a different picture of outcome.
A second consideration is that perhaps these Lithuanians are in very minor collisions. True, some of their vehicles were completely wrecked, but perhaps the vehicles were not very good quality and so were easily damaged. Perhaps that is why this cohort had such a good outcome and only minor injuries. This is an unhelpful consideration however, as studies in Canada have shown that with absolutely no vehicle damage, in very low velocity collisions, are just as likely to completely wrecked, but perhaps the vehicles 

A chapter on acute postoperative pain management is well written and informative as are the chapters on obstetric and paediatric pain. The chapter on chronic low back pain by Rauck is one of the best I have seen for some time. It is a comprehensive review of both acute and chronic low back pain. It is as excellent as it also mentions treatments that are often performed outside the medical specialist arena. I was pleased to see in it the mention of some of the newly evolving techniques such as facet denervations, spinal cord stimulation, and disc denervation. It was a pity that the randomised control trials which have shown facet denervation to be an outstandingly useful technique for chronic back pain were not mentioned. It was also a pity that the reference to the disc denervation procedure was to another text book rather than any original papers.

The chapter on cancer pain management has been written by internationally known authors and is an excellent summary of the subject. In the section on interventional pain techniques the emphasis was on spinal cord stimulation, radiofrequency, and cryosurgery. Again this chapter has been written by an internationally well known author who concentrated on general overview of the techniques rather than a how to do it approach, which I think was needed to make it bigger to a bigger for. In summary I think that this volume would make an excellent addition to the bookshelf of those involved in the treatment and management of pain.

RAJESH MUNGLANI

BOOK REVIEWS


This book purports itself to be a comprehensive reference. Certainly the title would suggest so. However, it is clear that this is not a comprehensive text, but a book that is an update on particular timely topics in the field of pain medicine. There are sections on pain mechanisms, the first chapter on the pharmacology of acute and chronic pain, and other chapters on postoperative pain, obstetric pain, and acute paediatric pain. There are three further chapters specifically on the management of chronic low back pain, cancer pain, and an overview of interventional pain techniques.

Many of the authors are internationally known and this is perhaps the book’s strongest point—one does get a state of the art review and to this end I warmly welcome this book as an addition to the bookshelf to update a busy anaesthetist or pain specialist, though the chapter on chronic low back pain and cancer pain will also be of interest to those in other fields.

The chapter on the anatomy and physiology of pain is excellent in that it has clear explanations and a number of very helpful diagrams. Unfortunately it fails to mention increasing understanding of the role of GABA in mediating analgesia within the spinal cord and furthermore does not mention some of the other neuroplastic changes which are well known to occur in chronic pain states such as central sprouting and phenotypic switching.

The chapter on pharmacology of acute and chronic pain is well written, but unfortunately a lot of time is spent on non-steroidal drugs. There is a review of the adjuvant drugs such as antidepressants and anticonvulsants that are used in chronic pain, however one is left at the end with a sense of knowing about the drugs but not quite to use them. There is no mention of the increasing use of gabapentin nor of other drugs that are sometimes used in chronic pain states such as clonidine and other sympatholytic agents or calcium channel blockers.

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RAJESH MUNGLANI


This is a really excellent book which is both comprehensive and amazingly up to date, with the inclusion of many references from as late as 1997.

As a clinical neurologist and neuropsychologist with a longstanding interest in the dementias, I found it extremely valuable. The editor has done a very good job in posing a coherence, format, and style, which is often lacking from multicounter textbooks.

The title of the book is perhaps a little misleading in that the book includes, as well as traditional neuropathology, a very comprehensive overview of the molecular biology and genetics of the dementias. As would be expected, a considerable proportion of the book is dedicated to Alzheimer’s disease with chapters on both the clinical features, genetics, and the neuropathology. The frontotemporal dementias are also well covered and the book includes a chapter on the opisthions related to chromosome 17 linked dementias. There are also sections on progressive supranuclear palsy, Huntington’s disease, corticobasal degeneration, dementia with lewy bodies, and prion diseases and vascular dementia.

The editor has managed to persuade many of the world’s experts to contribute. For instance, the chapter on prion diseases is by D’Almond and the recent Nobel laureate Prusiner, and the frontotemporal dementias are reviewed by Brun and Gasteron. Genetics of Alzheimer’s disease are dealt with by St George-Hyslop and the neuropathology of Alzheimer’s disease by Price and coworkers.


The standard of illustrations is excellent and the style generally very readable. I shall certainly find it extremely useful as a work of reference and for teaching purposes. The editor is to be complimented on producing such a delightful work.

JOHN HODGES


I very much enjoyed reviewing this textbook of instrumented spinal surgery written by Giuseppe Tabasso under the auspices of Jürgen Harms. Dr Harms is well known to all spinal surgeons and has made a very important contribution to the development of spinal surgery over the past 20 years, based on strong personal convictions. Many surgeons who manage spinal disorders would not choose to implement all of Professor Harms' solutions but all who have a serious interest in the surgical treatment of the spine admire and are grateful for his contribution. Within this book spinal surgeons will find a rational and practical approach which will allow them to treat a wide range of spinal disorders according to well thought out principles.

The opening chapter describes spinal biomechanics under normal and pathological circumstances mainly by using easily understood drawings and diagrams. Some of these drawings reminded me of images that I have recently seen on an interactive CD ROM that I have bought for my 4 year old son. This is not a criticism and I fully support any attempt to simplify the science of biomechanics which is often cloaked in seemingly contradictory jargon. Most spinal surgeons will be able to assimilate the two basic principles which underpin much of instrumented spinal surgery—namely, that the anterior column resists load compression forces and that the posterior column acts as a tension band which when disrupted should be reconstituted in compression. The remaining chapters cover fracture management, late kyphosis, metastatic tumours, spondylolisthesis, degenerative spinal disease, and infection. Each chapter sets out the principles of management which are illustrated schematically. There then follow case studies illustrated by radiological images including CT and MRI. These have reproduced well and surgeons will admire the technical precision and excellent anatomical reductions illustrated by these clinical cases. It is, however, a source of constant annoyance to spinal surgeons that perfect postoperative films do not always correlate with good clinical results and this discrepancy remains a source of fascination and mystery.

It is in the degenerative spine that this discrepancy between radiological and clinical findings is most apparent and it is partly for this reason that the management of these conditions is often controversial. It is difficult to disagree with much of the logic presented by the authors in planning their interventions but there is a danger that inexperienced surgeons may be misled into adopting complex solutions when often more simple operations will suffice. The authors' description of their approach to failed back surgery syndrome illustrates this problem and the inadequacies of attempting to treat a complex clinical problem by focusing on one aspect of it.

This book will be a useful addition to the shelves of spinal surgery textbooks and many orthopaedic and neurosurgical departmental libraries will wish to buy a copy.

RODNEY LAING


I wondered, when I received this book, how I could possibly say anything adverse about a book written by three such world renowned experts. I have heard them all lecture often and have seen them all at work. They have a vast knowledge and experience of treating disorders of peripheral nerves. In clinic and the operating theatre, they have shown myself and many trainees a clarity in their planning of management of complex problems that humbles one's own thoughts. That clarity has continued in this text book of over 500 pages. The field of peripheral nerve surgery is covered comprehensively, commencing with descriptions of anatomy, physiology, and pathological reaction to injury. This is followed in subsequent chapters with descriptions of approaches to virtually all the main peripheral nerves, and the operative management of brachial plexus injury and outcomes is covered in three detailed chapters. These are followed by chapters on nerve entrapment, neuropathy, iatropathic injury, and neoplasm within the peripheral nerve. The final section covers electrodiagnosis, pain, nerve recovery, reconstruction techniques, and rehabilitation.

The text is well written, easy to read, and supplemented by some excellent line drawings similar to those used in Lundborg's text. There are detailed plates showing histology and various imaging techniques. Each chapter is comprehensive, containing important historical aspects as well as up to date techniques, and there is an extensive reference section. I would recommend that trainees of all specialties dealing with peripheral nerve injuries should read much of this text and it would be extremely useful as a regular reference. It would also make an important and necessary addition to most medical libraries. All clinicians would be well advised to read the chapters on iatropathic injuries, not only for the extensive causes of such injuries encompassing all medical and surgical departments, but also for the précis of the changes occurring in medical negligence claims. This text represents good value for money.

IAN WHITWORTH