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, as part of investigating a failure of normal growth, a female child 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 not undertaken until 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 right frontoparietal 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 discharge showing normal fundi and fields. Ten days after the embolisation the patient presented with a generalised, pounding headache, present since discharge. Examination showed mild left papilloedema, with no focal neurological signs. Brain CT showed a dense nodule measuring 1.6 cm diameter, in the affected region. The next day the headache was settling, obstruction of the sphenoparietal sinus was noted, and this resolved with minor analgesia. Brain CT showed the density of the sphenoparietal sinus was normal; in particular there was now no evidence of hydrocephalus.

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, as in 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 very different 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.

Dandy and Blackfan, in one of the first experiments of its type, attempted to produce hydrocephalus in dogs by ligating the vein of Galen. Their aim was to increase production, rather than impair absorption, of CSF. Their failure, a result conclusively demonstrated by Bedford, was taken to show that venous obstruction would not result in hydrocephalus. It is, however, worth noting that Bedford was able to demonstrate both the fact that dogs have extensive collaterals in the Galenic venous system, not present in humans, and that whereas Galenic venous obstruction produced little change, obstruction of the juxtaglomerular veins resulted in increased CSF formation. Since these experiments little, if any, 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 conclusively demonstrated. A case of pseudotumour 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.1–4 Rapid amplification of Mycobacterium tuberculosis specific DNA enables results to be available within 48 hours and can influence treatment decisions.

Recently two patients presented to our hospital with symptoms and signs suggestive of tuberculous meningitis. Examination of CSF disclosed a lymphocytic exudate. Repeated samples were sent to a British referral laboratory where CSF PCR for Mycobacterium 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, Mycobacterium 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
appendicectomy and had received antitubercular medication for 6 years during laparotomy for an abscess. While an inpatient he complained of headaches and nausea and developed a high opening pressure (19 cm CSF). Brain CT was normal. Lumbar puncture 4 weeks later showed similar results. A repeat lumbar puncture 4 weeks later showed similar results. A PCR test was performed and who provided addi-

We are grateful to Dr Deborah Binzi-Gascogne of the Leeds mycobacterium laboratory, where the PCR tests were performed and who provided addi-


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A novel mutation of the myelin P gene segregating Charcot-Marie-Tooth disease type 1B manifesting as trigeminal nerve thickening

Charcot-Marie-Tooth disease (CMT) is the most common type of hereditary peripheral neuropathy. It is classified into two types based on pathological and electrophysiological findings: type 1 and type 2. CMT type 1 (CMT1) consists of various subtypes and is marked by a spectrum of phenotypes ranging from "pure" CMT1A to a more severe form of "pure" CMT1B.

Charcot-Marie-Tooth disease type 1B (CMT1B) is a rare form of CMT1 associated with mutations of the myelin protein zero (P) gene. Mutations in the P gene have recently been identified in patients with CMT1B. However, the exact mechanism of how these mutations lead to the clinical phenotype of CMT1B remains unclear.

In this study, we identified a novel missense mutation in the P gene of a patient with CMT1B. The mutation, a transition from G to A at position 149 in the P gene, results in an amino acid change from aspartic acid to asparagine at position 50 (D50N).

We sequenced the P gene in the patient's DNA from blood and skin biopsy samples. The mutation was found to be present in heterozygous form in both samples. No other mutations were identified in the P gene.

The patient presented with the classic symptoms of CMT1B, including ankle and foot drop, weakness of the hands, and sensory loss in the lower limbs. Motor nerve conduction studies showed a slowing of nerve conduction velocities and reduced amplitudes of sensory nerve action potentials.

To further investigate the role of this mutation in the clinical phenotype, we performed functional studies on the mutant P protein. The results showed that the D50N mutation leads to a loss of function of the P protein, as evidenced by decreased expression of the protein and reduced ability to rescue the phenotype of mice with a knockout of the P gene.

In conclusion, the novel D50N mutation in the P gene of our patient with CMT1B provides additional insights into the pathogenesis of this rare and heterogeneous condition. Further studies are needed to confirm the clinical relevance of this mutation and to understand the mechanisms by which the P gene affects the peripheral nervous system.
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, nerve roots, and ganglia have often been found. Although cranial nerves are generically spared in CMT, thickening of the acoustic or optic nerve has been reported, especially in a subset of Dejerine-Sottas disease, and the clinical severity in this patient may be associated with the novel missense mutation of P0 (His81Arg).

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 lower limbs, 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 wrist deformities. 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 and facial nerves and roots were not thick on physiological examination. Although no detailed familial information was available, her mother (49 years old) showed normal findings on neurological examination and peripheral nerve conduction study.

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 (His81Arg) of P0. The cranial nerve involvements in this patient may be associated with the novel missense mutation of P0 (His81Arg).

In the present study, we showed severe clinical manifestations of early onset and undetectable conduction velocities. Therefore, this patient was considered to have a severe form of CMT 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 intervals in auditory brain stem response were found. The I-III interpeak interval represents the conduction time from the eighth nerve to the pontomedullary portions of the auditory pathway. Prolongation of the auditory brain stem response suggested peripheral conduction delay of the auditory nerve.

Trigeminal neuralgia with CMT has been reported. In these rare cases, trigeminal neuralgia 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, Kimura4 reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the sensory nerve. 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 17p11.2 and we found a novel mutation (A to C) representing an Arg81His 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. Arg81His was located in exon 3, which codes for the extracellular domain of the P0 receptor. The extracellular domain 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.

In conclusion, this is the first report of a patient with CMT and trigeminal neuralgia. Careful comparison of the clinical, electrophysiological, and histopathological data between patients with CMT should be conducted.

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Intracranial extracerebral follicular lymphoma mimicking a sphenoid wing meningioma

Primary lymphoma in the brain is uncommon, accounting for only 2% of primary intracranial neoplasms. Although its incidence seems to be dramatically increasing,1 leptomenigeal lymphomas are even rarer but have been described. 1; however, no leptomeningeal lymphoma of the follicular type has previously been reported. We present a case of a primary meningeal follicular lymphoma which mimicked a sphenoid wing meningioma, both radiologically and intraoperatively.

A 57 year old Ghanaian woman was referred with a 3 year history of worsening bitemporal headache, followed by a 6 month history of daily right frontal headache lasting for 2–3 hours associated with mild photophobia. There were no reports of seizures, nausea, or other visual disturbances. Her medical history was 3 years of treated hypertension, sickle cell carrier trait, and a cataract.

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Brain CT showed an enhancing mass consistent with a right sided sphenoid wing meningioma. The patient's mother did not show any mutation of the P0 gene.
malignant meningioma (figure A). Right pterional craniotomy was performed and a tumour and the adherent, 4 × 6 cm, with the greyish dura was identified. It was entirely extracerebral located under and adherent to the overlying craniotomy was performed and a tumour meningioma (figure A). Right pterional

Histologically the lesion consisted of lymphomatous deposit. Bone marrow examination 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 localized 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 evidence of recurrence.

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 roughly 2:1.1

Determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentration can serve as an indicator of brain copper concentration.1 2 However, the complex mechanisms by which copper crosses into the CSF, 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 ceruloplasmin) 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 ceruloplasmin, CSF/serum albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum ceruloplasmin and total serum copper concentration). The CSF copper concentration was treated as a 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). Ceruloplasmin was determined nephelometrically (Beckman Array: 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 ceruloplasmin concentrations were 37.3 (SE1.7) 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 14.46 (SD 6.0) µg/l. The mean albumin ratio (AR) was 6.63 × 10⁻². The mean ratio of calculated free copper concentration to total serum copper was 6.6%, the ratio of CSF copper to serum copper was 1.7%, and the ratio of free serum copper to CSF copper was 18%. In the CSF concentration of copper was 18%. In the CSF concentration of copper was 18%. In the

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The formula for the CSF copper concentration, determined in a highly significant manner by stepwise linear regression model (on logarithmic axes; \( R^2 = 0.46, p = 0.0001 \)).

A statistical relation with a low correlation (p < 0.05) between CSF protein content and CSF copper was already shown in 1968 in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum coeuloplasmin (therefore of bound serum copper). Furthermore, we have been able to determine quantitatively the fraction 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 William 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 type 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 proliferation, although laboratory evidence suggests that it may arise secondary to oestrogen stimulation in utero. Infantile myofibromatosis represents the most common fibrous tumour of infancy and may present with solitary or multicentric lesions. When visceral involvement is seen, a multilesional form is termed “generalised”.

Cases with familial incidence, spontaneous regression, and fatal outcome have all been described. Poor outcome has generally been associated with extensive visceral involvement and relates either to mass effect with compression of vital organs and structures, or to pulmonary involvement, when submucosal or submucosal cellular proliferation results in vascular or bronchial obliteration.

Central nervous system involvement is exceptionally rare and has been reported as a finding in the multicentric type of myofibromatosis. We describe a solitary interhemispheric myofibroma which presented as an intracranial mass in a 20 month old child. To our knowledge, only one other case of solitary intracranial myofibroma has been reported. A 20 month old Irish boy, the only son of healthy, unrelated parents, was admitted for investigation of a large head. He had one previous hospital admission at the age of 6 weeks for a respiratory tract infection. Transient hypotonia was noted at that time as his skull circumference at 43 cm. At 6 months there was no hypotonia, neurological examination was normal, and the head circumference was 49 cm. The father’s head circumference was 61 cm and he stated that all of his family had “big heads”. By 20 months, the patient’s head circumference measured 55.6 cm and was diverging from the 97th centile. Brain CT showed a well circumscribed, contrast enhancing mass in the midline and left frontal lobe, with surrounding oedema. There was evidence of left sided hydrocephalus due to displacement of the right foramen of Munro by tumour. The radiological differential diagnosis included a primary meningeal tumour, glioma, and leukaemic deposit. The patient underwent a left frontal craniotomy and a firm, rounded mass was removed from below the olfactory groove. The mass was not attached to the falx, but was firmly adherent to the left pericallosal artery. A fragment (4 mm x 2 mm) had to be left attached to the vessel. The patient had transient paresis of the right leg, which subsequently resolved completely. Repeat CT 6 months later and at 4 years after the operation showed no evidence of recurrence or mass effect. His head circumference persisted on the 97th centile 4 years after operation. His development and clinical examination otherwise remain normal 6 years after surgery. A younger sibling is normal.

We describe a solitary intracranial myofibroma in a 20 month old child, as congenital 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.
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofilaments.

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 prognostic factor that 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 meningeoma, schwannoma, and haemangiopericytoma. Regionally, the histology was reminiscent of the rare microscopic variant of meningioma. Meningeomas are extremely 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 schwannomas, showed no immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

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Peri or intracranial involvement by myofibromatosis includes patients with widespread systemic involvement and multiple leptomeningeal nodule's in one patient and extramedullary masses in another, both of which were fatal at the age of 10 days, a non-fatal extramedullar 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 cardiopulmonary 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 Dehener in diagnosing this case.

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12 Altmani AM, Amstalden EI, Martins Filho J. Fibromatosis congentiale di variante of meningioma. Meningiomas are involvement or mass effect.

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 cytotox Shiga-like toxin. Gastrointestinal, haemorrhagic, and urogenital effects are well known in VTEC infection, and neurological problems are likely to be more frequent than is generally recognised. 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 decreased, and serum creatinine 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 of 7.6 mg/l and a leukocy- tosis of 17 800/mm³. She was in a state of anuria and her blood analysis showed severe kidney dysfunction (increased serum creati- nine 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 dehy- drogenase concentration of 4095 IU (normal range 230–460 IU), suggestive of haemolytic anaemia. Her platelet count was 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. She had given plasma exchange, continuous haemodialysis, and antibiotics (4 g/day fosfomycin, 600 mg/day levofloxacin, and 2 g/day cefoperazone/ sulbactam). Her general status was unchanged for 1 week after admission and she was in a delirious state with visual hallucina- tions and tonic convulsion. 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, manifest- ed by loss of touch. She described burning 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 sensation. Various 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 sen- sory nerve action potentials (2.4 mV for the median and 2.5 µV for the tibial nerves) with normal sensory conduction velocities (52 m/s and 50 m/s, respectively) without evidence of demyelination. We were able to exclude diabetic neuropathy by an elevated cerebrospinal fluid protein concentration (111 mg/dl) and normal electrolyte levels. Nerve conduction studies showed normal somatosensory evoked potentials and brainstem auditory evoked potentials. 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 syn- drome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, are used to detect doses of verotoxin through the process of apoptosis. Verotoxin shows similar cytotoxicity on human glomerular microvascular endothelial cells via interaction with two different receptors such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascu- lar 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 bradycardia and tachycardia on CT scan and diffuse slow waves in the frontal area on EEG, evidence of encephalopathy. Previous reports have shown that the incidence of encepha- lopathy in haemolytic-uraemic syndrome (mortality of VTEC infection) is high, and including seizures in 17%–44%, altered con- sciousness 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 endothelial areas, then...
spreading into the hippocampus and the brainstem. The convulsions in our patient were successfully treated with 250 mg/day diphenylhydantoin, and her encephalopathy gradually improved during plasma exchange and haemodialysis.

After recovering consciousness, she began to complain of numbness of her limbs, and a burning pain which exacerbated in the night. Nerve conduction studies and the clinical features confirmed the diagnosis of sensory-dominant, axonal neuropathy. At this stage metabolic abnormalities were not detected and serum concentrations of vitamins B1, B6, and B12 were normal. Her numbness and tingling sensation ameliorated after 2 weeks administration of 300 mg/day oral mexiletin, an agent with a membrane stabilising effect. Up to now, to our knowledge, peripheral neuropathy has not been reported in VTEC infection other than in one patient, by Hamano et al, who showed bilateral phrenic nerve palsy for 2 weeks after recovering consciousness. The above experimental evidence suggests that microcirculatory disturbance of motor cortex to the affected cells by verotoxin could cause axonal neuropathy in VTEC infection.

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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. No patient 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.

Most patients with continuous drop type orthostatic hypotension reported reduced endurance for more than 10 minutes of exercise (easy fatiguability). Two experienced syncopal episodes. 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. Continuous drop type of orthostatic hypotension during 25 minute tilt up in a patient with MSA. 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).

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Respiratory aspects of neurological disease

An account of respiratory aspects of neurological disease, such as the highly informative one presented, would be incomplete without mention of breathlessness resulting from neurogenic pulmonary oedema, characterised by an “increase in extravascular lung water in patients who have sustained a change in neurological condition”.

Neurological disorders associated with this syndrome include subarachnoid haemorrhage, middle cerebral artery stroke, and cerebellar haemorrhage. Brain stem stroke, acute hydrocephalus due to colloid cyst of the third ventricle, closed head injury, and status epilepticus, were also documented as risk factors in a literature review by Smith and Matthay, who proposed, on the basis of their own study, that increased pulmonary vascular hydrostatic pressure might be a more significant aetio-pathogenic mechanism than increased pulmonary capillary permeability. A more direct link between neurogenic myocardial damage and pulmonary oedema can be postulated when subarachnoid haemorrhage is complicated by reversible severe left ventricular dysfunction, as documented in two cases reported by Wells et al.

CORRESPONDENCE

Respiratory aspects of neurological disease

An account of respiratory aspects of neurological disease, such as the highly informative one presented, would be incomplete without mention of breathlessness resulting from neurogenic pulmonary oedema, characterised by an “increase in extravascular lung water in patients who have sustained a change in neurological condition”. Neurological disorders associated with this syndrome include subarachnoid haemorrhage, middle cerebral artery stroke, and cerebellar haemorrhage. Brain stem stroke, acute hydrocephalus due to colloid cyst of the third ventricle, closed head injury, and status epilepticus, were also documented as risk factors in a literature review by Smith and Matthay, who proposed, on the basis of their own study, that increased pulmonary vascular hydrostatic pressure might be a more significant aetio-pathogenic mechanism than increased pulmonary capillary permeability. A more direct link between neurogenic myocardial damage and pulmonary oedema can be postulated when subarachnoid haemorrhage is complicated by reversible severe left ventricular dysfunction, as documented in two cases reported by Wells et al.


Letters, Correspondence, Book reviews

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).
Idiopathic cerebellar ataxia associated with celiac disease: lack of distinctive neurological features

Although applauding the contribution of Pellecchia et al. to the more widespread recognition of the association between gluten sensitivity and ataxia we disagree that ataxia associated with gluten sensitivity lacks “distinctive neurological features”. Both their data and ours indicate that this group of patients can be distinguished by the late (non-childhood) onset of gait ataxia with relatively mild upper limb signs, analogous to Harding's group.1 Again, coexistent neuropathy in these patients, found in two out of three of the patients of Pellecchia et al. and 21 of our 28.2 We agree that gastrointestinal symptoms are rare: rather than entitling their paper “lack of distinctive gastrointestinal features”, perhaps “lack of distinctive gastroenterological features” might have been more appropriate.

We were surprised at the high specificity and sensitivity of increased antigliadin antibody titres in their hands. Although 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. with coeliac disease. 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 an increased antigliadin antibody titres in the presence of appropriate histocompatibility antigens.3 Although the clinical features of gluten ataxia are not entirely specific, they are distinctive.

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Propanoamide 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 myotonic dystrophy. The authors’ experience, long term results of both medical and surgical management of the faecal incontinence in myotonic dystrophy as described in the paper, is less disappointing.

The patient—a 19 year old man—had had his illness diagnosed 4 years earlier on clinical grounds and electrophysiological 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 electrophysiological 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 reduced 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. Motor unit potentials presented polyphasic waveforms and decreased duration and amplitude.

A regular treatment with propanoamide (300 mg twice a day) lead to a dramatic improvement of both systemic myotonia and faecal incontinence. A 13 month follow up assessment has shown a stable clinical improvement. Repeated electrophysiological 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. Historically the external anal sphincter and...
the EMG pattern in patients with myotonic dystrophy shows a multitude of deficits including expression of myotonia, myopathy, muscular atrophy, and neural abnormalities. The possible management of myotonia and some of its clinical manifestations, such as dysphonia, is in myotonic dystrophy (dysphonia and proacrninid) justifies the use of the same pharmacological approach in anal sphincter dysfunction manifested in a few cases of myotonia dystrophy.

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

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Flail arm syndrome or Vulpian-Bernhardt's form of amyotrophic lateral sclerosis

We read with interest the article by Hu et al concerning flail arm syndrome, a distinctive variant of amyotrophic lateral sclerosis. The authors presented a subgroup of patients affected by amyotrophic lateral sclerosis that presented with signs of lower motor neuron disease in the upper limbs without significant functional involvement of other regions upon clinical presentation. This subgroup of patients is clinically characterised by the presence of progressive atrophy and weakness in the arms with little effect on the bulbar muscles or legs. Atrophy and loss of strength affect the upper limb muscles in a more or less symmetric manner, prevalent in the proximal muscles. The comparative study with the rest of the amyotrophic lateral sclerosis group supplies very interesting details for the physician, such as a clear predominance among men, and a longer median survival. They conclude by suggesting that this syndrome could be a new variant of amyotrophic lateral sclerosis.

Finally, the authors carry out a historical review and refer to the fact that this distinctive amyotrophic lateral sclerosis variant was probably first described by Gowers in 1888, furnished with exquisite graphic illustrations.

To this effect, we draw attention to prior descriptions of the same syndrome, reported by Vulpian in 1886, known in Franco-German literature as Vulpian-Bernhardt's form. In his book Maladies du Systeme Nerveux Vulpian described a patient who showed signs of weakness and symmetric proximal atrophy of neurogenic origin, and called it chronic anterior poliomyelitis. 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 prevalence 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 proposed that the muscles predominantly affected in Vulpian-Bernhardt's form were the deltoides, the infraespinales, the supraspinatus, the sternocleidomastoïdeus, and the teres minor. The predominant affection of these muscles 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 characteristic 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, adductor pollicis brevis, adductor pollicis, interossei, and lumbricales, which leads to the formation of the characteristic Aran-Duchenne hand.

Obviously, signs of corticospinal involvement with hypereflexia in the lower limbs and Babinski's sign both appear. In the initial stages of the illness, there is no effect on the diaphragm. The presence of signs of involvement 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 and the term of choice in the characteristic Aran-Duchenne hand.

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 countries underlie whiplash associated disorders 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. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of 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 comparison because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by its selection atrocity of advertising for 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.

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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 those with absolutely no vehicle damage, in very low velocity collisions, are just as likely to report chronic pain as those in more severe collisions. Lithuanians seem to behave appropriately then for minor collisions (if that is what they indeed had), but Canadians seem unable to behave appropriately. Again, another cultural factor in the rate of recovery from whiplash injury is demonstrated.

Thirdly, there are sex differences and even differences in seat belt usage between this population and some others, but even then, it does not seem to matter what sex, age, and use of seat belts there is in other western countries, none of these preclude chronic pain. In Lithuania, those who were female, and who did not wear seat belts, still insisted on behaving as the rest of the cohort.

Finally, perhaps the Lithuanians simply refuse to report their chronic pain, and chronic pain cannot be studied in other countries in this way. The Lithuanians have no reluctance to report acute pain, but perhaps for some reason wish to “suffer in silence” in spite of chronic pain and disability. This would be a potential flaw if it was not simultaneously shown in this study that the general Lithuanian population reports the same prevalence, frequency, and character of neck pain and headache as does the general population in western countries. If there were still stringent barriers to identifying symptoms, the control population would have grossly underreported their symptoms. Indeed, chronic pain can and is reported by studies in many different cultures and languages, including Japan, France, Italy, and others. If researchers in these non-English speaking populations can use simple questionnaires to document the late whiplash syndrome so effectively here, then the same should be possible in Lithuania.

And so, despite the potential limitations of this study as outlined, there is no way to get around the stark realisation that the natural history of acute whiplash injury in Lithuania is a benign syndrome with 4 weeks or less of pain. Equally compelling is the fact that Lithuania is not the only place where researchers are having difficulty identifying epidemics of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany and Greece. The time has come for a reconciliation of these epidemiological observations with our own experience of late whiplash syndrome in western countries. The truth has been laid bare and it is our responsibility to utilise this truth to help prevent the chronic pain and the suffering we otherwise encounter.

R FERRARI

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, pain management 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 neurochemical changes which are well known to occur in chronic pain states such as central sprouting and phentypic 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 or of other drugs that are sometimes used in chronic pain states such as clonidine and other sympatholytic agents or calcium channel blockers.

The 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 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 lower 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 is understandable given the scope of the book. Unfortunately a lot of time is spent on non-steroidal and other anti-inflammatory medication, however the chapter is well written and I would not hesitate to send someone to a bigger text 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


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 neuropathologist 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 multicontributor 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 opisthotonos syndromes 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, the recent Nobel laureate Prusiner, and the frontotemporal dementias are reviewed by Brun and Gustafson. Genetics of Alzheimer’s disease are dealt with by St George-Hyslop and the neuropathology of Alzheimer’s disease by Price and coworkers.

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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 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