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 abnormal 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 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 discharge showing normal fundi and fields. It was thought to represent the thombosed varix and possibly thrombosis of the vein of Galen and straight sinus. There was no evidence of hydrocephalus.

At lumbar puncture several days later opening pressure was 27 cm H₂O, with 20 ml CSF of normal composition withdrawn, reducing the pressure to 9 cm H₂O. Acetazolamide was commenced and after an unknown period of 3 weeks later the headaches were settling, although occasionally present. Examination was normal; in particular there was no evidence of papilloedema.

Cerebral angiography 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 pressure, via lumbar puncture, was normal.

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 superficial transcapillary gradient, and hence a possibly increased 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 jugular 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 accounted for. 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.

This research was supported by the Madeline Foundation for Neurosurgical Research.

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False negative polymerase chain reaction on cerebrosplinal fluid samples in tuberculosis 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, 2 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 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 Loevenstein-Jensen slopes from CSF taken from both patients. False negative CSF PCR in tuberculous meningitis established by culture has rarely been reported. The two patients are described to emphasise the dangers of over reliance 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 tuberculous meningitis

J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.67.2.259c on 1 August 1999. Downloaded from http://jnnp.bmj.com/ on September 15, 2023 by guest. Protected by copyright.

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 is the most common, representing approximately 60% of all cases.

The myelin P gene, located on chromosome 17 (CMT1A), is the most common autosomal-dominant form of CMT.

The myelin P gene, located on chromosome 22 (CMT1B), is a rare form of CMT associated with mutations in the myelin protein zero (P0) gene.

The myelin P gene, located on chromosome X (CMTX), is a rare form of CMT associated with mutations in the myelin protein zero (P0) gene.
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.1,7 Although cranial nerves are generally spared in CMT, thickening of the acoustic or optic nerve has been reported in some cases.2 We report here on a Japanese woman who exhibited severe polyneuropathy, bilateral trigeminal thickening on MRI, and an abnormality of the auditory brainstem response. Gene analysis disclosed a novel missense mutation (His81Arg) of P0. The cranial nerve involvements 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 limbs, and distal hypesthesia. 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 fusiform 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 brainstem response showed normal findings in both proximal and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and distal hypesthesia. 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 fusiform deformities. Optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

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malignant lymphoma, was found to be positive. The Bcl-2 protein, which is an inhibitor of kappa light chain restriction. Staining for confirmed that these cells had a B lymphoid phenotype (CD20 positive) with lambda light chain restriction. Staining for Bcl-2 protein, which is an inhibitor of apoptosis and is expressed in 90% of follicular lymphoma, was found to be positive. The histological appearances and immunohistochemical profile confirmed a follicular lymphoma.

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 evidence. 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.2 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 material.2 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.2 Diffuse intracerebral lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellar pontine angle mimicking acoustic neuromlemoma or meningoima has been reported;4 Vigusin et al reported a patient with a calcified temporoparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular lymphoma resembling a meningioma. Rubinstein2 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.

We thank Professor Francesco Scaravilli, National Hospital for Neurology and Neurosurgery and Dr Mark Naper, The Meyerstein Institute of Oncology, Middlesex Hospital, for their help with this report.

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Determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentration can serve as an indicator of brain copper concentration.1 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 coeruleoplasmin) 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 coeruleoplasmin, CSF/serum albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum coeruleoplasmin and total serum copper concentration). The CSF copper concentration was treated as a dependent variable. 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). Coeruleoplasmin 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 coeruleoplasmin concentrations were 394.3 (SD11.7) µg/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 6.0) µg/l. The mean albumin ratio (AR) was 6.63 × 10−3. The mean ratio of calculated serum 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%. In the
Correlation of blood-CSF barrier (albumin ratio, AR) with total CSF copper concentration (on logarithmic axes; R=0.46, p=0.0001; 95% confidence bands for the true mean of the total CSF copper concentration are shown.

stepwise linear regression model (F to enter: 4.0, F to remove: 3.996), significant positive partial correlation of the CSF copper concentration were found to be AR (p=0.0001) and serum coeruloplasmin (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 CSF copper concentration (on logarithmic axes; R=0.46, p=0.0001).

The formula for the CSF copper concentration, derived from the multiple linear regression model, is: copper CSF (g/l)=3.32 × 10^(-6) × CSF/serum albumin ratio (×10^(-2)) + 0.012 serum coeruloplasmin (mg/l).

According to this analysis, CSF/serum albumin ratio and serum coeruloplasmin together determine 25.3% of the variation in CSF copper concentration (adjusted R^2=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 determination is carried out in a highly significant manner by disturbances in the blood-CSF barrier and by the serum coeruloplasmin concentration. It can be assumed that the in the case of normal blood-CSF barrier function and a normal serum coeruloplasmin concentration, 29.7% of the measured CSF copper originated from the brain, the CSF by passive diffusion bound to coeruloplasmin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20×10^(-2), this would mean that 60.6% of the measured CSF copper originated from the blood (bound to coeruloplasmin). 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 6.5×10^(-2)), that the CSF copper concentration is actually reduced by 27.4%, when the serum coeruloplasmin concentration falls from its normal value of 394 mg/l to 66 mg/l. 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 originates from the brain, the fraction entering the CSF by passive diffusion (bound to coeruloplasmin) 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 finding 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 coeruloplasmin-bound fraction originating from the plasma should be subtracted according to the formula we have given, or, better still, all measured copper concentrations in the CSF should be adjusted using the CSF/serum albumin ratio and serum coeruloplasmin concentration. A statistical relation with a low correlation (p<0.05) between CSF protein content and CSF copper was already shown in 1987 in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum coeruloplasmin (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 child 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 [3] and later subsequently renamed congenital generalised fibrous hamartoma [4] and finally fibromatosis congenita multiplex [5]. A rare case of 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. The main complaint was his skull circumference of 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 fornix of the CMYuro by tumour. The radiological differential diagnosis included a primary meningeal tumour, glioma, and leukenaemic deposit. The patient underwent a left frontal craniotomy and a firm, rounded mass was removed from brain. The histological examination showed a hamartomatous myofibroblast hyperplasia, 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 present, extensive involvement and relates either to mass effect with compression of vital organs and structures, or to pulmonary involvement, when subcutaneous 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.


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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 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 hamartomatous myofibroblast 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 present, extensive involvement and relates either to mass effect with compression of vital organs and structures, or to pulmonary involvement, when subcutaneous or submucosal cellular proliferation results in vascular or bronchial obliteration.

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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 afflicted, and generally involve the skin and soft tissues, especially of the head and neck. Solitary lesions are less commonly found in viscera or bones.1 Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported2 along with few cases of CNS involvement in the generalised form of infantile myofibromatosis.1,3-5 The prognostic outcome 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 variant of meningioma. Meningiomas are extremely rare in this age group, this lesion was not meningeal based and such lesions are usually reactive for epidermal 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.

Prior 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 reported6 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 Dr Lucy Roarte and Dr Louis Dehner in diagnosing this case.

V Mccurley, M J O'Sullivan, Lauren V Roarke and Dr Louis Dehner in diagnosing this case.

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.1 It is also called verotoxin producing E coli (VTEC), which produces a cytotoxic Shiga-like toxin. Gastrointestinal, haemorrhagic, and urological effects are well known in VTEC infection,2 and neurological problems are likely to be more frequent than is generally recognised.3 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 haemolytic colitis due to probable food poisoning. Then her urinary volume launched, 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 leucocytosis of 17 800/mm³. She was in a state of anuria and her blood analysis showed severe kidney dysfunction (increased serum creatinine 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 platelet count 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 also had a 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 in the legs. She described this feeling like frostbite 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 reflexes were decreased in the four limbs. Sensation for vibration was impaired in the lower legs, but preserved for pin prick, light touch, and joint sense. Routine laboratory data including haematochemical 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 potential (1.8 µV) and slow motor of conduction velocity (41.0 m/s) in the peroneal nerve. There were also markedly decreased amplitudes of the sensory nerve action potential (0.09 µV) and sural (0.989 µV) nerves. These findings and the clinical features confirmed the diagnosis of sensory dominant, axonal polyneuropathy. She was given 300 mg/day sulfamethoxazole (an anti-inflammatory agent) and 1500 µg/day meclobamatin (vitamin B12) without effect. Two weeks after administration of 300 mg/day oral meclozain, 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 of enterohaemorrhagic E coli O157:H7, and the detection of verotoxin in her stool. She had haemolytic-uraemic syndrome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, is killed by low doses of verotoxin through the process of apoptosis.4 Verotoxin shows similar cytokotoxicity on human glomerular microvascular endothelial cells via soluble factors such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis.4 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 convolution. She showed mild brain swelling, slowing 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 often of VTEC infection) is about 5%, 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.7 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 reticulohistiocytic areas, then
Crying spells as symptoms of a transient ischaemic attack

In the absence of depression, crying spells associated with neurological 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 paresthesias. Around 6:00am he awoke 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 bilateral 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 spells, but 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 nerve examination disclosed only flapping of the left nasolabial fold and decreased pin-prick sensation over his left face with an occasional mild facial twitching. Cranial nerves IX– XII were intact, and gag reflex and palate elevation were normal. It did not have dysarthria or a brisk jaw jerk. The rest of the neurological examination showed mild weakness in his left upper arm, and decreased pin-prick and temperature sensation over the left half of his body. Electrocardiogram were +2 and symmetric with downgoing toes.

The patient lacked prior depression, new depressive symptoms, 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 atrophy over the right frontotemporal cortex and an extensive fronto-temporal intermittent rhythmic delta activity but no epileptiform changes. Carotid Doppler studies showed atherosclerotic changes without haemodynamically relevant obstruction. He was discharged on amantadine therapy with aspirin.

These results suggest that crying spells can be a manifestation of a transient ischaemic attack. He presented with paroxysmal crying spells followed by a left sided hypoaesthesia and a mild left sided weakness, all of which resolved. His crying was non-emotional, inappropriate to the context, and did not correspond to his underlying mood. Moreover, the patient had multiple vascular risk factors supportive of a cerebrovascular aetiology for his episode.

The most common cause of pathological crying is pseudobulbar palsy, a complication of strokes and other diffuse or bihemispheric brain damage.1 Pseudobulbar palsy results from bilateral interruption of upper motor neuron innervation of bulbar motor nuclei and brainstem centres. In addition to crying, pseudobulbar palsy may include dysarthria, dysphagia, bifacial weakness, increased facial and mandibular reflexes, and weak tongue movements. There were no signs or symptoms of pseudobulbar palsy in this patient.

Crying or dacyrastic seizures also occur but are rare. These seizures are part of the range of complex partial seizures and usually emanate from the right temporal lobe system.2 Crying seizures may result from prior cerebrovascular infarctions.3 Although our patient had mild right sided lacunar involvement in his left hemisphere, he did not have any 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,4 crying also may result from right hemispheric strokes.5 Even more similar to our patient, sudden laughing spells, “le fou rire prodomique,” rarely precede strokes involving the left capsular-thalamic, lentico-caudate, or pontine regions.6 Our patient may have had a comparable phenomenon from the right hemisphere.7 Our case for the first time is of a crying spell syndrome that may have been temporary activation or stimulation of ischaemic motor pathways.

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Continuous drop type of orthostatic hypotension

Orthostatic hypotension has usually been evaluated for 2–10 minutes after standing.2–7 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 supine on a tilt 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 10 mm Hg from that at 5 minutes (mean 12.8 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. 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%).

Most patients with continuous drop type orthostatic hypotension reported reduced endurance for more than 10 minutes of exercise (easy fatigability). Two experienced vasodilatation of the volume vessels, is necessary for the difference in mechanisms between the early drop type and the continuous drop type. As we did not record heart rate and blood pressure continuously and did not evaluate ventricular function by echocardiography, the final conclusion and its interpretation require further study.

We think that more than a 20 minute tilt up study is needed to evaluate orthostatic hypotension and that reduced endurance of exercise and the syncope that occurs some time after standing should be considered symptoms of a continuous drop in blood pressure.

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CORRESPONDENCE

Respiratory aspects of neurological disease

An account of respiratory aspects of neurological disease, such as the highly informative one presented,1 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.2 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,2 who proposed, on the basis of their own study, that increased pulmonary vascular hydrostatic pressure might be a more significant aetiopathogenic mechanism than increased pulmonary capillary permeability.3 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.2

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

Although applauding the contribution of Pel-leckchia et al, we have noted more widespread recogni-
tion of the association between gluten sensi-
tivity and ataxia we disagree that ataxia associated with gluten sensitivity lacks “dis-
tinctive 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 1.2 Again, coexistent neu-
ropathy is common in these patients, found in two out of three of the patients of Pelleckchia et al and 21 of our 28.1 We agree that gastrointestinal symptoms are rare: rather than entitling their paper “lack of distinctive neurological features”, perhaps “lack of distinctive gastroenterological features” might have been more appropriate!

We were surprised at the high specificity and sensitivity of increased antigliadin anti-
boditles in their hands. Although we found both IgA and IgG antigliadin antibodies to be in-
visible 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 intriguing 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 Pelleckchia 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 in-
creased 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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1 Polkey MI, Lyall RA, Moxham J, et al. Respira-
tory aspects of neurological disease. J Neurol
2 Smith WS, Manthey MA. Evidence for a hydro-
static mechanism in human neurogenic pulmo-

Polkey replies: We thank Dr Jolobe for his interest in our article; we did not cover neurogenic pulmo-
nary oedema. We agree, however, that it can be a difficult clinical problem and therefore appreciate his contribution.

M I POLKEY

Procainamide for faecal incontinence in myotonic dystrophy

We read with interest the article by Aber-
crombie et al which describes the pathophysi-
ology and surgical management of faecal inconti-
ence in two siblings with severe myo-
tonic dystrophy.1

In the authors’ experience, long term results of both medical and surgical manage-
ment of the faecal incontinence were subject to variable outcomes.1 However, this report focused on the medical management of faecal incontinence in myotonic dystrophy.2

The authors’ pessimistic conclusions sug-
gest that “faecal incontinence in myotonic dystrophy is difficult to relieve 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 symp-
toms of sphincteric impairment developed soon after, including mild stress urinary incontinence and minor episodes of poor control of loose stool.

A complete diagnostic investigation, in-
cluding physical examination, defecography, and electrophysiological tests of pelvic floor musculature, was performed. At physical examination, digital ano-rectal evaluation showed low squeeze pressures. A reduced rectal diameter (4.9 cm), anal gaping, and barani loss at rest were found in defecog-
phy.

Motor evoked potentials elicited by cor-
tical 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.

A regular treatment with procainamide (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 dystro-
phy is still debated and controversial.3 His-

tological 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.  

The possible management of myotonia and some of its clinical manifestations, such as dystonia,1 by single-dose myotonic drugs (dipotassium-propanediol- and procainamide), 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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Flail arm syndrome or Vulpian-Bernhart'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 upper limbs that are less obvious than the signs of atrophy and weakness in the bulbar muscles or legs. Atrophy and loss of strength a predominantly showed signs of lower motor neuron involvement. Since then, in those countries and other countries under their influence,1,4 we have come to use the eponym of Vulpian-Bernhart'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 strongly suspected that the muscles predominantly affected in Vulpian-Bernhart's form were the deltoideus, the infraespinales, the supraespinales, the sternocleidomastoideus, and the teres minor. The predominant atrophy in these muscles permitted its distinction from that previously called Erb's dystrophy.10

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, abductor pollicis brevis, adductor pollicis, interossei, and lumbricales, which leads to the formation of the characteristic Aran-Duchenne hand.

Obviously, signs of cortiospinal involvement 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 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 form rebaptised as flail arm syndrome from other upper motor neuron syndromes such as of the spinal muscular atrophies, Kennedy's disease, multifocal motor neuropathy, and hypnic myotonia.  

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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 countries would have thought. There are some methodological issues which can be considered, as below, but the lesson of discarding “unisghly” data because it is too disturbing to one’s personal view and vested interest in the whiplash narrative has already been taught elsewhere.1 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 associations’ diagnostic orders 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 comparison 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 for subjects, and has a host of other reportedly fatal faults1, and giving some benefit of the doubt, the study is said to be an accurate representation of the state of affairs in Switzerland at that time, in that 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.2 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 grades (1 and 2) of whiplash associated disorders.3 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 report chronic pain as those in more severe collisions.6 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 culture 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, or 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, similarly 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.4 If there were study design 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 there, 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 Germany3 and Greece.4 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 knowledge to help prevent the chronic pain and the suffering we otherwise encounter.9

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, a 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, which I think one would have to go 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 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 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 other frontotemporal 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’Amold and 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.

8 Ferrari R, Russell AS. Whiplash: heading for a higher ground. A point of view.
10 Ferrari R, Russell AS. Whiplash: heading for a higher ground. A point of view.
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