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 infant underwent cerebral CT. This showed an unexpected arteriovenous malformation involving the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was 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 nodule measuring 1.5 x 0.1 mm above the vein of Galen and to the right of this (figure). This was thought to represent the thrombosed varix and possibly thrombosis of the vein of Galen and straight sinus. There was no evidence of new 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At lumbar puncture several days later opening pressure was 27 cm H2O, with 20 ml CSF of normal composition withdrawn, reducing the pressure to 9 cm H2O.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, in the absence of CSF protein or glucose, 3 weeks later the headaches were settling, although occasionally present. Examination was normal; in particular there was now 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 protein was normal, via lumbar puncture, was that 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.

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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 2 3 Successful culture 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 Lowenstein-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 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 Staphylococci.
appendicectomy and had received antitubercular medication for 3 years. She presented with a 3 month history of headache and coryza. CT brain showed a high opening pressure (15 cm CSF), was normal. Lumbar puncture results showed a high opening pressure (15 cm CSF), 90 white blood cells/µl, predominantly lymphocytes. The CSF/blood glucose ratio was normal. 3 weeks later from the first sample, growth of an organism was seen in culture. The organism showed sensitivity to isoniazid, 2 g pyrazinamide, and 10 mg pyridoxine daily. A CSF PCR was performed 4 weeks later showed similar results. A CSF PCR for M tuberculosis was again negative although a fully sensitive organism grew 12 weeks later from the first sample on Lowenstein-Jensen slopes.

The second patient was a 21 year old Kenyan woman living in the united Kingdom for 3 years. She had received antitubercular medication for 6 years during laparotomy for an ovarian cyst and had received antitubercular medication for 1 month only. On examination she had mild neck stiffness and a partial left third cranial nerve palsy. Brain CT was normal. Lumbar puncture results showed a high opening pressure (15 cm CSF), 90 white blood cells/µl, predominantly lymphocytes, a raised protein concentration (1.62 g/l), and a low CSF/blood glucose ratio. At the same referral laboratory CSF PCR for M tuberculosis was negative and 8 weeks later grew a fully sensitive organism. Despite the negative PCR antitubercular therapy was started empirically. After 2 months of treatment her symptoms had resolved although a partial third nerve palsy remains.

Adequate volumes of both patients' CSF (0.5 ml) were sent to our referral laboratory where a CSF PCR was performed using three primer sets and appropriate controls. The assay included primers for the target IS6110, an insertion sequence normally present in multiple copies in the M tuberculosis genome, which has been used successfully for the detection of M tuberculosis in CSF. Two primer sets were used as this is thought to increase the probability of detecting target DNA within a specimen.

Recent studies suggest that CSF PCR for M tuberculosis is more sensitive than culture in cases of clinically suspected tuberculous meningitis that responded to empirical treatment. Some authors have even suggested the usefulness of serial CSF PCR in assessing the efficacy of treatment. False negatives and positives are rarely reported in the literature and unless these results are critically reviewed patients could, tragically, have treatment prematurely stopped or be started on prolonged antitubercular chemotherapy. False negatives occurred in two studies, in which reported CSF PCR sensitivities were 32% and 85%.

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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 is a heterogeneous group of diseases which have been mapped to chromosome 17 (CMT1A), chromosome 1 (CMT1B), another unknown chromosome, (CMT1C) and the X chromosome (CMTX), CMT1B is a rare form of CMT1 associated with mutations in the myelin protein zero (P) gene. Mutations in the P gene have recently

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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 generally spared in CMT, thickening of the acoustic or optic nerve has been reported in some cases. We report here on a Japanese patient who exhibited severe polyneuropathy, bilateral trigeminal thickening on MRI, and an abnormality of the auditory brain stem 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 sensory atrophy of the hands. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand 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 normal prolongation of the I-III interpeak (2.81 ms on the right side, 2.88 ms on the left side). Brain MRI (figure) showed significant thickening of bilateral trigeminal nerves (7 mm) compared with that of controls (3.15 ± 1.62 mm (mean ± 2 SD), n=20). However, other cranial, spinal nerves and roots were not thick on physical examination or MRI study. Sural nerve biopsy was not performed. 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 (A→T at codon 81) resulting in a substitution of arginine for histidine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP 22) gene, was not present. Although multiple reports have addressed the possibility of a proportionately enlarged diameter of nerves in CMT type 1, and thickening of the cauda equina, nerve roots, and ganglia has also been found. Although blepharoptosis, ophthalmoplegia, facial weakness, deafness, dysphagia, and dysphonia in CMT have been previously reported, clinical involvement in the cranial nerves is rare and thickening of cranial nerves has not been reported except for the acoustic or optic nerves in some cases.

In the present study, our patient showed severe clinical manifestations of early onset and undetectable conduction velocities. Therefore, this patient was considered to have a severe 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 interval 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, Kimura et al. reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the I-V interpeak. 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 Arg81→His substitution in the P0 gene. Histidine 81 is conserved among many other species, including cows, rats, chickens, and sharks. This mutant allele was absent in the DNA from 100 controls. Therefore we identified this mutation as pathogenic. Arg81His was located in exon 3, which codes for the extracellular domain of P0. 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. A careful comparison of the clinical, electrophysiological, and histopathological data between patients with CMT should be conducted.
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 locoregional radiotherapy was discussed with the patient, however, she declined any further intervention. She has been closely reviewed in the follow up clinic and after 6 months there has been no clinical or radiological change.

Primary intracerebral lymphomas represent about 2% of intracranial neoplasms and 2% of all lymphomas. They occur most commonly in the 6th decade of life with a female to male ratio of roughly 2:1.1 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.2 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.3,4 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 peritumoural oedema and 92% enhance after administration of contrast medium.5 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.6 Diffuse primary lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellum (figure A). Right pterional craniotomy was performed and a tumour located under and adherent to the overlying dura was identified. It was entirely extracerebral, measuring 6×6×6 cm, with the greyish colour and hard consistency typical of a meningioma. The tumour and the adherent, thickened dura was macroscopically completely removed. Histologically the lesion consisted of lymphoid tissue with an ill defined follicular pattern. Follicular centres were composed of a mixture of centrocytes and centroblasts with mitotic activity (arrow). Haematoxylin and eosin, original magnification×30.

(A) Contrast enhanced CT of the head showing a 6×6×6 cm enhancing mass lesion in the region of the right lesser sphenoid wing. (B, C) Photomicrographs of the surgical specimen. (B) Section through the lesion showing triangular, ill defined lymphoid follicles. Haematoxylin and eosin, original magnification×30. (C) Follicle centre composed of a mixture of centrocytes and centroblasts with mitotic activity (arrow). Haematoxylin and eosin, original magnification×500.

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 calculated as a dependent variable in 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 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 394.3 (SD 87.7) 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 14.16 (SD 6.6) µg/l. The mean albumin ratio (AR) was 6.6×10⁻¹. 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%.
stepwise linear regression model (F to enter 4.0, F to remove: 3.996), significant positive predictors of the CSF copper concentration were found to be AR (p=0.0001) and serum ceruloplasmin (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 (µg/l)=5.32 µg/l/0.653 × CSF/serum albumin ratio (x10−3)+0.012 × serum ceruloplasmin (mg/l).

According to this analysis, CSF/serum albumin ratio and serum ceruloplasmin together determine 25.3% of the variation in CSF copper concentration (adjusted R²=0.253), implying that other (unknown) factors determine the remaining 74.7% of the variation. We have been able to demonstrate here that the CSF copper concentration is determined in a highly significant manner by disturbances in the blood-CSF barrier and by the serum ceruloplasmin concentration.

It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum ceruloplasmin concentration, 25.3% of the measured CSF copper results from the brain; the CSF by passive diffusion bound to ceruloplasmin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20×10³, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to ceruloplasmin). 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 an intact blood-CSF barrier (assuming a CSF/serum albumin ratio of 6.5×10³), that the CSF copper concentration is actually reduced by 27.4%, when the serum cerulo- plasmin concentration falls from its normal value of 394 µg/l to 66 µg/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 ceruloplasmin) 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 ceruloplasmin-bound fraction originating from the plasma should be subtracted according to the formula we have given, or better, all measured copper concentrations in the CSF should be adjusted using the CSF/serum albumin ratio and serum ceruloplasmin concentration.

A statistical relation with a low correlation (p=0.05) between CSF copper content and CSF copper was already shown in 1981 in various neurological diseases; our study shows a much higher significance and, in addition, the effect of serum ceruloplasmin (therefore of bound serum copper). Furthermore, we have been able to determine separately in the fraction of CSF copper which enters the CSF across the blood-CSF barrier.


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 Schram’ 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 tumour 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 myofibroblastic prolifer
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. Involution 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 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 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.

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

We present a patient with a solitary intracranial myofibroma with an excellent postoperative course. The child was well in 7 days after admission. The lesion was completely excised and the child made an uneventful recovery with no recurrence at 1 year after surgery.

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 cytotoxic Shiga-like toxin. Gastrointestinal, haemorrhagic, and uremic 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, her 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 leukocytosis 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/dl). 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 230–460 IU), suggestive of haemolytic anaemia. Her platelet count also decreased to 21 000/mm³. The culture of her stool showed the growth of E.coli O157:H7 and analysis of the bacterial toxins showed the presence of verotoxin, which confirmed the diagnosis of VTEC infection. Given plasma exchange, chronic haemodialysis, and antibiotics (4 g/day fosfomycin, 600 mg/day levofloxacin, and 2 g/day cefepirazon sulbactam). Her general status was unchanged for 1 week after admission and she was in a delirious state with visual hallucinations and tonic convolution, indicative of encephalopathy. Brain CT disclosed mild brain swelling and there were diffuse slow waves in the EEG. She was given 250 mg/day diphenylhydantoin. During the next two weeks her kidney function, haemolytic anaemia, and encephalopathy gradually improved.

After recovery of consciousness she began to complain of numbness of the limbs, manifested by the legs. She described walking like from 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 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 sensation. Routine laboratory data including haematological studies, serum chemistry, urinalysis, and CSF analysis were normal. Serum concentrations of vitamin B1, B6, and B12 were normal. Nerve conduction studies were carried out on her right limbs, and showed normal findings in the distal latencies, motor conduction velocities, and F wave latencies of the median, ulnar, and tibial nerves, and no evidence of conduction block. However, there were demyelinating and muscle action potentials (1.18 mV) and mild slowing of motor conduction velocity (41.0 m/s) in the peroneal nerve. There were also markedly decreased amplitudes of the sensory nerve action potentials (150 µV) and sural (0.98Vµ) nerves. These findings and the clinical features confirmed the diagnosis of sensory dominant, axonal polyneuropathy. She was given 300 mg/day sulbactam (an anti-inflammatory agent) and 1500 µg/day mecholobamin (vitamin B12) without effect. Two weeks after administration of 300 mg/day oral mexiletin, her numbness and pain gradually disappeared.

The patient was diagnosed as having VTEC infection, because of a typical history of an acute haemorrhagic colitis, the cultured growth of enterohaemorrhagic E.coli O157:H7, and the detection of verotoxin in her stool. She had haemolytic-uraemic syndrome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, can be killed by low doses of verotoxin through the process of apoptosis. 1, 2 Verotoxin shows similar cytotoxicity on human glomerular microvascular endothelial cells via inflammation mediators such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis.3 Our patient was treated with antibiotics, plasma exchange, and chronic 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 on CT and diffuse slow waves in the frontal area on EEG, evidence of encephalopathy. Previous reports have shown that the incidence of encephalopathy in haemolytic-uraemic syndrome (most of VTEC infections) is low, up to 40%, 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.3 There is experimental evidence that verotoxin has direct virulence to both endothelial cells and neurons in the nervous system, and its initial lesion is in the hypothalamic areas, then...
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 polyneuropathy. 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 melixin, 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 or loss of integrity to the neuronal cells by verotoxin could cause axonal neuropathy in VTEC infection.

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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 paraesthesia. Around 6.00 am he woke with a dull headache and 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 peripheral nerves. He did not have 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 these events, 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 revealed flattening of the left nasobulbar fold and decreased pinprick 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. The patient 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 pinprick and temperature sensation over the left half of his body. Numbness 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 frontal horns, and an ECoG showed frontal intermit-}

cent rhythmic delta activity but no epileptiform changes. Carotid Doppler studies showed atherosclerotic changes without haemodynamically relevant obstruction. He was discharged on antipateptide therapy with aspirin.

These results suggest that crying spells can be a manifestation of a transient ischaemic attack. He presented with paroxysmal crying spells following by a left sided hypaesthesia and 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 dyscaric seizures also occur but are rare. These seizures are part of the range of complex partial seizures and usually emanate from the right temporalis lobe.2 Crying seizures may result from prior cerebrovascular infarctions.3 Although our patient had mild left sided weakness in his left face, he did not exhibit 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, crying also may result from right hemispheric strokes.4 Even more similar to our patient, sudden laughing spells, “le fou rire prodromique,” rarely precede strokes involving the left capsular-thalamic, lenticulo-caudate, or pontine regions.5 Our patient may have had a comparable phenomenon from the right hemisphere. The pathophysiology for this phenomenon may have been a 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.1,2 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 down on a tilting table, and an intravenous cannula was introduced into the cubital vein more than 30 minutes after recovering consciousness, diastolic blood pressure fell immediately after tilting, while in 13 patients (early drop type); whereas, in 13 patients blood pressure fell immediately after tilting but kept decreasing by more than 8 mm Hg from that at 5 minutes (mean 12.9 mm Hg;
We used a Swan-Ganz catheter to investigate orthostatic hypotension in three patients with MSA. 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 type (figure). The other five patients could not remain standing for more than 5 minutes to reach the minimum (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.

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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, 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 aetiopathogenic 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.

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

Although applauding the contribution of Pellecchia et al, we note 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 our own 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 is common in these patients, found in two out of three of the patients of Pellecchia et al and 21 of our 28. 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 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 antidesmoyosin 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 antidesmoyosin antibodies in gluten ataxia.

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

We thank Hadjivassiliou et al for their interesting comments on our paper. They suggest that patients with gluten ataxia can be distinguished by the late onset of gait ataxia and the relatively mild upper limb signs. Our results support the finding of a late onset in these patients, but this feature cannot be considered a distinctive one. In fact, in our population 11 out of 24 patients with idiopathic cerebellar ataxia had a late onset, but only three of them were affected by celiac disease.

Furthermore, we do not think that celiac patients may be distinguished by mild upper limb signs and coexistent neuropathy; in our study 20 out of 24 patients with idiopathic cerebellar ataxia, including the three patients with celiac disease, had ataxic gait as the presenting and prominent clinical feature. Similarly, nerve conduction studies, performed in 17 out of 24 patients, showed a peripheral neuropathy in nine, including two out of the three patients with celiac disease.

We understand that some discrepancies arise comparing our study with that of Hadjivassiliou et al. Firstly, only six out of their 28 patients had evidence of cerebellar atrophy on MRI, whereas all of our patients had cerebellar atrophy. Secondly, many of their patients had a peripheral neuropathy in the absence of cerebellar atrophy. This finding could explain the increased upper limb signs. Although two of our three celiac patients had a clinically silent peripheral neuropathy, we think that their ataxia was explained by cerebellar atrophy. Thirdly, we found a high prevalence (12.5%) of celiac disease on duodenal biopsy among patients with idiopathic cerebellar ataxia, whereas none of the six patients with cerebellar atrophy described by Hadjivassiliou et al showed histological features of celiac disease.

It would be interesting to know the prevalence of gluten ataxia among all ataxic patients screened for antigliadin by Hadjivassiliou et al. Our series is too small to estimate the sensitivity of both antigliadin and antidesmoyosin antibodies in gluten ataxia; unfortunately Hadjivassiliou et al did not report any data on antidesmoyosin antibody screening in their patients. On the other hand, we were surprised at the high prevalence of antigliadin antibody positivity (12%) in the normal population studied by Hadjivassiliou et al in a previous report. This is in contrast with the 2% of antigliadin antibody positivity found in a large population by Catassi et al. Further studies are required to better characterise the syndrome of cerebellar ataxia associated with celiac disease or gluten sensitivity.

Procainamide for faecal incontinence in myotonic dystrophy

We read with interest the article by Abercrombie et al which describes the pathophysiology and surgical management of faecal incontinence in two siblings with severe myotonic dystrophy. In the authors' experience, long term results of both medical and surgical management of the faecal incontinence, using procainamide, has been disappointing. We would like to report on our experience in a patient with severe myotonic dystrophy presenting with severe faecal incontinence.

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 muscle function, was performed. At physical examination, defecography revealed a reduced rectal diameter (4.5 cm), anal gaping, and barium loss at rest. Sphincter motor units showed evidence of denervation. Motor evoked potentials elicited by cortical and lumbar magnetic stimulation and recorded from the external anal sphincter showed a normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed, as in the first patient of Abercrombie et al, a decreased number of motor units and multiple myotonic discharges. Few motor unit potentials presented polyphasic waveforms and decreased duration and amplitude.

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

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

The possible management of myotonia and some of its clinical manifestations, such as dystonia,1 by use of myotonic drugs (disopyramide 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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1 Abercrombie JF, Rogers J, Swash M. Faecal incontinence with procainamide should always be attempted before any surgical option in patients with myotonic dystrophy. The predominant involvement in these muscles: opponens pollicis, flexor brevis, abductor pollicis brevis, adductor pollicis, interossi, and lumbricales, which leads to the characteristic distribution of weakness and symmetric proximal 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 appears that the muscles predominantly affected in Vulpian-Bernhardt's form were the deltoideus, the infraspinatus, the supraspinatus, the sternocleidomastoideus, and the teres minor. The predominant involvement in these muscles permitted its distinction from its previous entity called Erb's dystrophy. As a consequence of the atrophy of these muscles, the upper limbs adopt a characteristic posture, with the shoulders slumped, and the arms, forearms, and hands in pronation. As the illness progresses, the hand muscles are affected, with atrophy of the following muscles: opponens pollicis, flexor brevis, abductor pollicis brevis, adductor pollicis, interossi, and lumbricales, which leads to the formation of the characteristic Aran-Duchenne hand. Obviously, signs of corticospinal involvement with hypertreflexia 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 rebaptised as flail arm syndrome from other motor neuron syndromes such as of the spinal muscular atrophies, Kennedy's disease, multifocal motor neuropathy, and monomelic amyotrophy.

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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 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 display 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 1886, 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 Système 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 appears that the muscles predominantly affected in Vulpian-Bernhardt's form were the deltoideus, the infraspinatus, the supraspinatus, the sternocleidomastoideus, and the teres minor. The predominant involvement in these muscles permitted its distinction from its previous entity called Erb's dystrophy. As a consequence of the atrophy of these muscles, the upper limbs adopt a characteristic posture, with the shoulders slumped, and the arms, forearms, and hands in pronation. As the illness progresses, the hand muscles are affected, with atrophy of the following muscles: opponens pollicis, flexor brevis, abductor pollicis brevis, adductor pollicis, interossi, and lumbricales, which leads to the formation of the characteristic Aran-Duchenne hand.

Obviously, signs of corticospinal involvement with hypertreflexia 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 rebaptised as flail arm syndrome from other motor neuron syndromes such as of the spinal muscular atrophies, Kennedy's disease, multifocal motor neuropathy, and monomelic amyotrophy.
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.8 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 bias 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 cultures 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.14 If there were strong 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 Germany and Greece.1 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.15

RAJESH MUNGALI


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 in the psychobiology 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. 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 MUNGALI

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 Ruck 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 denervation, 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 in chronic low 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 would only lead one to a bigger text for. In summary I think that this would make an excellent addition to the bookshelf of those involved in the treatment and management of pain.

RAJESH MUNGALI


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

Surgical Disorders of the Peripheral Nerves. Edited by R BIRCH, G BONNEY, and C W WYNN PARRY. (Pp 539, £95.00). Published by Harcourt Brace and Co Ltd. London 1998. ISBN 0 443 04443 0.

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 textbook 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 précised of the changes occurring in medical negligence claims. This text represents good value for money.

IAN WHITWORTH
Pain after whiplash

R FERRARI

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