Pseudotumour after arteriovenous malformation embolisation

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

At 2 years of age, as part of investigating a failure of normal growth, a female child underwent cerebral CT. This showed an unexpected arteriovenous malformation involving the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was not undertaken until the age of 5 years when vertigo and intermittent numbness of the left arm and leg had been present for about 12 months.

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

The patient complained of right sided headache for 24 hours after the procedure, resolving with minor analgesia. Brain CT the following day showed a large thrombus in the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was not undertaken until the age of 5 years when vertigo and intermittent numbness of the left arm and leg had been present for about 12 months.

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

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

Ten days after the embolisation the patient presented with a generalised, pounding headache, present since discharge. Examination showed mild left papilloedema, with no focal neurological signs. Brain CT showed a dense nodule measuring 1.6 x 1.0 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 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 3 weeks 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 production, via lumbar puncture, was 0.3 ml/hour.

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 subtotal transcillary gradient, and hence a possibly decreased CSF production. If this were the case, with decreased production serving to retard the normal development of absorptive capility, 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, and it is possible that increases in the rate of CSF production is the pressure 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’s ability 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 pseudotumor developing in the setting of minimal venous thrombosis, particularly in part of the venous system not thought to play a major role in the absorption of CSF, must force us to reconsider our opinions as to the relation between venous obstruction and CSF dynamics.

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

The polymerase chain reaction (PCR) has been reported to be of diagnostic value when performed on CSF samples in tuberculous meningitis.1–4 Rapid amplification of Mycobacterium tuberculosis specific DNA enables results to be available within 48 hours and can influence treatment decisions.

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

The first patient was a 28 year old Asian man, last in India 8 years previously. He was sent from a clinic to hospital for incision and drainage of two deep seated Staphylococcus
auraless absences. While an inpatient he complained of headaches and nausea and developed a low grade pyrexia and meningitis. Brain CT was normal. Lumbar puncture disclosed a high opening pressure (19 cm CSF), 133 white blood cells/µl, predominately lymphocytes, a raised protein concentration (1.62 g/l), and a low CSF/blood glucose ratio (1.7/0.1). A sample of 0.5 ml CSF was sent to a British referral laboratory and PCR for M tuberculosis was negative. Twenty four hours later, because of increasing confusion and agitation, treatment with intravenous acyclovir, antituberculcer chemotherapy (600 mg rifampicin, 300 mg isoniazid, 2 g pyrazamide, and 10 mg pyridoxine daily), and dexamethasone was commenced. Clinically he showed signs of improvement and was discharged home 2 weeks later on the above treatment. A repeat lumbar puncture 4 weeks later showed similar results. A PCR CSF for M tuberculosis was again negative although a fully sensitive M tuberculosis 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 was referred with a 3 month history of photophobia and occipital headaches. She had no other systemic symptoms. She had had peritoneal tuberculosis diagnosed at the age of 6 years during laparotomy for an appendicectomy and had received antituberculosis 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 (5 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 but culture after 2 weeks grew a fully sensitive organism. Despite the negative PCR antituberculosis 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 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. Multiple primer sets were used as this is thought to increase the probability of detecting target DNA within a specimen.

Recent studies suggest that 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 antituberculous chemotherapy. False negatives occurred in two studies, in which reported PCR CSF sensitivities were 32% and 85%.

In one study 6.1% of CSF specimens received from patients with no evidence of tuberculous meningitis were falsely PCR positive. These results also show that sensitivity and specificity can vary when different assays and laboratories are used. Claims that PCR can detect 1–10 M tuberculosis organisms “in vitro” seems not to be the case in clinical samples such as CSF.

In the two patients presented above adequate volumes and repeated samples of CSF were assayed using suitable primers and appropriate controls at a British referral laboratory. Results for these two patients show the dangers of overreliance on PCR CSF when tuberculous meningitis is clinically suspected.

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

There have been few studies in the literature concerned solely with the use of the polymerase chain reaction (PCR) to identify Mycobacterium tuberculosis DNA directly from CSF. These studies suggest that in some cases, PCR may be more sensitive than culture; however, in the largest study, performed by Nguyen et al, specimens from seven patients who were culture positive for M tuberculosis were not positive by PCR. The study did report on 22 culture negative, PCR positive patients, suggesting that PCR can be more sensitive than culture. Studies comparing PCR with culture of M tuberculosis using other clinical specimens, particularly respiratory specimens, have reported that PCR may be less sensitive than culture for the detection of M tuberculosis and that the low sensitivity correlated with low colony counts on culture. Dalovisio et al also reported that multiple specimens may be required to improve the sensitivity of the test in some patients. In the two cases described above, colonies were seen after incubation for 12 and 8 weeks on LJ slopes, suggesting a low inoculum.

The PCR has been reported to detect the equivalent of 1–10 mycobacteria in in vitro testing. However, lower sensitivity is found with clinical specimens. The sensitivity of PCR may be the result of inhibitors of PCR present in the reaction, poor lysis of mycobacteria, and the uneven distribution of mycobacteria in clinical specimens.

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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 lack of 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 pes cavus deformities. Optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

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

Although no detailed familial information was available, her mother (49 years old) was available, her mother (49 years old) showed normal findings on neurological examination. She also had atrophy of the hands and pes cavus deformities. Optic atrophy, 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-II interpeak interval in auditory brain stem response were found. The I-II interpeak interval represents the conduction time from the eighth nerve to the pontomedullary junction. Prolongation of the auditory brain stem response suggested peripheral conduction delay of the auditory nerve.

Trigeminal neuropathy 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 \textsuperscript{1} reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of the motor fibers. 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 Arg\textsuperscript{81} to His substitution in the P0 gene. Histidine 81 is conserved among many other species, including cows, rats, chickens, and sharks. This mutant allele was absent in the DNA from 100 controls. Therefore we identified this mutation as pathogenic. Arg\textsuperscript{81}His 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.

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References


Intracranial extracerebral follicular lymphoma mimicking a sphenoid wing meningioma

Primary lymphoma in the brain is uncommon, accounting for only 2% of primary intracranial neoplasms. Although its incidence seems to be dramatically increasing,\textsuperscript{7,8} leptomeningeal lymphomas are even rarer but have been described.\textsuperscript{7,8} However, no leptomeningeal lymphoma of the follicular type has previously been reported. We present a case of a primary meningeal follicular lymphoma which mimicked a sphenoid wing meningioma, both radiologically and intraoperatively.

A 37 year old Ghanaian woman was referred with a 3 year history of worsening bitemporal headache, followed by a 6 month history of daily right frontal headache lasting for 2–3 hours associated with mild photophobia. There were no reports of seizures, nausea, or other visual disturbances. Her medical history was 3 years of treated hypertension, sickle cell carrier trait, and a cataract extraction. The patient was obese but physical examination was otherwise normal. Neurological examination showed no papilloedema and there were no cranial nerve or long tract signs.

Brain CT showed an enhancing mass consistent with a right sided sphenoid wing

Axial T1 weighted (TR 600/TE 15) brain MRI at 1.5 Tesla of our patient with CMT. Note the thickness of the bilateral trigeminal nerves.
leukemic 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 rather than a meningioma (figure A). Right pterional craniotomy was performed and a tumour measuring 6 cm × 500. 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. meningioma. The tumour and the adherent, colour and hard consistency typical of a dural, measuring 6 cm × 6 cm, with the greysish dura was identified. It was entirely extracerebral, measured 6 cm × 6 cm, with the greysish 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 architecture (figure B). The follicles varied in size and shape and infiltrated the overlying dura. Follicular centres were composed of a mixture of centrocytes and centroblasts with frequent mitotic figures and apoptotic bodies (figure C). Immunohistochemical staining confirmed that these cells had a B lymphocytic phenotype (CD20 positive) with kappa 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 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 1:3. The association between primary intracerebral lymphoma and immunodeficiency has long been established, and it is not surprising, therefore, that the incidence has increased 10-fold over the past 3 decades with the onset of transplant surgery and, particularly, the AIDS epidemic. In postmortem studies, these neoplasms are found, on average, in 5.5% of AIDS cases, and malignant cerebral lymphoma is the most common diagnosis of a focal intracranial lesion in patients with AIDS. Malignant primary lymphoma can occur throughout the CNS and they often have a periventricular distribution. Multifocality seems to be more common in patients with AIDS. The CT scan usually shows hyperdense masses with peritumorous oedema and 92% enhance after administration of contrast medium. Leptomeningeal lymphoma is usually encountered as a late complication of systemic non-Hodgkin’s lymphoma, although primary leptomeningeal lymphoma is occasionally seen. The prognosis for these tumours is poor. Diffuse intracerebral lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellopontine angle mimicking acoustic neurilemoma or meningioma has been reported; Vigushin et al reported a case of a calcified temporoparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular rather than diffuse intracranial lymphoma which has been described. Rubinstein described a case of follicular lymphoma metastasis found in the dura of a 61 year old man at necropsy. We found no report of a primary follicular extracerebral lymphoma. Similar radiological and intraoperative appearances of the tumour in our case to sphenoid wing meningioma suggest that this entity should be considered as a rare differential diagnosis.

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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. 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 albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum ceruloplasmin and total serum copper concentration). The CSF copper concentration was treated as a dependent variable of 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). Coeruloplasmin was determined nephelometrically (Beckman Instruments, Brea, CA, USA). The age of the patients was 50.0 (SD15.5) years; 50 were women and 63 were men. Mean serum ceruloplasmin concentrations were 394 (SD 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 6.0) µg/l. The mean albumin ratio (AR) was 6.63x10⁻¹. The range of calculated ratio of 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

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Correlation of blood-CSF barrier (albumin ratio, AR) with total CSF copper concentration (on logarithmic axes). R²=0.46, p=0.0001; 95% confidence bands for the true mean of the total CSF copper concentration are shown.

A 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 serum ceruloplasmin concentration. A statistical relation with a low correlation (p=0.05) between CSF protein 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 experimentally the fraction of CSF copper which enters the CSF across the blood-CSF barrier.

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Solitary intracranial myofibroma in a child

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

A case of congenital fibrosarcoma was first diagnosed by William and Schram1 and was subsequently renamed congenital generalised fibromatosis by Stout in 19542 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 cells of origin and the occurrence of this condition in infants, as well as congenitally, it was renamed infantile myofibromatosis by Chung and Enzinger in 1981.3 This disorder is considered to represent a hamartomatous myofibroblastic prolifer-
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofibrils.

Solitary lesions of infantile myofibromato-
sis 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.11 Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported12 along with few cases of CNS involvement in the generalised form of infantile myofibromatosis.13

The provisional 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 microscopic 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 anti-
gen like this tumour. This lesion, unlike some meningioma, showed no immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

Periorbital and intracranial involvement by myofibromatosis includes patients with widespread systemic involvement and multiple leptomeningeal nodules1 in one patient and extramedullary masses in another, both of which were fatal at the age of 10 days, a non-fatal, extramedial in 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 reported13 in which the patient died within 24 hours of surgery, secondary to cardiorespiratory arrest.

We present a patient with a solitary intracra-
nial myofibroma with an excellent postop-
erative 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 Roeke 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.2 It is also called verotoxin producing E. coli (VTEC), which produces a cytotoxic Shiga-like toxin. Gastrointestinal, haemorrhagic, and ureamic effects are well known in VTEC infection,3 and neurological problems are likely to be more frequent than is generally recognised.4 Here we describe axonal polyneuropathy and encephalopathy in a young female patient associated with haemolytic-uraemic syndrome caused by VTEC infection.

A 26 year old woman began to have abdominal pain and haemorrhagic diarrhoea. She was admitted to an emergency hospital and diagnosed as having haemorrhagic colitis due to probable food poisoning. Then her urinary volume rapidly increased and serum creatinine increased, and she was transferred to our hospital. On the 9th day she had a high fever of 39.7°C with increased C reactive protein of 7.6 µg/ml and a leukocytosis of 17 800/mm³. She was in a state of anuria and her blood analysis showed severe kidney dysfunction (increased serum creati-
ine 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 dehy-
drogenase concentration of 4095 IU (normal range 230–460 IU), suggestive of haemolytic anaemia. Her platelet count was decreased to 21 000/mm³. The culture of the 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.4 She showed diffuse plasma exchange, continuous haemodialysis, and antibiotics (4 g/day fosfomycin, 600 mg/day levofloxacin, and 2 g/day cefoperazon/ sulfactam). Her general status was un-
changed for 3 weeks after admission and she was in a delirious state with visual hallucina-
tions and tonic convulsion, indicative of encephalopathy. Brain CT disclosed mild brain swelling and there were diffuse slow waves in the frontal area on 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, manifest-
est in the legs. She also started feeling like frost bite when she was lying on the bed, and this gradually exacerbated to be a burning pain. On examination she was alert and cooperative. Her cranial nerves were normal. Muscle strength was normal and coordination was intact. Deep tendon re-

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spreading into the hippocampus and the brainstem. The convulsions in our patient were successfully treated with 250 mg/day diphenhydantoin, 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 methylxoth, an agent with a membrane stabilising effect. Up to now, to our knowledge, peripheral polyneuropathy 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 disturbances or toxicity to the peripheral 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 6am he awoke with a diffuse, 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 paraesthesia in his trunk or lower extremities. He had photophobia, nausea or vomiting, blurred vision, visual obscurations, difficulty swallowing, dysarthria, or focal weakness. Over the next 2 to 3 hours, he had five more crying spells, each lasting 5 to 10 minutes, occurring out of context, without precipitating factors or sadness, with an acute onset and offset, and without alteration of consciousness. The patient’s left face and arm numbness persisted during and between these 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 bruises. His mental status was normal. Cranial nerve examination showed flattening of the left nasolabial 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. He 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. His reflexes 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, paraesthesiaes, 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 capping over the right frontal and parietal lobes, and an EDX study showed right frontal intermit-}

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 at this patient in his episode.

Crying or dacyratic 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.1 Crying seizures may result from prior cerebral infarctions.2 Although our patient had mild left hemispheric signs, he fulfilled none of the 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,3 crying also may result from right hemisphere strokes.4 Even more similar to our patient, sudden laughing spells, “levire prodromiqum,” rarely precede strokes involving the left capsular-thalamic, lenticulocaudate, or pontine regions.5 We our patient may have had a comparable phenomenon from the right hemisphere, which is a unilaterial cell anism for this phenomenon may have been temporary activation or stimulation of ischae-
We used a Swan-Ganz catheter to investigate 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 Mathay,3 who proposed, on the basis of their own study, that increased pulmonary vascular hydrostatic pressure might be a more significant aetio-pathogenetic mechanism than increased pulmonary capillary permeability.4 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.5

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

Although applauding the contribution of Pellecchia et al. to the more widespread recognition of the association between gluten sensitivity and ataxia, we disagree that ataxia associated with gluten sensitivity lacks “distinctive neurological features”. Both their data and ours indicate that this group of patients can be distinguished by the late (non-childhood) onset of gait ataxia with relatively mild upper limb signs, analogous to Harding’s group.1 Again, coexistent neuropathy is common in these patients, found in two out of three of the patients of Pellecchia et al and 21 of our 28.2 We agree that gastrointestinal symptoms are rare: rather than entitling their paper “lack of distinctive 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 idiopathic ataxia 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 antidiemysium 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 antidiemysium 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 antigens3. Although the clinical features of gluten ataxia are not entirely specific, they are distinctive.

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Pellecchia et al reply:

We thank Dr Jobole for his interest in our article; we did not cover neurogenic pulmonary oedema. We agree, however, that it can be a difficult clinical problem and therefore appreciate his contribution.


Polkey replies:

We agree that the clinical features of 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 our 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 relatively mild 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 cerebellar disease on duodenal biopsies among patients with idiopathic cerebellar ataxia, whereas none of the six patients with cerebellar atrophy described by Hadjivassiliou et al showed histological features of cerebellar 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 antidiemysium antibodies in gluten ataxia; unfortunately Hadjivassiliou et al did not report any data on antidiemysium 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.4 This is by contrast with the 2% of antigliadin antibody positivity found in a large population by Catassi et al.5 Further studies are required to better characterise the syndrome of cerebellar ataxia associated with celiac disease or gluten sensitivity.


Procarainamide for faecal incontinence in myotonic dystrophy

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

In the authors’ experience, long term results of both medical and surgical management of the faecal incontinence were unsatisfactory. It should be noted, however, that the medical treatment used is not specified in the text.

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

A complete diagnostic investigation, including physical examination, defecography, and electrophysiological tests of pelvic floor musculature, was performed. At physical examination, digital anorectal evaluation showed low squeeze pressures. A reduced rectal diameter (4.5 cm), anal gaping, and barium loss at rest were found in defecography. Motor evoked potentials elicited by cortical and lumbar magnetic stimulation and recorded from the external anal sphincter showed a normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed a normal latency and decreased amplitude. Somatosensory evoked potentials after anal stimulation and sacral reflex latency were normal. EMG recording of the external anal sphincter showed a normal latency and decreased amplitude.

A regular treatment with procarainamide (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 rest 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 myoton, myopathy, muscular atrophy, and neural abnormalities. The possible management of myotonia and some of its clinical manifestations, such as dysmorphism, by means of myotonic drugs (diprery- maide 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 features 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 data for the physician, such as a clear predominance among men, and a longer median survival. They conclude by suggesting that this syndrome could be a new variant of amyotrophic lateral sclerosis.

Finally, the authors carry out a historical review and refer to the fact that this distinctive amyotrophic lateral sclerosis variant was probably first described by Gowers in 1888, furnished with exquisite graphic illustrations. To this effect, we draw attention to prior descriptions of the same syndrome, reported by Vulpian in 1886, known in Franco-German literature as Vulpian-Bernhard'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 atrophy, and signs of denervation and upper motor neuron involvement. Since then, in those countries and other countries under their influence, we have come to use the eponym of Vulpian-Bernhard'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 noted that the muscles predominantly affected in Vulpian-Bernhard's form were the deltoideus, the infraespinales, the supraespinales, the sternocleidomastoidea and the teres minor. The predominant involvement in the two muscular regions in these muscular clines permitted its distinction from that previously called Erb's dystrophy.

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 corticospinal 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-Bernhard's variant (classically termed as flail arm syndrome from other upper motor neuron syndromes such as of the spinal muscular atrophies, Kennedy's disease, multifocal motor neuropathy, and monomelic amyotrophy.

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

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

The first consideration is that there were 98 accident victims who reported acute symptoms, and thus were at risk for the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome? The Swiss study may be useful for comparison, it was completed in 1997 and, however, its greatest strength. The study has, however, its limitations.

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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, it was completed in 1997, and, however, its greatest strength. The study has, however, its limitations.
A second consideration is that perhaps these Lithuanians are in very minor collisions. True, some of their vehicles were completely wrecked, but perhaps the vehicles were not very good quality and so were easily damaged. Perhaps that is why this cohort had such a good outcome and only minor injuries. This is an unhelpful consideration however, as studies in Canada have shown that those with absolutely no vehicle damage, in very low velocity collisions, are just as likely to report chronic pain as those in more severe collisions. Lithuanians seem to behave appropriately then for minor collisions (if that is what they indeed had), but Canadians seem unable to behave appropriately. Again, another culture difference 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 make much sense, as seat belt 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 so obvious in this study that the general Lithuanian population reports the same prevalence, frequency, and character of neck pain and headache as does the general population in western countries. If there were study design barriers to identifying symptoms, the control population would have grossly underreported their symptoms. Indeed, chronic pain can and is reported by many cultures, 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 reality 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 the epidemic of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany and France. The time has not yet come for a reconciliation of these epidemiological observations with our own experience of late whiplash syndrome in western countries. The truth has been laid bare and it is our responsibility to utilise this truth to help prevent the chronic pain and the suffering we otherwise encounter.

R FERRARI


BOOK REVIEWS


This book purports itself to be a comprehensive reference. Certainly the title would suggest so. However, it is clear that this is not a comprehensive text, but a book that is an update on particular timely topics in the field of pain medicine. There are sections on pain mechanisms, and the pharmacology of acute and chronic pain, and other chapters on postoperative pain, obstetric pain, and acute paediatriic 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. 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 for 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 how to do it approach, 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 neuropathologist with a longstanding interest in the dementias, I found it extremely valuable. The editor has done a very good job in posing a coherence, format, and style, which is often lacking from multicontributor textbooks.

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

The editor has managed to persuade many of the world’s experts to contribute. For instance, the chapter on prion diseases is by D’Almond and the recent Nobel laureate Prusiner, and the frontotemporal dementias are reviewed by Brun and Gustafson. Genetics of Alzheimer’s disease are dealt with by St George-Hyslop and the neuropathology of Alzheimer’s disease by Price and coworkers.
The standard of illustrations is excellent and the style generally very readable. I shall
certainly find it extremely useful as a work of reference and for teaching purposes. The
editor is to be complimented on producing such a delightful work.

JOHN HODGES

Instrumented Spinal Surgery. Principles and Technique. Edited by
JÜRGEN HARMS and GIUSEPPE TABASSO. (Pp 198, DM248.00). Published by Georg

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 practi-
cal 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 jar-
gon. 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 reconstit-
tuted in compression. The remaining chapters cover fracture management, late kypho-
sis, metastatic tumours, spondylolisthesis, degenerative spinal disease, and infection.
Each chapter sets out the principles of management which are illustrated schemati-
cally. 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 fascina-
tion 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 diffi-
cult to disagree with much of the logic presented by the authors in planning their interventions but there is a danger that inex-
perienced 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 B 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 themselves and many trainees a clarity in their
planning of management of complex prob-
lems that humbles one’s own thoughts. That
clarity has continued in this text book of over
500 pages. The field of peripheral nerve sur-
ery is covered comprehensively, commencing
with descriptions of anatomy, physiology, and pathological reaction to injury. This is
followed in subsequent chapters with de-
scriptions of approaches to virtually all the
main peripheral nerves, and the operative
management of brachial plexus injury and outcomes is covered in three detailed chap-
ters. 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 tech-
niques, and rehabilitation.

The text is well written, easy to read, and
supplemented by some excellent line draw-
ings similar to those used in Lundborg’s text.
There are detailed plates showing histology and various imaging techniques. Each chap-
ter is comprehensive, containing important historical aspects as well as up to date
techniques, and there is an extensive refer-
ence section. I would recommend that train-
ees 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 inju-
ries 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