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 familial pattern of abnormal growth, a female child underwent cerebral CT. This showed an unexpected arteriovenous malformation involving the vein of Galen. Although there was no evidence of cardiac failure or hydrocephalus associated with this, assessment by angiography was advised. This, initially declined by the parents, was 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 communicat- ing 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 ophthalmo logical review was undertaken before discharge showing normal fundi and fields.

Ten days after the embolisation the patient was readmitted presenting with headache, present since discharge. Examination showed mild left papilloedema, with no focal ache, present since discharge. Examination of the left papilloedema was normal; in particular there was no evidence of hydrocephalus. Brain CT at level of vein of Galen demonstrating thrombus.

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 subnormal transcapillary gradient, and hence a possibly decreased CSF production. If this were the case, with decreased production serving to retard the normal development of absorptive capacity, then the increase in the pressure in the choroid plexus capillaries brought about by both the closure of the fistulae and the subsequent venous thrombosis may have resulted in a rate of CSF production greater than could be handled by the absorptive system. Resolution of the thrombus, recruitment of venous collaterals, and possibly an increase in absorptive capacity would have resulted in the resolution of the syndrome.

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 relationship 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.

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

CHRISTOPHER D KOLLAR
Madeline Foundation Laboratory,
University of Sydney, Australia

IAN H JOHNSTON
Department of Neurosurgery, Royal Alexan dra Hospital for Children, Sydney, Australia

Correspondence to: Correspondence to: Dr Christopher Kollar, Madeline Foundation Laboratory, Room 323, Building D06, University of Sydney, 2006, Sydney, Australia. Telephone 0061 2 9351 3359; fax 0061 2 9351 4887; kollar@ surgery.usyd.edu.au

1 Symonds CP. Hydrocephalic and focal cerebral symptoms in relation to thrombophilias of the dural sinuses and cerebral veins. Brain 1937;60:251–50.
5 Bedford THB. The great vein of Galen and the syndrome of increased intracranial pressure. Brain 1934;57:1–24.

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

The polymerase chain reaction (PCR) has become reported to be of diagnostic value when performed on CSF samples in tuberculous meningitis.1–4 Rapid amplification of Mycobacterium tuberculosis specific DNA 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, antitu berculous 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...
M. tuberculosis is a highly contagious airborne organism that primarily affects the lungs, leading to pulmonary tuberculosis. However, it can also cause extrapulmonary manifestations, including meningitis, pericarditis, and disseminated disease.

In summary, the use of PCR in the diagnosis of tuberculosis is an important and valuable tool, especially in cases where culture is not feasible or negative. It offers a rapid and sensitive method for the detection of M. tuberculosis DNA, which can help in the early and accurate diagnosis of tuberculosis.


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 cerebrospinal fluid (CSF). These studies suggest that in some cases, PCR may be more sensitive than culture; however, in the largest study, performed by Nguyen et al., 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. Dalvi et al. also reported that multiple specimens may be required to improve the sensitivity of the test in some patients.

The PCR has been reported to detect the equivalent of 1–10 mycobacteria in vitro. However, lower sensitivity is found with clinical specimens. PCR positivity 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.

We are grateful to Dr Deborah Binzi-Gascogne of the Leeds mycobacterium laboratory, where the PCR tests were performed and who provided additional information for the manuscript.


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 characterized by distal muscle weakness and atrophy, and CMT type 2 by sensory symptoms, such as pain and numbness.

In this study, a novel mutation of the myelin P gene was identified in a patient with Charcot-Marie-Tooth disease type 1B, manifesting with trigeminal nerve thickening. The mutation was a missense mutation leading to a change in amino acid sequence of the myelin P protein. This mutation was absent in unaffected family members, suggesting a mendelian inheritance pattern.

Further studies are needed to investigate the molecular mechanisms underlying this mutation and its clinical implications. This study highlights the importance of genetic testing in the diagnosis and management of patients with Charcot-Marie-Tooth disease.
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).

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.

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” to C at codon 81) resulting in a substitution of arginine for histidine only in the patient. A DNA duplication in chromosome 17p11.2-12, including the peripheral myelin protein-22 (PMP 22) gene, was not present. The patient’s mother did not show any mutations in the P0 gene. CMT type 1 is caused by abnormalities in myelin protein of Schwann cells. Repeated demyelinating and remyelinating responses in the peripheral nerves causes diffusely enlarged diameters of nerves in CMT type 1, and thickening of the cauda equina, nerve roots, and ganglia has also been found. Although blepharoptosis, ophthalmoplegia, facial weakness, deafness, dysphagia, and 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 brainstem response suggested peripheral conduction delay of the auditory nerve. Trigeminal neurolgia with CMT has been reported. In these rare cases, trigeminal neurolgia 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 reported that the sensory component of the trigeminal nerve was relatively spared, despite extremely delayed conduction of this nerve. However, the MRI study of our patient suggested that the fifth cranial nerves were subjected to the same pathological process that affects other peripheral nerves.

Our patient showed no DNA duplication on chromosome 17p11.2 and we found a novel mutation (A to C) representing an Arg 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. 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.

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, leptomeningeal lymphomas are even rarer but have been described. However, no leptomeningeal lymphoma of the follicular type has previously been reported. We present a case of primary meningeal follicular lymphoma which mimicked a sphenoid wing meningioma, both radiologically and intraoperatively.

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

Brain CT showed an enhancing mass consistent with a right sided sphenoid wing meningioma. Careful comparison of the clinical, electroencephalographic, and histopathological data between patients with CMT should be conducted.

We are indebted to the families studied for their cooperation and support. This work was partly supported by Uehara Memorial Foundation, the Sasaki Health Science Foundation, the Primary Amelioration Research Committee, and the Ministry of Education, Science and Culture of Japan 10832002, 18832993.

MASAMI SHIZUKA
YOSHIKO IKEDA
MITSUONRI WATANABE
KOICHI OKAMOTO
MIKIO SHOJI
Department of Neurology, Gunma University School of Medicine, 3–39–22 Showa-machi, Maebashi, Gunma 371–8511, Japan

TORU I. KEGAMI
KIYOSHI HAYASAKA
Department of Pediatrics, Yamagata University School of Medicine, Yamagata, Japan

Correspondence to: Dr Masami Shizuka, Department of Neurology, Gunma University School of Medicine, 3–39–22 Showa-machi, Maebashi, Gunma 371–8511, Japan. Telephone 0081 27 220 8806; fax 0081 27 220 8086; email mshizuka@news.sb.gunma-u.ac.jp


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, leptomeningeal lymphomas are even rarer but have been described. However, no leptomeningeal lymphoma of the follicular type has previously been reported. We present a case of primary meningeal follicular lymphoma which mimicked a sphenoid wing meningioma, both radiologically and intraoperatively.
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 2:1.2

Malignant primary lymphoma can occur throughout the CNS and they often have a periventricular distribution. Multifocality seems to be more common in patients with AIDS. The CT scan usually shows hyperdense masses with peritumoral oedema and 92% enhance after administration of contrast medium.3

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.4

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;5 Vigushin et al.6 described a case of meningioma with a calcified temporoparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular rather than diffuse histological appearance.7

Rubinstein8 described a case of leptomeningeal 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.

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

Dominic J Hodgson
Karoly M David
Michael Powell
Department of Surgical Neurology

Jan L Holton
Department of Neuropathology, The National Hospital for Neurology and Neurosurgery

Francesco Pezzella
Department of Pathology, University College Hospital, London, UK

Determinants of the copper concentration in cerebrospinal fluid

The measurement of CSF copper concentration can serve as an indicator of brain copper concentration.1 However, the complex mechanisms by which copper crosses into the CSF, and the factors determining the CSF copper concentration in humans are largely obscure. Copper can pass into and out of the CSF by various mechanisms. For example, active transport through the blood-brain barrier or the blood-CSF barrier, or passive diffusion of the free or the bound fraction (bound to albumin or coeuroplasmin) 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 coeuroplasmin, CSF/serum albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum coeuroplasmin 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). Copeuroplasmin was determined nephelometrically (Beckman Array: Beckman Instruments, Brea, CA, USA). The age of the patients was 50.0 (SD15.5) years; 50 were women and 63 were men. Mean serum coeuroplasmin concentrations were 394.3 (SE1.7) mg/l. Mean serum copper concentrations were 1194 (SD 335) µg/l. Mean calculated free copper concentrations in serum were 78.5 (SD 1285) µg/l. Mean CSF copper concentrations were 14.16 (SD 6.0) µg/l. The mean albumin ratio (AR) was 6.63×10⁻³. The mean ratio of calculated free copper concentration to total serum copper was 6.6%, the ratio of CSF copper to serum copper was 1.2%, and the ratio of free serum copper to CSF copper was 18%. In the
Correlation of blood-CSF barrier (albumin ratio, (AR)) with total CSF copper concentration (on logarithmic axes): R^2=0.46, p=0.0001; 95% confidence bands for the true mean of the total CSF copper concentration are shown.

The data are fitted with a stepwise linear regression model (F to enter 4.0, F to remove: 3.996), significant positive parameters of the CSF copper concentration were found to be AR (p=0.0001) and serum coeruleoplasmin (p=0.0057). The other independent variables mentioned above showed no statistically significant relation with CSF copper concentrations. The figure shows the simple linear regression between CSF/serum albumin ratio and CSF copper concentration (on logarithmic axes): R^2=0.46, p=0.0001. The formula for the CSF copper concentration, derived from the multiple linear regression model, is: copper (g/l) = 5.32 μg/L/0.635 × CSF/serum albumin ratio (×10^-3) + 0.012 × serum coeruleoplasmin (mg/l).

According to this analysis, CSF/serum albumin ratio and serum coeruleoplasmin concentration together determine 25.3% of the variation in CSF copper concentration (adjusted R^2=0.253), implying that other (unknown) factors determine the remaining 74.7% of the variation. We have been able to demonstrate here that the CSF copper concentration is determined in a highly significant manner by disturbances in the blood-CSF barrier and by the serum coeruleoplasmin concentration. It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum coeruleoplasmin concentration, 20.7% of the measured CSF copper originates from the brain, the CSF by passive diffusion bound to coeruleoplasmin, and only around 0.9% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20×10^-3, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to coeruleoplasmin). 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 6.5×10^-3), that the CSF copper concentration is actually reduced by 27.4%, when the serum coeruleoplasmin concentration falls from its normal value of 394 μg/ml to 66 μg/ml. In consequence, CSF copper in patients with Wilson's disease is evidently substantially free, implying that a larger fraction than previously assumed of the raised CSF copper in patients with untreated Wilson's disease is originated from the plasma, the fraction entering the CSF by passive diffusion (bound to coeruleoplasmin) 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 coeruleoplasmin-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 coeruleoplasmin concentration. A statistical relation with a low correlation (p=0.05) between CSF protein content and CSF copper was already shown in 1987 in various neurological diseases;1 our study shows a much higher significance and, in addition, the effect of serum coeruleoplasmin (therefore of bound serum copper). Furthermore, we have been able to determine specifically the fraction of CSF copper which enters the CSF across the blood-CSF barrier.

HANS JOERGSTUERENBURG
MATTHIAS OECHSSNER
SVEN SCHROEDER
KLAUS KUNZIE
Neurological Department, University Hospital Hamburg-Eppendorf, Hamburg, Germany. Telephone 0049 40 4717 4832; fax 0049 40 4717 4086.

Correspondence to: Dr Hans Joerg Stuermen, Neurological Department, University Hospital Hamburg-Eppendorf, Martinistrasse 52, 20246 Hamburg, Germany.

References

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 clinical and radiological investigation results in vascular or bronchial obliteration.

1. A 20 month Irish boy, the only son of healthy, unrelated parents, was admitted for investigation of a large head. He had one previous hospital admission at the age of 6 weeks for a respiratory tract infection. Transient hypotonia was noted at that time as was his skull circumference of 43 cm. At 6 months there was no hypotonia, neurological examination was normal, and the head circumference was 49 cm. The father's head circumference was 61 cm and he stated that all of his family had “big heads”. By 20 months, the patient's head circumference measured 55.6 cm and was diverging from the 97th centile. Brain CT showed a well circumscribed, contrast enhancing mass in the midline and left frontal lobe, with surrounding oedema. There was evidence of left sided hydrocephalus due to displacement of the right foramen of Munro by tumour. The radiological differential diagnosis included a primary meningeal tumour, glioma, and leukaemic deposit. The patient underwent a left fronto-craniotomy and a firm, rounded mass was removed in brain. Histologically, this was a myofibroma. focal necrosis, or calcification. The tumour cells appeared to blend with the vessel walls. An atypical distribution of the vessels led to erroneous diagnosis. The mass was not attached to the falx, but was firmly adherent to the left pericoronal artery. A fragment (4 mm×2 mm) had to be left attached to the vessel. The histological picture was compatible with solitary intracranial myofibroma. The patient underwent a left fronto-craniotomy and a firm, rounded mass was removed in brain. Histologically, this was a myofibroma. Histologically, this was a myofibroma. The patient underwent a left fronto-craniotomy and a firm, rounded mass was removed in brain. Histologically, this was a myofibroma. The patient underwent a left fronto-craniotomy and a firm, rounded mass was removed in brain. Histologically, this was a myofibroma.
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofilaments.

Solitary lesions of infantile myofibromatosis are more common than multiple lesions, with twice as many males as females being afflicted, and generally involve the skin and soft tissues, especially of the head and neck. Solitary lesions are less commonly found in viscera or bones. Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported along with few cases of CNS involvement in the generalised form of infantile myofibromatosis.10 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 meningeoma, schwannoma, and haemangiopericytoma. Regionally, the histology was reminiscent of the rare microcystic variant of meningioma. Meningeomas are extremely rare in this age group, this lesion was not meningeal based and such lesions 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 schwannoma, did not show any 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 extracranial masses in another, both of which were fatal at the age of 10 days, a non-fatal extracranial mass in one patient, and a patient with systemic involvement, in which there was recurrence of orbital and temporal lesions 2 years after operation. A single previous case of solitary intracranial myofibroma has been reported in which the patient died within 24 hours of surgery, secondary to cardiorespiratory arrest.

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

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

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

Escherichia coli serotype O157:H7 causes serious food poisoning worldwide, especially in children and elderly people.1 It is also called verotoxin producing E coli (VTEC), which produces a potent Shiga-like toxin. Gastrointestinal, haemorrhagic, and uremic effects are well known in VTEC infection,2 and neurological problems are likely to be more frequent than is generally recognised.3 Here we describe axonal polyneuropathy and encephalopathy in a young female patient associated with haemolytic-uraemic syndrome caused by VTEC infection.

A 26 year old woman began to have abdominal pain and haemorrhagic diarrhoea. She was admitted to an emergency hospital and diagnosed as having haemorrhagic colitis due to probable food poisoning. Then her urinary volume gradually decreased and anuria and her blood analysis showed severe haemolytic-uraemic syndrome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised kidney cell line, are highly sensitive to doses of verotoxin through the process of apoptosis.4 Verotoxin shows similar cytotoxicity on human glomerular microvascular endothelial cells via lysis and apoptosis such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis.5 Our patient was treated with antibiotics, plasma exchange, and continuous haemodialysis, with benefit.

During the course of the disease, our patient was in a delirious state with visual hallucinations and tonic convulsion. She showed mild brain swelling on CT and diffuse slow waves in the frontal area on EEG, evidence of encephalopathy. Previous reports have shown that the incidence of encephalopathy in haemolytic-uraemic syndrome (haemolytic-uraemic syndrome caused by VTEC infection) is 3.2%–10%, 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.6 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 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, manifesting as tingling of the legs. She described her feet 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 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 haematochemical studies, serum chemistry, urinalysis, and CSF analysis were normal. Serum concentrations of vitamin B1, B6, and B12 were normal. Nerve conduction studies were carried out on her right limbs, and showed normal findings in the distal latencies, motor conduction velocities, and F wave latencies of the median, ulnar, and tibial nerves, and no evidence of conduction block. However, there were decreased amplitudes of the sensory nerve action potentials (1.18 mV) and slow motor of conduction velocity (41.0 m/s) in the peroneal nerve. There were also markedly decreased amplitudes of the sensory nerve action potentials (2.86 mV) and sural (0.98aV) nerves. These findings and the clinical features confirmed the diagnosis of sensory dominant, axonal polyneuropathy. She was given 300 mg/day sulindac (an anti-inflammatory agent) and 1500 µg/day mechofolamin (vitamin B12) without effect. Two weeks after administration of 300 mg/day oral mecloretin, 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 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 kidney cell line, are highly sensitive to doses of verotoxin through the process of apoptosis.4 Verotoxin shows similar cytotoxicity on human glomerular microvascular endothelial cells via lysis and apoptosis such as tumour necrosis factor-a, which induced an increase in the numbers of verotoxin receptors, leading to a microvascular thrombosis.5 Our patient was treated with antibiotics, plasma exchange, and continuous haemodialysis, with benefit.

Correspondence to: Dr Ryuji Sakakibara, Department of Neurology, University of California at Los Angeles, West Los Angeles Veterans Affairs Medical Center, Los Angeles, CA, USA

Correspondence to: Dr MF Mendez, Neurobehavioral Unit (H91116A/F), West Los Angeles VA Medical Center, 11301 Wilshire Blvd, Los Angeles, CA 90073, USA. Telephone 001 310 487 3711 ext 42606; fax 001 310 268 4181; email mmendez@ucla.edu


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 followed by left sided parasthesias. Around 600 am he awoke with a 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 paraesthesia in his trunk or right leg. 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 crying spells but abruptly resolved shortly after his last crying spell. This patient had hypertension, diabetes mellitus, coronary artery disease, an old myocardial infarction, raised cholesterol concentrations, and a history of heavy smoking.

On examination between recurrent crying spells, his blood pressure was 143/92 with a regular pulse of 62, and there were no carotid bruits. His mental status was normal. Cranial nerve examination was normal, with no 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, 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 capping over the right frontal horn and an EDV of the right frontal interictal rhythmic delta activity but no epileptiform changes. Carotid Doppler studies showed atheroocclusive changes without haemodynamically relevant obstruction. He was discharged on antiplatelet therapy with aspirin.

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

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

Crying or dacrystic seizures also occur but are rare. These seizures are part of the range of complex partial seizures and usually emanate from the right temporolimbic system.2 Crying seizures may result from prior cerebrovascular infarctions.3 Although our patient had mild right hemisphere ischaemia, he had no other evidence suggesting definite seizure activity.

It is likely that this patient had a single transient ischaemic attack with multiple crying spells. The localisation of his attack is unclear; involvement of the right thalamus or neighbouring internal capsule is a possibility. Similar to spells of laughter, spells of crying may occur in relation to unilateral cerebrovascular events. Although most reports of crying after unilateral strokes have reported left hemispheric lesions,4 crying also may result from right hemispheric strokes.5 Even more similar to our patient, sudden laughing spells, “le fou rire prodromique,” rarely precede strokes involving the left capsular-thalamic, lenticulocaudate, or pontine regions.6 Our patient may have had a comparable phenomenon from the right hemisphere or unilateral cells anism for this phenomenon may have been temporary activation or stimulation of ischaemic motor pathways.

MARIO F MENDEZ
YURI L BRONSTEIN
Department of Neurology, University of California at Los Angeles, West Los Angeles Veterans Affairs Medical Center, Los Angeles, CA, USA

Correspondence to: Dr MF Mendez, Neurobehavioral Unit (H91116A/F), West Los Angeles VA Medical Center, 11301 Wilshire Blvd, Los Angeles, CA 90073, USA. Telephone 001 310 487 3711 ext 42606; fax 001 310 268 4181; email mmendez@ucla.edu

Continuous drop type of orthostatic hypotension

Orthostatic hypotension has usually been evaluated for 2–10 minutes after standing.7,8 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 before the 25 minute test of 60° head up tilt. Blood pressure and heart rate were recorded every minute with an automatic sphygmomanometer. Patients could clearly be classified into two groups in terms of the time taken to reach the minimum blood pressure. In 12 patients systolic blood pressure fell rapidly, reached a minimum within 5 minutes, and then remained stable or partially recovered (early drop type); whereas, in 13 patients blood pressure fell immediately after tilting but kept decreasing by more than 8 mm Hg from that at 5 minutes (mean 12.8 mm Hg;
with orthostatic hypotension of the continuous drop type. 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. As we did not record heart rate between the early drop type and the continuous drop type, the mechanisms for continuous reduction of cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline (norepinephrine) level (+0.05 ng/ml) during the decrease in blood pressure.

Our results suggest that in many patients with MSA the blood pressure drops continuously on standing. The continuous blood pressure drop is caused by continuous reduction of cardiac output. A part of the mechanism for continuous reduction of cardiac output should be lack of reflex tachycardia and no significant release of noradrenaline which are caused by interruption of the baroreflex arc, as is known in MSA. However, further explanation, such as continuous vasodilatation of the volume vessels, is necessary for the difference in mechanisms between the early drop type and the continuous drop type. We did not record heart rate and blood pressure continuously and did not evaluate ventricular function by echocardiography, the final conclusion and its interpretation require further study.

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

TAKANORI YOKOTA
KAZUO MITANI
YUKINOBU SAITO
Department of Neurology

TOSHIYUKI ONIKI
Third Department of Internal Medicine, Tokyo Medical and Dental University, Tokyo 113, Japan

MICHIYUKI HAYASHI
Department of Neurology, Tokyo Metropolitan Neurological Hospital, Tokyo 183, Japan

Correspondence to: Dr Takanori Yokota, Department of Neurology, Tokyo Medical and Dental University, 1-5-45 Yushima, Bunkyo-ku, Tokyo 113-8519, Japan. Telephone +81-3-5803-5234; fax +81-3-5808-0169.


CORRESPONDENCE

Respiratory aspects of neurological disease

An account of respiratory aspects of neurological disease, such as the highly informative one presented,1 would be incomplete without mention of breathlessness resulting from neurogenic pulmonary oedema, characterised by an “increase in extravascular lung water in patients who have sustained a change in neurological condition”. Neurological disorders associated with this syndrome include subarachnoid haemorrhage, middle cerebral artery stroke, and cerebellar haemorrhage.2 Brain stem stroke, acute hydrocephalus due to colloid cyst of the third ventricle, closed head injury, and status epilepticus, were also documented as risk factors in a literature review by Smith and Matthay,2 who proposed, on the basis of their own study, that increased pulmonary vascular hydrostatic pressure might be a more significant aetiopathogenic mechanism than increased pulmonary capillary permeability.3 A more direct link between neurogenic myocardial damage and pulmonary oedema can be postulated when subarachnoid haemorrhage is complicated by reversible severe left ventricular dysfunction, as documented in two cases reported by Wells et al.4

O M P JOLOBE
Department of Medicine for the Elderly, Tameside General Hospital, Fountain Street, Ashton under Lyne OL6 9RW, UK
Idiopathic cerebellar ataxia associated with celiac disease: lack of distinctive neurological features

Although applauding the contribution of Pelliceria et al on 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 gastrointestinal 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 cerebellar 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 antientomysiom 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 antientomysiom 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.

M HADJIVASSILIOU
R A GRUNEWALD
G A B DAVIES-JONES
Department of Neurology, Royal Hallamshire Hospital, Glossop Road, Sheffield S10 2JF, UK

Correspondence to: Dr G A B Davies-Jones, Department of Neurology, Royal Hallamshire Hospital, Glossop Road, Sheffield S10 2JF, UK


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.

M P O'PKEY

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 myotonic dystrophy in two siblings with severe myotonic dystrophy.

In the authors’ experience, long term results of both medical and surgical management of the faecal incontinence are disappointing. The patient—a 19 year old man—had had his illness diagnosed 4 years earlier on clinical grounds and electro-physiological and genetic tests. Early sympotms of sphincter 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 on 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, 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 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 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 patterns in patients with myotonic dystrophy shows a multitude of deficits 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,3 by using 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.

GPELLICCI
O SCARPINO
Department of Neurology, IRRCAG, Geric Hospital, Ancona, Italy

V PILONI
Department of Radiology, Azi. N 7, Ancona, Italy

Correspondence to: Dr Giuseppe Pelliccion, Department of Neurology, Geric Hospital, via della Macerata, 66110 Ancona, Italy. Telephone: 0039 071 8003432; fax 0039 071 8003530; email: o.scarpino@fastnet.it


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 1888, furnished with exquisite graphic illustrations. To this effect, we draw attention to prior descriptions of the same syndrome, reported by Vulpian in 1886 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, we have come to use the eponym of Vulpian-Bernhardt’s syndrome to describe those forms of amyotrophic lateral sclerosis with more or less symmetric involvement of the proximal muscles of the upper limbs at the clinical onset. A certain enigma exists surrounding the characteristic distribution of weakness and muscle atrophy. The reason for the prevalence in the proximal muscles of the upper limbs is unknown. We can furnish little more information in this respect. However, in the 1960s, in the differential diagnosis of this syndrome, it was proposed that the muscles predominantly affected in Vulpian-Bernhardt’s form were the deltoideus, the infraespinaeous, the supraspinatus, the sternocleidomastoideus, and the teres minor. The predominance of these muscular involvements in these muscular classes permitted its distinction from that previously called Erb’s dystrophy.4

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, abductor 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 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, since it too has only 117 subjects, yet the selection criteria are intriguing. This is the first true inception cohort study where 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.

The Swiss study may be useful for comparison because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by the selection atrocity of advertising for subjects, and has a host of other reportedly fatal faults, and giving some benefit of the doubt, the study is said to be an accurate representation of the state of affairs in Switzerland at that time. The figures expounding myths of whiplash would find the Swiss study’s data most interesting.

The study of 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, since it too has only 117 subjects, yet the selection criteria are intriguing. This is the first true inception cohort study where 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.

The Swiss study may be useful for comparison because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by the selection atrocity of advertising for subjects, and has a host of other reportedly fatal faults, and giving some benefit of the doubt, the study is said to be an accurate representation of the state of affairs in Switzerland at that time. The figures expounding myths of whiplash would find the Swiss study’s data most interesting.

Thus, while the sample size is small in this Lithuanian study, it is comparable with others reporting the prognosis of whiplash, and yet gives a different picture of outcome.
A second consideration is that perhaps these Lithuanians are in very minor collisions.

True, some of their vehicles were completely wrecked, but perhaps the vehicles were not very good quality and so were easily damaged. Perhaps that is why this cohort had such a good outcome and only minor injuries. This is an unhelpful consideration however, as studies in Canada have shown that those with absolutely no vehicle damage, in very low velocity collisions, are just as likely to report chronic pain as those in more severe collisions. Lithuanians seem to behave appropriately then for minor collisions (if that is what they indeed had), but Canadians seem unable to behave appropriately. Again, another cultural rift 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.

If there were strict exclusion criteria to identify chronic pain, 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. The time has indeed 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 opportunity to help prevent the chronic pain and the suffering we otherwise encounter.

R FERRARI

Acknowledgements


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 management. There are sections on pain mechanisms and pain as a chapter in the pharmacology of acute and chronic pain, and other chapters on postoperative pain, obstetric pain, and acute paediatric pain. There are three further chapters specifically on the management of chronic low back pain, cancer pain, and an overview of interventional pain techniques.

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

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

The chapter on pharmacology of acute and chronic pain is well written, but unfortunately a lot of time is spent on non-steroidal drugs. There is a review of the adjuvant drugs such as antidepressants and anticonvulsants that are used in chronic pain, however one is left at the end with a sense of knowing about the drugs but not quite to use them. There is no mention of the increasing use of gabapentin or nor of other drugs that are sometimes used in chronic pain states such as cloxidine and other sympathetic agents or calcium channel blockers.
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 reconstructed in compression. The remaining chapters cover fracture management, late kyphosis, metastatic tumours, spondylolisthesis, degenerative spinal disease, and infection. Each chapter sets out the principles of management which are illustrated schematically. There then follow case studies illustrated by radiological images including CT and MRI. These have reproduced well and surgeons will admire the technical precision and excellent anatomical reductions illustrated by these clinical cases. It is, however, a source of constant annoyance to spinal surgeons that perfect postoperative films do not always correlate with good clinical results and this discrepancy remains a source of fascination and mystery.

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

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

RODNEY LAING


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

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

IAN WHITWORTH