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

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

The patient complained of right sided headache for 24 hours after the procedure, resolving with minor analgesia. Brain CT the next day was reported as normal. A full ophthalmological review was undertaken before discharge showing normal fundi and fields. Ten days after the embolisation the patient presented with a generalised, pounding headache, present since discharge. Examination showed mild left papilloedema, with no focal meningitis. CSF showed mild left papilloedema, with no focal meningitis.

A week later she 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 meningitis. CSF showed mild left papilloedema, with no focal meningitis.

A week later she 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 meningitis.
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 when tuberculous meningitis is clinically suspected.

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.

M. MELZER
T J BROWN
Department of Microbiology, St Thomas’ Hospital, Lambeth Palace Road, London SE1 7EH, UK

J FLOOD
S LACEY
L R BAGG
King George Hospital, Bartley Lane, Goodmayes, Essex IG1 6YB, UK

Correspondence to: Dr M Melzer, Department of Microbiology, St Thomas’ Hospital, Lambeth Palace Road, London SE1 7EH, UK.


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. Dalovich 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. PCR 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.

D M GASCOYNE-BINZI
P M HAWKEY
Department of Microbiology, The General Infirmary at Leeds, Great George Street, Leeds LS1 3UK, UK

Correspondence to: Dr D M Gascoyne-Binzi, Department of Microbiology, The General Infirmary at Leeds, Great George Street, Leeds LS1 3UK, UK.


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 includes CMT1A, CMT1B, CMT2, and X chromosome (CMTX). CMT1A and CMT2 are the most common forms, accounting for about 70% of all cases. CMT1A is caused by mutations in the myelin protein zero (P) gene. Mutations in the P gene have recently
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 decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A→C at codon 81) resulting in an amino acid substitution of histidine for arginine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. However, other cranial, spinal, and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A→C at codon 81) resulting in an amino acid substitution of histidine for arginine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. However, other cranial, spinal, and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A→C at codon 81) resulting in an amino acid substitution of histidine for arginine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. However, other cranial, spinal, and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A→C at codon 81) resulting in an amino acid substitution of histidine for arginine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. However, other cranial, spinal, and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.

The six exons of the P0 gene were amplified by the polymerase chain reaction using primers, and analysed by single strand conformational polymorphism (SSCP) and sequencing analyses. DNA sequencing of exon 3 showed a novel point mutation (A→C at codon 81) resulting in an amino acid substitution of histidine for arginine only in the patient. A DNA duplication in chromosome 17p11.2-p12, including the peripheral myelin protein-22 (PMP-22) gene, was not present. However, other cranial, spinal, and distal muscles of the legs, decreased grasping power, sensory disturbance of distal limbs, and decreased hearing acuity. Facial sensation, mastication power, and hearing acuity were normal. She also had atrophy of the lower limbs, drop foot, a steppage gait, claw hand deformities, optic atrophy, incoordination, autonomic dysfunction, and cardiac involvement were not evident.

In laboratory findings, creatinine kinase was 343 IU/L. A peripheral nerve conduction study showed undetectable sensory and motor action potentials in all limbs. Auditory brain stem response showed 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 and peripheral nerve conduction study. Blood samples were obtained from the patient and her mother with informed consent. DNA was extracted from the blood by a standard phenol/chloroform protocol.
chemical 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 negative. Postoperative adjuvant whole brain or localised radiotherapy was discussed with the patient, however, she declined any further intervention. She has been closely reviewed in the follow up clinic and after 6 months there has been no clinical or radiological evidence of relapse.

Primary intracerebral lymphomas represent about 2% of intracranial neoplasms and 2% of all lymphomas. They occur most commonly in the 6th decade of life with a female to male ratio of 2:1. The association between primary intracranial lymphoma and immunodeficiency has long been established, and it is not surprising, therefore, that the incidence has increased 10-fold over the past 3 decades with the onset of transplant surgery and, particularly, the AIDS epidemic. In postmortem studies, these neoplasms are found, on average, in 5.5% of AIDS cases, and malignant cerebral lymphoma is the most common diagnosis of a focal intracranial lesion in patients with AIDS. 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.

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 intrinsic 2:1 lymphomas have been mistaken for more common lesions: solitary primary B cell lymphoma of the cerebellar pontine angle mimicking acoustic neurilemoma or meningioma has been reported; Vugshin et al reported a patient with a calcified temporoparietal lymphoplasmacytic lymphoma which resembled a meningioma; however, this tumour was entirely extradural. There is only one previous report of a follicular rather than diffuse intrinsic primary leptomeningeal lymphoma. 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 splenoid wing meningioma suggest that this entity should be considered as a rare differential diagnosis.

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 coeulioplasmin) 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 coeulioplasmin, CSF serum albumin ratio, total serum copper concentration, and calculated serum free copper concentration (based on serum coeulioplasmin 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). Coeulioplasmin 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 coeulioplasmin concentrations were 394.3 (SD 74.7) mg/l. Mean serum copper concentrations were 1194 (SD 335) mg/l. Mean calculated free copper concentrations in serum were 78.5 (SD 1285) mg/l. Mean CSF copper concentrations were 11.16 (SD 6.00) mg/l. The mean albumin ratio (AR) was 6.63×10^-4. 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%.
stepwise linear regression model (F to enter 4.0, F to remove: 3.996), significant positive predictive power of this model. CSF copper concentration were found to be AR (p=0.0001) and serum coeruloplasmin (p=0.0057). The other independent variables mentioned above showed no statistically significant relation with CSF copper concentration. The figure shows the simple linear regression between CSF/serum albumin ratio and CSF copper concentration (on logarithmic axes; R=0.46, p=0.0001). The formula for the CSF copper concentration, derived from the multiple linear regression model, is: copper (µg/l)=5.32 µg/l0.653 × CSF/serum albumin ratio (×10-3)+0.012×serum coeruloplasmin (mg/l). According to this analysis, CSF/serum albumin ratio and serum coeruloplasmin together determine 25.3% of the variation in CSF copper concentration (adjusted R=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 coeruloplasmin concentration. It can be assumed from this that in the case of normal blood-CSF barrier function and a normal serum coeruloplasmin concentration, 25.3% of the measured CSF copper concentration is due to the CSF by passive diffusion bound to coeruloplasmin, and only around 0.09% by passive diffusion bound to albumin. In the case of a markedly raised CSF/serum albumin ratio of 20×10³, this would mean that 60.6% of the measured CSF copper originated from the blood (bound to coeruloplasmin). A variable fraction of the CSF copper concentration, depending on the degree of damage to the blood-CSF barrier, therefore crosses from the blood into the CSF and can be measured there. Our formula would therefore predict, in patients with Wilson’s disease with 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 coeruloplasmin concentration falls from its normal value of 394 mg/l to 66 mg/l. In consequence, CSF copper in patients with Wilson’s disease is evidently substantially free, implying that a larger fraction than previously assumed of the raised CSF copper in patients with untreated Wilson’s disease originates from the brain, the fraction entering the CSF by passive diffusion (bound to coeruloplasmin) tends towards zero. It can be concluded from this that, when the aim of therapy is considered in terms of the total CSF copper concentration, a region around 30% lower than the upper limit of the normal range should be aimed for. This is supported by the clinical finding that patients report feeling better when the CSF copper concentration is below this value. This analysis also shows that the raised copper concentrations in the CSF can only originate from the brain. In particular, it is not associated with free serum copper, but evidently only via storage in the brain. The investigation here also shows that, after determining the CSF copper concentration, the coeruloplasmin-bound fraction originating from the plasma should be subtracted according to the formula we have given, or better, all measured copper concentrations in the CSF should be adjusted using the CSF/serum albumin ratio and serum coeruloplasmin concentration. A statistical relation with a low correlation (p=0.05) between CSF protein content and CSF copper was already shown in 1981 in this variety of neurological diseases; our study shows a much higher significance and, in addition, the effect of serum coeruloplasmin (therefore of bound serum copper). Furthermore, we have been able to determine quantitatively the fraction of CSF copper which enters the CSF across the blood-CSF barrier.

HANS JOERGSTUERENBURG
MATTHIAS OECHSNER
SVEN SCHREIDER
KLAUS KUNZE
Neurological Department, University Hospital Hamburg-Eppendorf, Martinistrasse 52, 20246 Hamburg, Germany.

Correspondence to: Dr. Hans Joerg Stuerenberg, Neurological Department, University Hospital Hamburg-Eppendorf, Martinistrasse 52, 20246 Hamburg, Germany. Telephone 040 40 4717 4832; fax 040 40 4717 5086.

2. Weisner B, Hartard C, Dies C. CSF copper concentration: a new parameter for diagnosis and was seen. No mitotic figures were present and transient parasites of the right leg, which subsequently resolved completely. Repeat CT 6 months later and at 4 years after the operation showed no evidence of recurrence or mass effect. His head circumference persisted on the 97th centile 4 years after operation. His development and clinical examination otherwise remain normal 6 years after surgery. A younger sibling is normal.

A 20 month old Irish boy, the only son of healthy, unrelated parents, was admitted for investigation of a large head. He had previous hospital admission at the age of 6 weeks for a respiratory tract infection. At birth, muscle 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 meningioma tumour, glioma, and leukaemic deposit. The patient underwent a left frontal craniotomy and a firm, rounded mass was removed from between the frontal lobes. The mass was not attached to the falx, but was firmly adherent to the left pericarinal artery. A fragment (4 mm x 2 mm) had to be left attached to the vessel wall. The mass had a fibrous, white-yellow cut surface appearance. Microscopically, it consisted of hypercellular fasciculated and storiform areas, alternating with hypocellular, hyalinised regions. Central, a haemorrhagic necrosis-like area was seen. No mitotic figures were present and there was no evidence of haemorrhage, necrosis, or calcification. The tumour cells appeared to blend with the vessel walls. Immunohistochemical studies showed strong reactivity for vimentin and smooth muscle actin. Scattered cells showed immunoreactivity for desmin. No reactivity was noted for cytokeratin, epithelial membrane antigen, factor VIII, gial fibrillary acidic protein, or...
myoglobin. Ultrastructural examination showed elongated cells with surrounding collagen fibrils, some showing intracytoplasmic myofilaments.

Solitary lesions of infantile myofibromatosis are more common than multiple lesions, with twice as many males as females being affected, and generally involve the skin and soft tissues, especially of the head and neck.1 Solitary lesions are less commonly found in viscera or bones.1,2 Involvement of the CNS is exceedingly rare and only one other case of a solitary mass is reported3 along with few cases of CNS involvement in the generalised form of infantile myofibromatosis.3 The progno-

sis 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 schwannoma, showed no immunoreactivity for S-100 protein. Haemangiopericytoma is a diagnosis of exclusion and shows no reactivity for actin, unlike this tumour.

Perivascular and intracranial involvement by myofibromatosis includes patients with widespread systemic involvement and multiple leptomeningeal nodules1 in one patient and extramedullary masses in another,2 both of which were fatal at the age of 10 days, a non-fatal extramedullar mass in one patient, and a patient with systemic involvement, in which there was recurrence of orbital and temporal lesions 2 years after operation. A single previous case of solitary intracranial myofibroma has been reported3 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 postopera-
tive outcome. Although rare, infantile myofibroma should be included in the differential diagnosis of intracranial neoplasms in children.

We acknowledge the expert assistance of Drs Lucy Roarte and Dr Lewis Dehner in diagnosing this case.

C B O’SUILLIEBHAIN
C J MARKS
Department of Neurosurgery
D RYDER
Department of Radiology
C KEOHANE
M J O’SULLIVAN
Department of Pathology, University College Cork, Cork University Hospital, Wilton, Cork, Ireland

Correspondence to: Dr. M.J. O’Sullivan, Lauren V Ackerman Laboratory of Surgical Pathology, Washington University Medical Center, PO Box 8118, 660, South Euclid Avenue, St Louis, MO 63110, USA. Telephone 001 314 362 0101; fax 001 314 362 9590.

1 William JO, Schrum D. Congenital fibrobro-


3 Chung EB, Enzinger FM. Infantile myofi-


neouveau-ne a evolution regressive. Ann Paedi-
atre (Paris) 1965;92:96–98.

5 Teng P, Warden MJ, Cohn WL. Congenital gen-


8 Adickes ED, Goodrich, P, AuchMoody, J, et al. Central nervous system involvement in con-


12 Enzinger F, Weiss S. Fibrous tumors of infancy. In: Soft tissue tumors, ed. C KEOHANE

C J MARKS

2 Second soft tissue tumor, ed. D RYDER


14 Zschoch H, Poley F. Angeborene generalisierte


16 Chung EB, Enzinger FM. Infantile myofi-


C J MARKS

20 Second soft tissue tumor, ed. D RYDER

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.2 Gastrointestinal, haemorrhagic, and uremic effects are well known in VTEC infection,1,2 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 syn-
drome 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 renal dysfunction (increased serum creati-
inase, 230–460 IU), suggestive of haemolytic anemia, was noted. The patient died 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 joint sensation. Routine laboratory data including haematological studies, serum chemistry, urinalysis, and CSF analysis were normal. Serum concentrations of vitamin B1, B6, and B12 were normal. Nerve conduction studies were carried out on her right limbs, and showed normal findings in the distal latencies, motor conduction velocities, and F wave latencies of the median, ulnar, and tibial nerves, and no evidence of conduction block. However, there were decreased amplitudes of the sensory nerve action potentials (1.18 mV) and mild slowing of motor conduction velocity (41.0 m/s) in the peroneal nerve. There were also markedly decreased amplitudes of the sen-
sory nerve action potentials (0.27 mV) and 1500 µg/day mecholamin (vitamin B12) without effect. Two weeks after administra-
tion of 300 mg/day oral meexitlin, her numb-
ness and pain gradually disappeared.

The patient was diagnosed as having VTEC infection, because of a typical history of an acute haemorrhagic colitis, the cultured strain of E coli O157:H7, and the detection of verotoxin in her stool. She had haemolytic-uraemic syn-
drome (haemolytic anaemia, thrombocytopenia, and uraemia, following diarrhoea), which is the main complication of VTEC infection. Experimentally, vero cells, an immortalised primate kidney cell line, are killed by low doses of verotoxin through the process of apoptosis.3 Verotoxin shows similar cytotoxicity on human glomerular mesangial vascular endothelial cells via inflammation, such as tumour necrosis factor alpha, which induces an increase in the numbers of verotoxin receptors, leading to a microvascular-
lar thrombosis.4 Our patient was treated with antibiotics, plasma exchange, and continuous haemodialysis, with benefit.

During the course of the disease, our patient was in a delirious state with visual hallucinations and tonic 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 encephalopa-
thapy in haemolytic-uraemic syndrome (mostly of VTEC infection) is 30% to 52%,5 having seizures in 7%–40%, and paralysis in 1%–16%. Many of the patients, including ours, seemed to have metabolic encephalopathy,5 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 injurious lesion is in the hypothalamic areas, then 254

Letters, Correspondence, Book reviews
Crying as a symptom of a transient ischaemic attack

In the absence of depression, crying spells associated with neurological disease usually result from pseudobulbar palsy or, more rarely, from crying seizures. To our knowledge, there are no prior reports of crying spells heralding or signifying a transient ischaemic attack. We report on a patient with prominent cerebrovascular risk factors who had a transient episode of intractable crying and focal neurological findings.

The patient was a 55 year old right handed man who presented with acute, uncontrolled crying spells following by left sided paresthesias. Around 6:00 am he awoke with a cataclysmic 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 left side of the body. He did not have photophobia, nausea or vomiting, blurred vision, visual obscurations, difficulty swallowing, dysarthria, or focal weakness. Over the next 2 to 3 hours, he had five more crying spells, each lasting 5 to 10 minutes, occurring out of context, without precipitating factors or sadness, with an acute onset and offset, and without alteration of consciousness. The patient’s left face and arm numbness persisted during and between the 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 showed right palpebral fissure narrowing, 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 which 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, paraesthesiae, 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 edema over the right frontal horns, and an ECG revealed focal, intermittent rhythmic delta activity but no epileptiform changes. Carotid Doppler studies showed atherosclerotic changes without haemodynamically relevant obstruction. He was discharged on antplatelet 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 hypesthesia 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 nerve innervation of bulbar motor nuclei and brainstem centres. In addition to crying, pseudobulbar palsy may include dysarthria, dysphagia, bifocal weakness, increased facial and mandibular reflexes, and weak tongue movements. There were no signs or symptoms of pseudobulbar palsy in this patient.

Continuous drop type of orthostatic hypotension

Orthostatic hypotension has usually been evaluated for 2–10 minutes after standing.2,3 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 12.9 mm Hg;
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 (norepinephrine) level (+0.05 ng/ml) during the decrease in blood pressure. 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. As we did not record heart rate and blood pressure continuously and did not evaluate ventricular function by echocardiography, the final conclusion and its interpretation require further study.

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

Continuous drop type of orthostatic hypotension during 25 minute tilt up in a patient with MSA. SBP=systolic blood pressure; HR=heart rate; CO=cardiac output; SVR=systemic vascular resistance; NA=plasma noradrenaline concentration.
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 comments 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 neurological features are common in these patients, found in two out of three of the patients of Pellecchia et al. and 21 of our 28. 1 We agree that gastrointestinal symptoms are rare: rather than entitling their paper “lack of distinctive neurological features”, perhaps “lack of distinctive gastroenterological features” might have been more appropriate!

We were surprised at the high specificity and sensitivity of increased antigen gliadin antibodies to be invaluable screening tools in patients with ataxia, only 11 of our 28 patients with increased antigen gliadin antibodies had histology of overt coeliac disease on duodenal biopsy, the remainder having normal or non-specific inflammatory changes but with an HLA genotype in keeping with gluten sensitivity. It is interesting to note that despite the often quoted high sensitivity for coeliac disease of increased antigliadin antibody titres, such was found in only one of three patients of Pellecchia et al. with coeliac disease. This concurs with our impression of the very modest sensitivity of antigliadin antibodies in gluten ataxia.

Gluten sensitivity is common in patients with ataxia, and can be identified by increased antigen gliadin antibody titres in the presence of appropriate histocompatibility antigens. 2 Although the clinical features of gluten ataxia are not entirely specific, they are distinctive.


Pellecchia et al. reply:

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

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

We understand that some discrepancies arise comparing our study with that of Hadijvassiliou et al. Firstly, only six out of their 28 patients had evidence of cerebellar atrophy on MRI, whereas all of our patients had cerebellar atrophy. Secondly, many of their patients had a peripheral neuropathy in the absence of cerebellar atrophy. This finding could explain the 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 celiac disease on duodenal biopsy among patients with idiopathic cerebellar ataxia, whereas none of the six patients with cerebellar atrophy described by Hadijvassiliou et al. showed histological features of celiac disease. It would be interesting to know the prevalence of gluten ataxia among all ataxic patients screened for antigliadin by Hadijvassiliou et al. Our series is too small to estimate the sensitivity of both antigliadin and antidentomyosin antibodies in gluten ataxia; unfortunately Hadijvassiliou et al. did not report any data on antidentomyosin 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 Hadijvassiliou et al. in a previous report. 3 This is in contrast with the 2% of antigliadin antibody positivity found in a large population by Catassi et al. 4 Further studies are required to better characterise the syndrome of cerebellar ataxia associated with celiac disease or gluten sensitivity.

M T PELLECCHIA
R SCALA
A FILLA
G DE MICHELE
P BARONE
Department of Neurological Sciences, Via S Panini 5, 80131 Naples, Italy


We read with interest the article by Abercrombie et al. which describes the pathophysiological and surgical management of faecal incontinence in two siblings with severe myotonic dystrophy. 1

In the authors’ experience, long term results of both medical and surgical management of the faecal incontinence was achieved, as proved by the fact that postanal sphincter repair restored faecal continence only for a brief time.

The authors’ pessimistic conclusions suggest that “faecal incontinence in myotonic dystrophy is difficult to relieve by any currently available treatment other than colostomy”. It should be noted, however, that the medical treatment used is not specified in the text.

Our experience with medical treatment using procainamide in a patient with severe myotonic dystrophy and faecal incontinence is less disappointing. 2 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-muscarlature, was performed. At physical examination, digital anorectal evaluation showed low squeeze pressures. A reduced rectal diameter (4.5 cm), anal gaping, and barometric 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, as in the first patient of Abercrombie et al., a decreased number of motor units and multiple myotonic discharges. Few motor unit potentials presented polyphasic waveforms and decreased duration and amplitude.

A regular treatment with procainamide (300 mg twice a day) 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 electromyographical investigation showed disappearance of myotonic discharges at the external anal sphincter, whereas defecography disclosed an improved rectal compliance (5.2 cm in diameter) at capacity and no more than a barium leak on straining.

The pathophysiology of motor disorders of the gastrointestinal tract in myotonic dystrophy is still debated and controversial. Histological study of the external anal sphincter and...
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 and symptoms of lower motor neuron disease in the upper limbs without significant functional involvement of other regions upon clinical presentation. This subgroup of patients is clinically characterised by the display of progressive atrophy and weakness in the arms with little effect on the bulbar muscles or legs. Atrophy and loss of strength affect the upper limb muscles in a more or less symmetric manner, prevalent in the proximal muscles. The comparative study with the rest of the amyotrophic lateral sclerosis group supplies very interesting details for the physician, such as a clear predominance among men, and a longer median survival. They conclude by suggesting that this syndrome could be a new variant of amyotrophic lateral sclerosis.

Finally, the authors carry out a historical review, and refer to the fact that this distinctive amyotrophic lateral sclerosis variant was probably first described by Gowers in 1886, known in Franco-German literature as Vulpian-Bernhardt’s form.

In his book Maladies du Système Nerveux Vulpian described a patient who showed signs of weakness and symmetric proximal atrophy of neurogenic origin, and called it chronic anterior poliomyelitis. The patient showed symptoms of proximal amyotrophy, and signs of denervation and upper motor neuron involvement. Since then, in those countries and other countries under their influence, 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 critical onset.

A certain enigma exists regarding 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 infraspinatus, the supraspinatus, the sternocleidomastoideus, and the teres minor. The predominant involvement of these muscles 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 Arman-Duchenne hand.

Obviously, signs of corticospinal involvement with hypereflexia in the lower limbs and Babinski’s sign both appear. In the initial stages of the illness, there is no effect on the diaphragm. The presence of signs of involvement of the upper motor neuron, its different clinical evolution, and the data supplied by genetic molecular investigation allow us to distinguish the syndrome previously known as Vulpian-Bernhardt’s rebaptised as flail arm syndrome from other motor neuron syndromes such as of the spinial muscular atrophies, Kennedy’s disease, multifocal motor neuropathy, and monomelic amyotrophy.

JOSEP GAMEZ
CARLOS CERVERA
AGUSTIN CODINA
Servicio de Neurología, Hospital Gral Universitari Vall d’Hebron, Passeig Vall d’Hebron 119–135, 08035 Barcelona, Spain

Correspondence to: Dr Josep Gamez, Servicio de Neurologia, Hospital Gral Universitari Vall d’Hebron, Passeig Vall d’Hebron 119–135, 08035 Barcelona, Spain. Email: j2784@hsbc.com


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 disorder grades 1 and 2. This is the study’s greatest strength. The study has, however, its limitations.

The first consideration is that there were 98 accident victims who reported acute symptoms, and thus were at risk for the late whiplash syndrome. How does this compare with other studies documenting the natural evolution of the late whiplash syndrome? The Swiss study may be useful for comparison because it too has only 117 subjects, yet is much quoted. Setting aside for the moment that the Swiss study is hampered by the selection atrocity of advertising for subjects, and has a host of other reportedly fatal 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. Yet, in Switzerland, not even 20% manage to recover fully by 3 months and many of these were reporting total disability during that time, whereas the Lithuanians fully recover in 4 weeks or less, with little or no therapy and no treatment whatsoever. Studies in other western countries disclose an even greater contrast, with 50%–70% of patients reporting pain even after 3–6 months, despite the fact that all these studies are examining the same grades (1 and 2) of whiplash associated disorders. 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 culture in the rate of recovery from whiplash injury is demonstrated.

Thirdly, there are sex differences and even differences in seat belt usage between this population and some others, but even then, it does not determine whether 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.1, 2, 3, 4 If there were strict design criteria to identifying symptoms, the control population would have grossly underreported their symptoms. Indeed, chronic pain can and is reported by studies in many different cultures and languages, including Japan, France, Italy, and others. If researchers in these non-English speaking populations can use simple questionnaires to document the late whiplash syndrome so effectively there, then the same should be possible in Lithuania.

And so, despite the potential limitations of this study as outlined, there is no way to get around the stark realisation that the natural history of acute whiplash injury in Lithuania is a benign syndrome with 4 weeks or less of pain. Equally compelling is the fact that Lithuania is not the only place where researchers are having difficulty identifying epidemics of chronic pain. Recovery from acute whiplash injury without neurological injury or fracture routinely occurs within 4–6 weeks in Germany1 and Greece.1 The time has now 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 time in order to help prevent the chronic pain and the suffering we otherwise encounter.1, 2

R FERRARI


BOOK REVIEWS


This book purports itself to be a comprehensive reference. Certainly the title would suggest so. However, it is clear that this is not a comprehensive text, but a book that is an update on particular timely topics in the field of pain medicine. There are sections on pain mechanisms, a chapter on the pharmacology of acute and chronic pain, and other chapters on postoperative pain, obstetric pain, and acute paediatric pain. There are three further chapters specifically on the management of chronic low back pain, cancer pain, and an overview of interventional pain techniques.

Many of the authors are internationally known and this reinforces 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 outstandingly useful technique of spinal cord stimulation, and disc denervation. Again this chapter has been written by an internationally well known author who concentrated on general overview of the techniques rather than a how to do it approach, which I think one would expect from a chapter of this size.

The chapter on 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 cryoneuromodulation. Again this chapter has been written by an internationally well known author who concentrated on general overview of the techniques rather than a how to do it approach, which I think one would expect from a chapter of this size.

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 neuropyschologist with a longstanding interest in the dementia, 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 multic Contributor textbooks.

The title of the book is perhaps a little misleading in that the book includes, as well as traditional neuropathology, a very comprehensive overview of the molecular biology and genetics of the dementias. As would be expected, a considerable proportion of the book is dedicated to Alzheimer’s disease with chapters on both the clinical features, genetics, and the neuropathology. The frontotemporal dementias are also well covered and the book includes a chapter on the new developments 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, one of the chapters 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


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 textbook of over 500 pages. The field of peripheral nerve surgery is covered comprehensively, commencing with descriptions of anatomy, physiology, and pathological reaction to injury. This is followed in subsequent chapters with descriptions of approaches to virtually all the main peripheral nerves, and the operative management of brachial plexus injury and outcomes is covered in three detailed chapters. These are followed by chapters on nerve entrapment, neuropathy, iatropathic injury, and neoplasm within the peripheral nerve. The final section covers electrodiagnosis, pain, nerve recovery, reconstruction techniques, and rehabilitation.

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

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

M HADJIVASSILIOU, R A GRÜNEWALD and G A B DAVIES-JONES

*J Neurol Neurosurg Psychiatry* 1999 67: 257
doi: 10.1136/jnnp.67.2.257