Continuous intrathecal baclofen infusion (CIBI) is a widely accepted therapy for the treatment of severe spinal spasticity. There is increasing evidence that CIBI has similar effects on patients with supraspinal spasticity resulting from either hypoxic or traumatic brain injury. A proportion of patients with supraspinal spasticity present with additional autonomic dysfunction. These dysfunctions are often unresponsive to conventional antidiuretic medication. Patients may present with severe arterial hypertension, tachycardia, hyperhidrosis, hypersalivation, and bronchial hypersercretion. Mortality is mainly influenced by these symptoms. In addition to antidiuretic medication sedatives and analesics are often required. During intensive care treatment such medication often leads to a prolongation of artificial ventilation and delayed rehabilitation. During rehabilitation autonomic instability additionally interferes with physotherapeutic activity.

In the follow up of patients with severe supraspinal spasticity we noted the positive influence of intrathecal baclofen infusion on autonomic instability. Study of the literature did not disclose similar findings. So far, it had not yet been pointed out that CIBI has a positive influence on symptoms originating from severe hypoxic or traumatic brain damage.

Eighteen patients with severe tetraparesis from either hypoxic or traumatic brain injury were treated with CIBI. Before admission all patients were treated with maximum doses of various oral antispastic agents.

The interval between event and bolus test varied between 1 and 62 months (median 8 months).

After a positive response to an intrathecal baclofen bolus application a pump (Medtronic Synchromed 8611H) was implanted for CIBI.

During the further treatment of these patients it was noted that six of 18 patients had severe autonomic dysfunction, unresponsive to conservative medical treatment (30% of the original communication). Tachycardia, arterial hypertension or hypotension, often associated with agitation, hyperhidrosis, hypersalivation, and tracheobronchial hypersercretion were regarded as autonomic dysfunction associated with severe spasticity. These symptoms were graded qualitatively as not present, present and improved, or present and unchanged. This assessment was made from physicians’ and nurses’ documentation of symptoms in the patient files and from the observations of relatives. Due to the retrospective character of the study, a further quantitative assessment could not be performed.

Six out of the 18 patients had severe autonomic dysfunction and spasticity. Three patients had hypoxic and three severe traumatic brain injury. Two patients were severely disabled and four were in a vegetative state.

The interval from primary event to pump implantation ranged from 4 to 24 months.

The mean follow up was 16.8 (8–23 months) and the mean daily dose of intrathecal baclofen was 408 µg (range 100–600 µg).

Four patients presented with severe spasticity (Ashworth score 5); one with an initial Ashworth score of 4 and one with an Ashworth score of 3. The following autonomic dysregulation symptoms were seen: tachycardia (3/6), arterial hyper-hypotension (2/6), hyperhidrosis (5/6), hypersalivation (5/6), tracheobronchial hypersercretion (5/6). Agitation was seen in five of six patients. In five patients the symptoms disappeared with intrathecal baclofen therapy. In one patient (No 3), a 33 year old woman, in a persistent vegetative state after hypoxic brain injury originating from non-suicidal strangulation, the autonomic dysfunctions did not improve with intrathecal baclofen but disappeared after the additional application of intrathecal morphine. The outcome concerning the autonomic dysfunctions is shown in the table.

Patients surviving severe traumatic or hypoxic brain injury regularly have a variable period of deep coma with flexion and extension spasms and severe autonomic dysfunction that might persist over months. Later, many patients die from autonomic dysfunction or severe infections. Most of the survivors develop severe spasticity and still have autonomic dysfunction. The effects of CIBI on autonomic dysfunction have not yet been examined in the literature, although these symptoms are present in about 30% of patients with supraspinal spasticity in our series.

The patients presented had severe autonomic dysfunction and were unresponsive to conservative medical treatment. Intrathecal baclofen infusion improved most of the symptoms so that no additional medication was necessary. One patient did not improve until additional intrathecal morphine was applied. The improvement of symptoms after intrathecal morphine administration might suggest a central analgesic effect of baclofen enabling a reduction of autonomic dysfunction. Spinal inhibition of excitatory transmission or severe infections. Most of the survivors develop severe spasticity and still have autonomic dysfunction. The effects of CIBI on autonomic dysfunction have not yet been examined in the literature, although these symptoms are present in about 30% of patients with supraspinal spasticity in our series.

The pharmacological background of these findings needs further investigation. Our preliminary findings should be confirmed in pharmacological testing series.

3 Amano M, Kubo T. Involvement of both GABAergic systems, especially in pathological conditions, is unknown. Tachycardia, arterial hypertension or hypotension, often associated with agitation, hyperhidrosis, hypersalivation, and tracheobronchial hypersercretion were regarded as autonomic dysfunction associated with severe spasticity. These symptoms were graded qualitatively as not present, present and improved, or present and unchanged. This assessment was made from physicians’ and nurses’ documentation of symptoms in the patient files and from the observations of relatives. Due to the retrospective character of the study, a further quantitative assessment could not be performed.

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muscles. The EEG records show a slowing of background activity and bursts of spike and wave which are generalised, multifocal, and often sensitive to intermittent luminous stimulation. The disease provokes progressive deterioration leading to cortical and subcortical dementia and eventual death.

The cases in the literature to date, including the families which we studied, all possessed these clinical criteria. The aim of this letter is to highlight one of our latest cases of late onset Lafora’s disease, which occurred when the patient was 25 years old and who, although not showing evidence of either tonic-clonic or focal seizures, did, however, have dementia and erratic myoclonus over a long period. Blood taken from this patient served in the identification of the Lafora chromosome map.

The patient was a 40 year old woman who, at 25, after giving birth to a healthy baby girl, began to show symptoms of depression and notions of persecution allied to paranoid psychosis.

At the age of 28 she came into our care with a dystonic reaction to neuroleptic treatment which affected the mouth, neck, and trunk but which cleared up when treatment was suspended. Her family informed us that the patient’s condition had not improved over the previous 2 years and in addition to having memory loss she had become unable to perform normal household tasks.

The patient showed general disinterest and irritability, she was poorly oriented regarding time and space, but was, however, capable of reading, writing, and making small calculations; on the other hand she was unable to interpret a newspaper correctly or remember times and dates, including the care of her child. A Benton and Endo test was done which showed temporal disorientation, short term memory loss, digital agnosia, ideomotor apraxia, and difficulty with reading and writing, expressive language, and logical argument. The patient could not distinguish right from left. The EEG record showed background theta activity as well as slow and sharp waves with a paroxysmal tendency. Intermittent photic stimulation did not induce further paroxysmal discharges. During subsequent years the patient deteriorated mentally without any major epileptic seizures. The EEG record showed background theta activity and presents multifocal paroxysmal discharges. 

During subsequent years the patient deteriorated mentally without any major epileptic seizure and only slight myoclonus. Use of valproic acid and clonazepam was suspended. An auxiliary biopsy of sweat glands ruled out the presence of Lafora inclusion bodies, although a right frontal cerebral biopsy did show their presence (figure).

The patient is currently 40 years old and has obvious dementia. She moves from bed to armchair in a position of tetraparesis of limbs with pseudobulbar signs, she displays generalised rigidity with cogwheeling phenomenon and amnesia, her deep reflexes are brisk, she has only slight erratic myoclonus but no other seizures. The EEG register is slow in background activity and presents multifocal paroxysmal discharges of spike and spike waves. In the literature about Lafora’s disease, there have been few reported cases with late onset.

The peculiarity of our patient is that after having given birth to a child at the age of 25 she was shown to have the disease in a late onset form.

Initially she had psychiatric disturbances leading to progressive dementia with myoclonus. The patient at no point had generalised tonic-clonic or focal seizures.

Treatment with valproic acid and clonazepam was initiated as a precaution, but as the illness progressed and the patient deteriorated and still no major attacks happened, use of both was suspended. The EEGs recorded during evolution were typical with generalised paroxysmal discharges and focal or multifocal and slow background activity. The only clinical seizure that the patient presented was provoked shortly after admission and consisted of massive myoclonus with no loss of consciousness. This was caused by photostimulation in the EEG laboratory. What interested us about the patient’s evolution was the evidence of dementia rather than epilepsy.

An auxiliary biopsy proved negative, but the cerebral cortex showed inclusion bodies characteristic of Lafora’s disease, most of which were intraneuronal.

Despite the late onset of the illness in this case, studies of blood samples from the patient showed the chromosomal map of Lafora illness.

A patient with dementia and Lafora inclusion bodies but without epilepsy has only previously been described by Suzuki et al. This was a patient aged 59 with dementia who showed the histological findings typical of the illness. Our patient, who has now had the disease for 15 years, continues to have dementia and shows no evidence of tonic-clonic or focal seizures. Only slight myoclonus is present.

We think that Lafora’s disease could be more common than previously thought and other forms of the illness should be sought, such as late onset dementia without epilepsy, accompanied or not by slight myoclonus. We also think that an auxiliary cerebral or other tissue biopsy, or a genetic study can show different clinical forms of the disease.

We are grateful to Professor Barry Noonan for help in revising the text.

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Light microscopy of biopsy of the frontal cortex. There are characteristic intraneuronal inclusion bodies of different morphological forms with Nissl granules. Periodic acid Schiff originallyx1000.
Hyperlipidaemia (%) 9 (53) 23 (48) NS  
Hypertension (%) 6 (35) 24 (50) NS  
Mean age (range) (y) 61 (35–81) 61 (40–80) NS  
Smoking (%) 8 (47) 31 (64) NS  
Atrial fibrillation (%) 2 (12) 2 (4) NS  
Between the two groups by Fisher’s exact test.

Abundance of left hemispheric embolic strokes complicating coronary angiography and PTCA

Stroke is a well known complication of coronary catheterisation but there are only few reports on the incidence of stroke after coronary angiography (CoAng) and angioplasty (PTCA), and the unique characteristics of this complication are not well documented. We therefore examined stroke rate as well as minor health changes in our institution, and whether certain stroke patterns are more common than others.

We retrospectively examined medical records of all patients that were coded in the computerised hospital database in an intensive care unit of discharges of both stroke and CoAng or PTCA between the years 1987 and 1994. Only patients that had a stroke within 24 hours after the cardiac procedure were included. All patients were evaluated by a neurologist. The neurological deficit was classified as mild (for example, mild hemiparesis or dysphasia) or severe (for example, severe disability or aphasia). Patients with non-localising neurological symptoms such as vertigo were not included in the study. Brain CT was performed in all patients, as soon as the haemodynamic condition allowed it, for identification of haemorrhagic strokes. We further classified ischaemic events as embolic in patients with hyperacute presenta- tion of stroke and maximal neurological def-icit at onset. We then compared the stroke group with a control group for the frequency of cerebrovascular disease risk factors, for the indication for the cardiac procedure, and for the degree of coronary disease that was subsequently found. The control group consis- ted of 48 randomly selected patients who underwent CoAng or PTCA without develop- ing stroke. All catheterised patients rou- tinely received antiplatelet therapy before, and heparin during the cardiac procedure. The extent of coronary disease was graded as mild (single vessel disease), moderate (double vessel disease), or severe (triple vessel dis- ease). A χ2 analysis and Fisher’s exact test were used for statistical evaluation.

A total of 7151 patients underwent cardiac procedures (3460 had PTCA and 3691 had CoAng) during the study period. Seventeen patients had strokes that fulfilled the entry criteria, accounting for an incidence of 0.23%. Sixteen patients had an ischaemic stroke and one had a haemorrhage. The major- ity of the ischaemic strokes were severe (13/16, 81.5%) and occurred during the card- iac procedure itself (12/16, 75%). All of the strokes evolved hyperacutely, with maximal deficit at onset, including those that appeared several hours after the catheterisation. Brain CT obtained in the acute phase, showed acute infarcts in the distribution of the middle cerebral artery in five patients and was interpreted as normal in all other patients. The severity, distribution, and hyperacute presentation strongly suggest that the neuro- logical events were embolic in nature.

Stroke locations were distributed unevenly between the major vascular territories. Thir- teen patients (81.5%) had a left hemispheric stroke, whereas only one patient (6%) had a minor right hemispheric event (p<0.0001, χ2), and two patients (12.5%) had a vertebro- basilar event (p=0.0002, χ2). There were no differences in patient distribution and one patient was left with severe disability. All three deaths occurred during the initial hospital stay and were related to the acute stroke.

Table 1 shows that the group of stroke patients did not differ from the control group for risk factors for cerebrovascular disease and severity of coronary disease. However, the indication for the cardiac procedure in most stroke patients was urgent (75% with unstable angina pectoris or acute myocardial infarction), whereas it was non-urgent in the majority of controls (63% with stable angina pectoris or congestive heart failure). This dif- ference was significant (p=0.001, χ2).

The stroke rate of 0.23% after cardiac proce- dures found in our study is similar to other reports. Identification of patients who are prone to have a stroke after cardiac procedures for its development might enable us to lower the risk of this serious complication. Brown and Topol1 reported that stroke patients had more risk factors for cerebrovascular disease than controls. Alternatively our study suggests that performance of cardiac proce- dures on an urgent basis due to acute activity of the heart disease may increase the risk of cerebrovascular complication. This may be explained by the presence of a generalised hypercoaguable state at the time of the procedure.

The clinical features of most stroke pa- tients in our series are typical for an embolic event. Our finding that these emboli involve preferentially the left hemisphere, may pro- vide a clue as to their source and mechanism of dislodgement. We cannot rule out com- pletely a selection bias in this retrospective study as minor right hemispheric events may escape diagnosis more often than left hemi- spheric ones. However, such a bias does not hold for major neurological events. In addi- tion, all patients were subject to close monitoring in an intensive care unit. A consulting neurologist was routinely called in all cases of suspected neurological deficit, including minor ones, thus minimising a selection bias. Therefore, we think that the relative abundance of major left hemispheric strokes is genuine.

Uneven distribution of stroke location does not support a cardiac origin of emboli. Alternatively, brain emboli could arise in these patients from the aortic arch or carotid arteries. Such emboli could preferentially reach the left hemisphere due to the vascular anatomy of the aortic arch region. We suggest that manipulation of the catheter tip at the dis- tal bend of the aortic arch, which is closer to the origin of the left common carotid artery, may cause dislodgement of atheromatous mate- rial preferentially into that artery. It was previ- ously suggested that scraping the ascending aorta while searching for the ostia of coronary arteries was the cause of stroke. However, Tunick et al showed that most protruding aor- tic atheromas are located distal to the innomi- nate artery, and are therefore not prone for left hemispheric strokes in most patients. This is in good agreement with our proposed mechan- ism for embolism in CoAng/PTCA patients. Indeed, one of our more recent patients underwent transoesophageal echocardiography after the stroke, and an atheromatous plaque was found in the aortic arch near the origin of the left common carotid artery.

In two studies in which cardiac catheterisa- tions were performed by passing the catheter at a right antecubital or a brachial site, an increased incidence of vertebrobasilar strokes was found. In these cases the catheter tip may have released atheromatous material while advancing in this strong artery that subclavian artery, preferentially into the vertebral system, spar- ing the carotid arteries. In our study, the femoral artery was used for catheter insertion in all stroke patients except for one. The one stroke patient in whom the catheter was inserted via the radial artery had a vertebro- basilar event.

In conclusion, we suggest that cardiac catheterisation still carries a measurable risk for stroke, especially in patients with acute

<table>
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<th>Stroke</th>
<th>Control</th>
<th>p Value</th>
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<tr>
<td>Number</td>
<td>16</td>
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</tr>
<tr>
<td>Male (%)</td>
<td>10 (62)</td>
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</tr>
<tr>
<td>Mean age (range) (y)</td>
<td>61 (35–81)</td>
<td>61 (40–80)</td>
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<tr>
<td>Hypertension (%)</td>
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<td>Hypertension (%)</td>
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<td>Smoking (%)</td>
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<tr>
<td>Atrial fibrillation (%)</td>
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<td>CoAng/PTCA (%)</td>
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<td>24/24</td>
</tr>
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<tr>
<td>Unstable AP</td>
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<tr>
<td>Acute MI</td>
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<tr>
<td>Non-urgent</td>
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<td>Severe AP</td>
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<tr>
<td>CHF</td>
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<tr>
<td>CoAng/PTCA findings: Severe CAD</td>
<td>4 (25)</td>
<td>20 (42)</td>
</tr>
<tr>
<td>Moderate CAD</td>
<td>9 (56)</td>
<td>16 (33)</td>
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<tr>
<td>Mild CAD</td>
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</tr>
<tr>
<td>no CAD</td>
<td>1 (6)</td>
<td>3 (6)</td>
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</table>

AP=Angina pectoris; M=myocardial infarction; CHF=congestive heart failure; CAD=coronary artery disease.

*The number of procedures performed on an urgent versus non-urgent basis was compared between the stroke and control groups by Fisher’s exact test.
†The number of patients with moderate-severe coronary artery disease versus no-mild disease was compared between the two groups by Fisher’s exact test.
coronary disease. We also suggest that stroke location depends on the route of catheterisation, with left hemispheric strokes being more common when the femoral artery is used for access. This finding calls for special care to be taken to avoid excessive catheter manipulation near the origin of the left common carotid artery.

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Cavernomas in the central nervous system and the relevance of multiple intracranial lesions in the familial form of this disease

The availability of MRI has greatly increased the detection of cavernous malformations of the CNS in both symptomatic and asymptomatic patients. These lesions may be responsible for previously unexplained neurological events or may even have been incorrectly diagnosed. Cavernomas have a characteristic MRI appearance consisting of an area of mixed signal intensity, thought to be due to extracellular methaemoglobin, surrounded by an area of reduced signal intensity reflecting a zone of haemosiderin. Missing an angiographically occult vascular malformation on MRI seems likely only if the lesion contains no haemoglobin breakdown products or is microscopically so small as to be unidentifiable. This may explain the sudden appearance or “growth” of cavernomas occasionally described.

We report on a family, spanning three generations, in which at least 10 members are affected. The original reference patient was a woman aged 34. She developed a sudden weakness of the left leg subsequently found on MRI to be due to a rare intramedullary cavernoma at C3. Two intracranial cavernomas were also found during the same study. All three were subsequently removed. It has been suggested that in patients in whom multiple lesions are found a familial link is more likely. We therefore took a detailed family history. The patient told us of an aunt who has epilepsy. She had recently been diagnosed by CT as having a low grade glioma. Subsequent MRI studies have shown the lesion to have the characteristics of a cavernoma. As the family tree (figure) was constructed it became apparent that five first cousins—four males (two of whom are monozygotic twins) and one female, all siblings—had presented independently to different consultants at our institution with either seizures or unexplained intracerebral haemorrhages. Brain MRI studies in the males had shown multiple intracerebral cavernomas. Their sister has two epileptic children, both shown to have cerebral cavernomas. She was symptom free and declined investigation, until the development of persistent headaches. Brain MRI has now shown intracranial cavernomas. When the medical history of the siblings’ parents was reviewed, their father admitted to a sudden spontaneous resolving hemiparesis when aged 20. He was noted to have the cutaneous angiomas sometimes associated with this condition. He was anxious to undergo investigation. Brain MRI has disclosed multiple cavernomas in the brain.

The familial occurrence of cavernomas has been reported previously, notably in Mexican-American families. As in the family we report, it takes the form of multiple intracranial lesions. The inheritance would seem to be autosomal dominant with strong penetrance. Recently the gene implicated has been mapped to the 7q locus. The finding of more than one cavernoma in one person should alert investigators to the possibility that other family members may be affected.

The surgical treatment of CNS cavernomas remains controversial. It seems that most cavernomas show evidence of previous haemorrhage to varying degrees. Most surgeons would agree on surgery in a symptomatic patient with a readily accessible lesion. However, the argument for surgery can also be made with lesions producing repetitive or progressive symptoms where there is significant neurological disability. Unfortunately, to date predictors of timing and size of haemorrhage are unclear. However in a recent study involving 145 patients the authors suggest that risk factors for “aggressive behaviour” include pregnancy, familial or multiple form, previous wholemale, brain or stereotactic radiotherapy, incomplete removal, associated venous malformation, and female sex. A conservative approach is best adopted when a clinically silent lesion in an eloquent area is discovered incidentally or in the case of multiple clinically silent lesions. In cases of epilepsy, well controlled on drugs, many would adopt a conservative approach. Further controversy surrounds management strategies after a single bleed in a vital area such as the brain stem.

Options here include MR directed stereotactic radiosurgery or direct surgery, which has been achieved with acceptable morbidity.

Review of the literature has shown this to be the largest number of affected members in a single family

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Molecular genetic diagnosis of Friedreich’s ataxia in a pedigree with apparent autosomal dominant spinocerebellar degeneration

Friedreich’s ataxia is a progressive neurodegenerative disorder of autosomal recessive inheritance, in which gait ataxia followed by upper limb ataxia, dysarthria, nystagmus, areflexia, loss of joint position sense, and spastic paraparesis develop from the second decade of life. It is the commonest hereditary ataxia, with a prevalence of 1 in 50 000 and a deduced carrier frequency in European populations of 1 in 120. Recently, Friedreich’s ataxia has been associated with mutations of the frataxin gene on chromosome 9.
Dysarthria has been present for four years. She is wheelchair bound. She has taken valproate for presumed complex partial seizures since the age of 8. Examination disclosed limb incoordination, worse on the left. Lower limb reflexes were hyperactive, especially on the left, and both plantar responses were flexor. He has marked truncal ataxia. Brain MRI was normal.

The proband's sister (IV.2) was seen at the age of 8 months after becoming floppy, unable to sit, or control her head acutely. She was intermittently irritable and sleepy, and had vomited repeatedly. She was admitted to hospital, where she was afebrile and well perfused. She could not sit unsupported, was unable to reach out for toys, and was hypotonic. She had gazedependent very fast vertical nystagmus with no failure of upgaze. Ultrasound examination of the head, blood gases, blood and urinary amino acids, liver function tests, and blood ammonia were normal. At review 6 months later, a further episode of acute ataxia in the context of a febrile infection was reported. This had settled within a few hours, and she had not been admitted to hospital. Since then she has been well (currently 3.5 years) with normal development.

Polymerase chain reaction for the GAA triplet repeat in Friedreich's ataxia was carried out using the primers and method described by Campuzano et al. 1 III.1 showed two expanded alleles carrying 320 and 840 GAA repeats. III.4 and IV.2 were heterozygous for the GAA expansion, the expanded allele bearing a repeat of 840 triplets consistent with their carrier status. II.5 was also heterozygous for the expansion, and III.2 had no expanded alleles. The presentation of this family with ataxic features in three generations had suggested a form of autosomal dominant cerebellar ataxia. However, the proband and his sister have clinical phenotypes consistent with Friedreich's ataxia, and this diagnosis has been established by genome analysis. The carrier status of the two other members of this pedigree manifesting ataxic features has been confirmed. This finding raises the possibility that Friedreich's ataxia carriers are at risk of developing ataxia, especially in the context of environmental insults (such as alcohol in II.4 and and viral infections in IV.2). In a recent series of 56 pedigrees, at least two heterozygous parents (both fathers) manifesting ataxic features were identified. 3

No data are available on possible environmental insults in these members. The current finding lends support to the conclusion of Lamont et al. that a history of ataxia preceding (or successive) generations should not preclude a diagnosis of Friedreich's ataxia. Finally, it may be fruitful to investigate those who develop spinocerebellar ataxia secondary to recognised environmental insults for their carrier status of Friedreich's ataxia.

We are grateful to Professor Tetsuo Ashizawa, Dr Pragna Patel, and Dr Sanjay Bidichandani at the Department of Neurology, Baylor College of Medicine, Houston, Texas for assistance with genome analysis and helpful discussion.


Intracerebral haemorrhage due to possible venous obstruction in the neck

Multiple concurrent cerebral haemorrhages in the absence of trauma or a bleeding diathesis suggest venous sinus thrombosis, multiple haemorrhagic infarcts, and haemorrhagic metastases. Iatrogenic venous obstruction is another possible cause. Patient 1 was a 55 year old obese woman who underwent a left posterior fossa craniotomy (Jannetta procedure) for trigeminal neuralgia. Examination and a CT of the head were normal. At surgery she was positioned with her head held in slight lateral flexion to place the left occipital area uppermost. During the operation the anaesthetist reported that on two occasions the pulse rate slowed only to return to normal when the retractor was immediately removed. The patient's head was repositioned to provide increased lateral flexion and because of her short neck and broad shoulders, slight Trendelenburg tilt was applied. A large artery was found indenting...
the left fifth nerve. On repositioning the retractor, swelling of the left hemisphere was noted which bulged though the craniectomy. A small cerebellar resection was performed and the head repositioned with upward tilt. The operation was then abandoned.

Postoperatively she failed to regain consciousness. Brain CT showed moderate swelling of the left cerebellum and both occipital lobes appeared hypodense. There were also high density haemorrhagic lesions in the left temporal, right fronto-occipital (30x20x20 mm), right parietal (15x10x10 mm), right temporal (20x20x10 mm), right occipital (25x10x20 mm) and left posterior (30x50x40 mm) lobes. The overlying subcutaneous space and sulci in the affected areas were widely distended with blood (sulcal haematomas) and adjacent cerebral cortex and underlying subcuticular white matter were disrupted by confluent small haemorrhages. The diagnosis of haemorrhagic infarction of venous type. Microscopical assessment of the haemorrhagic lesions disclosed venous and capillary congestion and coalescence of multiple microhaemorrhages typical of venous obstruction. The left cerebellum was lacereated and swollen.

In 1978 Jannetta et al reviewed 825 cases of posterior fossa surgery. Five were complicated by supratentorial haemorrhage, and in four the cause remained elusive. Two came to postmortem examination disclosing haemorrhage without evidence of underlying neoplasia or vascular malformation but it was not clear whether they were arterial or venous. It was considered that patient position (modified sitting) may have been in some way implicated and subsequent procedures were done in the lateral decubitus position. In 1988 Hanakita and Kondo reviewed 278 patients with head injuries and microvascular decompression all in the lateral decubitus position. There were two fatal intracerebral haemorrhages but there was no postmortem. Excessive cerebellar laceration and a disturbance of venous return with an increase in blood pressure was assumed to be the cause in one, but no explanation was given for the second.

The haemorrhages which occurred in our case were simultaneous and widespread and pathologically were venous yet the venous sinuses were patent macroscopically.

We postulate then that the venous haemorrhages were due to acute intraoperative venous compression secondary to neck positioning, possibly contributed to by the short thick neck. It is not the lateral decubitus position versus the sitting position which so often are cited as important but the neck positioning.

In support of the hypothesis of mechanical venous obstruction, Goody and Stimac have demonstrated in animal and infant cadaver models that turning the head to one side causes both torsion and compression of the ipsilateral jugular vein. They postulate that jugular venous occlusion on the side of the dominant venous drainage can result in severely limited cerebral venous drainage. Emerson et al, and Parker showed that with obstruction there was a marked reduction in cerebral blood flow with an increase in cerebral venous pressure to 25 mm Hg. Doppler studies have shown that cranial venous drainage is often asymmetric, being more dominant on the right and with unilateral venous occlusion, efferent contralateral shunting of blood is only possible to the dominant side. It was the right side of the neck which was compressed at surgery in our case.

Case 2 was a 40 year old woman with cryptogenic cirrhosis who underwent orthotopic liver transplantation. On admission for transplantation a central venous line was inserted into the right internal jugular vein. A Swan-Ganz catheter was inserted into the left internal jugular vein intraoperatively. Postoperatively transient thrombocytopenia was noted, the platelet count falling to 68 000 rising to 100 000 by day 2 and 335 000 by day 9. On day 4 the Swan-Ganz catheter was removed and on day 7 the central venous line was taken out. In addition to cirrhosis of uncertain aetiology the liver showed multifocal well differentiated hepatocellular carcinoma.

On the ninth post-operative day she complained of dizziness, blurred vision, and headache and the next day she was transiently dysphasic. Brain CT disclosed multiple superficial and deep haemorrhages in both hemispheres (fig 1). There were no cutaneous or systemic bleeding manifestations at this or any other time. Brain MRI confirmed the haemorrhagic lesions and disclosed patent venous sinuses. A subsequent ultrasound of the jugular veins 4 days later showed circumferential thrombus in both the right and left internal jugular veins with some venous flow. A repeat CT 1 month later was normal (fig 2). The patient remains well.

Neurological complications after liver transplantation include cerebral haemorrhage but we could find no case report of multiple perioperative intracranial haemorrhages without a significant bleeding diathesis.

Radiologically there were multiple haemorrhagic lesions without venous sinus thrombosis. Clinically there was no coagulopathy, no vasculitis, or evidence of haemorrhagic metastases. The complete resolution of the haemorrhages without evidence of encephalomalacia and the multifocal haemorrhages not conforming to any particular arterial territory supported the hypothesis that they were venous.

We therefore postulate that jugular venous catheterisation induced jugular venous thrombosis and venous hypertension with secondary multiple haemorrhages. There is one case reported of an infant aged 2 months who died of multiple cerebral haemorrhages thought to be produced by bilateral internal jugular vein thromboses caused by jugular vein catheters.

The concept of venous hypertension causing cerebral haemorrhage is not generally recognised. Neck positioning or traction and jugular vein catheterisation causing extracranial venous obstruction are two possible mechanisms with important management implications.

References:
Contribution of in vivo $^1$H spectroscopy to the diagnosis of deep-seated brain abscess

Brain abscesses are associated with high mortality and morbidity even in the antibiotic era and with complex imaging techniques, mainly because of delayed diagnosis. Several reasons can be identified: (1) the incidence of abscesses is low, about 1.1/100 000 person-year; (2) clinical features may be non-specific without evidence of infection and with neurological signs similar to tumours, which are 50 times more frequent; (3) neuroradiological imaging may be confusing specially in the early stages.

The only way to improve the outcome of this pathology is to consider an abscess when faced with a space occupying lesion and to rely only on clinical and radiological features; stereotactic aspiration is mandatory.

Recently, in vivo $^1$H MR spectroscopy (MRS) was reported to give metabolic information on brain abscesses very different from that of brain tumours, permitting a non-invasive differential diagnosis between these two diseases. We report a case in which the diagnosis of an abscess was made on the basis of in vivo $^1$H MR spectroscopy resulting in complete recovery.

A 50 year old woman with headache and fever was transferred to our institution. Brain CT was inconclusive and MRI showed a focal contrast enhancement near the right ventricle on T1 weighted images, after gadolinium injection. This region appeared heterogeneous on T2 and a diagnosis of glioblastoma was evoked (figure A). At the time of admission, the patient was confused and agitated. Neurological examination disclosed a left hemiparesis. There was no evidence of infection, with white blood cells at 9000/mm$^3$ and only a slight rise in C reactive protein, despite a temperature of 37.8°C. The family reported benign dental caries 2 weeks earlier. Intravenous methyl prednisolone resulted in clinical improvement and a stereotactic biopsy was planned after the performance of an in vivo $^1$H MR spectroscopy, applied for brain tumours at our institution. This procedure was performed on a Magnetom Vision 1.5T imager (Siemens, Erlangen, Germany) with the patient mildly sedated for 30 minutes. The volume of interest (VOI) was 20 mm×20 mm×20 mm. Two types of sequences were used: a STEAM sequence with TR 1500 ms and a short TE of 20 ms which enables many metabolites to be visualised including those with short T2 and a spin echo sequence with TR 1500 ms and a TE of 135 ms for lactate identification. Each sequence was taken from the lesion and the controlateral area. This spectrum was very unusual for a tumour, with large amounts of amino acids (0.9 ppm), lactate (1.35 ppm, inverted with TE 135 ms), and unexpected signals at 1.85 and 2.4 ppm attributed respectively to acetate and succinate. With respect to a $^1$H MRS spectrum of glioblastoma, N-acetyl-aspartate (NAA 2.0 ppm), creatine, and choline were present in smaller quantities (figure B). The clinical status of the patient was worsening rapidly, the possibility of an abscess was considered, and an emergency biopsy was performed. Pus (20 ml) was drawn off stereotactically and Gram stain with bacterial cultures isolated multiple organisms with an anaerobic streptococcus and haemophilus.

The pus was analysed with in vitro $^1$H NMR (figure C). The resonances were assigned by spiking samples with authentic standards. The main signals in the complex spectrum were those of lactate (1.33 ppm), alanine (1.48 ppm), acetate (1.92 ppm), succinate (2.41 ppm), and glycine (3.56 ppm), thus confirming the in vivo $^1$H NMR study. The status of the patient improved rapidly with a 3 week intravenous antibiotic therapy with cefotaxime and metronidazole, then ceftriaxone and metronidazole for another 6 weeks. At the end of this period, in vivo $^1$H MRS showed amino acids remaining but no lactate present, and the NAA signal rose. The steady increase of the choline and creatine peaks expressed a process of slow recovery. Six months later, the patient had totally recovered, MRI only showed a punctiform gadolinium uptake, and the general aspect of the in vivo $^1$H MR spectrum was nearly normal.

The clinical diagnosis of brain abscess is difficult because barely half the patients exhibit the four main groups of symptoms:

1. increased intracranial pressure;
2. focal neurological signs;
3. systemic indications of infection;
4. known extracranial infection.

Thus diagnosis cannot rely only on clinical and radiological features; stereotactic aspiration is mandatory.

In vivo $^1$H MRS can provide metabolic information on the development of a brain abscess. Actually, a metabolic degradation due to bacteria is responsible for the appearance of characteristic compounds such as acetate and succinate, end products of carbohydrate metabolism, and an amino acid peak linked with the reaction of polynuclear leucocytes against bacterial aggression. Lactate was also found but its bacterial origin was not certain. Moreover, a decrease of neuron density, energy metabolism failure, and phospholipid membrane breakdown were expressed respectively by loss of NAA, creatine, and choline. The brain tumour spectra are different as acetate, succinate, and amino acids have never been reported. High levels of succinate and acetate have been detected in the $^1$H NMR spectrum of aspirated abscess material.

After 3 weeks of intravenous antibiotic therapy, succinate, acetate, and lactate were absent, indicating a return to an aseptic condition. Nevertheless, aminoacids were still present. As they are only indirect consequences of bacterial development, they were not modified sooner by antibiotic treatment. The increases in NAA, choline, and creatine indicated a progressive return to a normal metabolism. Six months later, MRI and the brain spectrum were normal.
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CORRESPONDENCE

A triptan too far?

We read with interest the excellent editorial from Goadsby “A triptan too far?”1. As he points out, both neurologists and general practitioners are now faced daily with the problem of which triptan to choose. However, we think that it is difficult to choose between the different triptans and their dosages from the figures therapeutic gain and therapeutic penalty. The efficacy for 5 mg zolmitriptan, for example, is somewhat higher than for 100 mg sumatriptan, but this is acquired at the cost of more adverse events. To make a choice between the different options, it might be appropriate to relate the increased therapeutic gain with the increase in therapeutic penalty.

It is therefore interesting to compare the ratio of therapeutic gain/therapeutic penalty (rTG/TP) for the different triptans and their dosages. This direct comparison is allowed as the compounds all belong to the family of the triptans and therefore the characteristics of the adverse events are expected to be comparable.

As Goadsby suggests, the principles he outlines could be applied to the newer triptans when they become available. We propose the addition of a figure in which the therapeutic gain on the x axis is plotted against the therapeutic penalty on the y axis (figure). Here as well, it is possible to indicate a reference for the rTG/TP. Compounds which are situated below the reference line have a relatively favourable rTG/TP, whereas those situated above the reference line, rTG/TP is relatively unfavourable.

We have found a difference in the data mentioned in the text and the ones shown in figure 3.1. For sumatriptan 100 mg the therapeutic gain is 17% (95% CI 10%–24%) according to the text, but in figure 3 the penalty is shown as about 8% (95% CI 3%–11%). Because sumatriptan 100 mg is used as a reference, this difference is even more important. In our figure we have used the sumatriptan 100 mg data as mentioned in the text.

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Goadsby's editorial says many sensible things, which the first time it is a difficult question of how to meaningfully compare the outcomes of different treatments for migraine studied in separate clinical trials. However, in promoting the notion of “therapeutic gain” for between study comparisons he has not fully heeded the caveats he suggests for such an approach. I am aware of no compelling data supporting his statements that “the benefit of the active drug is added to that of placebo” or that “the active drug gain corrects for the effect of placebo.”

Consider the exceptional drug, meclozine, which is effective in almost all patients. In one study, in which the investigators are very excited about the drug’s prospects and are also excellent caregivers, the placebo response is 45%, whereas the meclozine response rate is 95%. In another study, performed at study centres where patients are mechanically processed with little enthusiasm, the placebo response is only 15%, but the meclozine response remains at 95%. In the second study the therapeutic gain is 80%, whereas in the first it is only 50%. Surely these two figures for therapeutic gain tell us more about the variability of the placebo response than about the response rates of meclozine.

In irritable bowel syndrome, which has many similarities to migraine, the placebo response rate in clinical trials varies from 0% to over 70%, due to differences in study design, patient and investigator expectations, treatment setting, patient population, and other factors. Subtracting out the placebo response in such studies does not necessarily help elucidate the true benefit of the active therapy. Indeed, in migraine there are similar examples. In zolmitriptan trials of the 5 mg dose, the 2 hour placebo rate has varied over threefold, from 15% to around 50% (Diener HG et al, IIIrd European Headache Federation Meeting, Sardinia, 1996). However, in those and other studies the zolmitriptan response rate remained remarkably constant, between 62%–67%.14 Rather than concluding that the therapeutic gain of zolmitriptan is variable, another interpretation would be that there is a ceiling effect for the drug of about 65%, and the placebo response is elastic, depending on various factors. This is not to say that the placebo response should be ignored in interpreting outcomes, but that it must not be mechanically “taken away” from the active treatment response in evaluating either absolute or comparative efficacy. Surely both the active treatment and placebo response rates must be considered as well as
the differences between them in fully evaluating migraine studies. And because of differences in the design and execution of such studies, it remains hazardous to draw comparative conclusions from independent studies, as Goadsby states.

I also suggest caution in the use of the concepts of “therapeutic penalty,” and the “number needed to harm,” as discussed by Goadsby. Such approaches treat all adverse events as equally important. However, the frequency of occurrence of relatively trivial events, such as mild dry mouth is of far less consequence than the infrequent occurrence of, say, myocardial ischaemia. It would not be appropriate to give them the same weight, as is done with both of these approaches.

For truly meaningful comparisons of both efficacy and safety, head to head comparative trials remain the definitive approach. Hopefully, such trials will be forthcoming to allow for truly valid comparisons among the triptans.

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Goadsby replies:
I thank van der Kuy and Lohman for their interest in the editorial1 and agree that treatment is crucial. We have all struggled with a construct that would absorb both the benefit and payment for headache relief and considered a gain/penalty ratio as suggested, being aware of the issues of measurement that surround the end points currently employed in clinical studies. I have tried to do something similar for number needed to treat and found, as the correspondents show, that it is not very sensitive, and there are issues with the confidence intervals. This is, in part, an indictment of the problems with current meta-analyses that matter indeed the field, represent a moving target. The data quoted in the text were from a single large study that was used in the figure were from a meta-analysis that included many more patients and perhaps is likely to be a more accurate representation of the differences (Tfelt-Hansen, personal communication). The problem is that the meta-analysis covers studies that are older and the newer study was more or less contemporaneous with the newer drug developments. One could argue for either data set and we all await with interest a comparative study between sumatriptan and zolmitriptan that has been completed but not yet formally reported.

In regard to Meijer’s comments, I would agree that the placebo provides an assessment of various things including the therapeutic effect, which I am not sure is different from the therapeutic gain, given that in some settings the treatment may be worse than the natural history of the condition. In any event the therapeutic gain is merely the reciprocal of the number needed to treat analysis which is a well developed concept.2 Certainly in the editorial the point was made that direct comparative studies to date (Barton et al3) however, by standard criteria the triptan developments are reasonably suited to this methodology.4

I also suggest caution in the use of the construct that would absorb both the placebo effects and the drug effects. Others have found that taking account of placebo responses is useful across a range of clinical disciplines, including neuropathic pain,5 epilepsy,6 and myocardial infarction,7 although not without criticism. The example of the zolmitriptan development programme, of which Klein was directly involved, is curious. The very large placebo response quoted in the editorial that has been published (Diener, H-C et al, 3rd European Headache Federation Meeting Sardinia, 1996), may have resulted from a very skewed randomisation of patients into the different treatments, so that there were very few patients entered in the placebo arm which was then a noisy data point with wide confidence intervals that contributed little to the meta-analysis. This outcome really makes the point for me that such overall analyses can be very useful when individual studies are problematic to interpret. In regard to making hazardious conclusions, I would suggest that ignoring the placebo effects offers a tool by which efficacy outcomes can be inflated for rather obvious purposes somewhat more hazardous than those employed in the editorial. Concerning side effects, similar comments may be applied. Calculating therapeutic penalty is nothing more than making an overall tolerability index. It does not consider safety and was not suggested by myself to do so. No clinician would weigh myocardial infarction equal with dry mouth and to suggest that this has been done is simply incorrect. Safety is a completely different issue and is considered by practitioners on another level. If there is any confusion about safety and tolerability is it in the mind of industry, not in the clinical side. I would agree that more head to head comparisons are necessary and stated this before commenting on the available triptans. In addition, I would like to correct the naratriptan logDpH7.4 which is ~0.2 not ~0.02, a typographical error pointed out to me by an alert neurologist colleague (Dr. Purdy, Halifax, Canada) to whom I am grateful. Triptans seem to have come of age in that we can have a mature, data driven debate about their use and misuse in clinical practice. Headache as a subspecialty is better for this, as is neurology in general. Patient care is well served by such discussions as we seek to do the best job for this very common clinical problem.

1 Goadsby PJ. A triptan too far? J Neurol Neurosurg Psychiatry 1999;64:143–7

1 Goadsby PJ. A Triptan too far? J Neurol Neurosurg Psychiatry 1999;64:143–7
4 McCrory HJ, Moore RA. Numerical results from systematic reviews in clinical prac- tice. 1998;104:1121-3

Line bisection in hemianopia
Barton and Black indicated in a recent report that hemianopic subjects without unilateral neglect may demonstrate a horizontal line bisection bias towards the scotomatous field.1 Their study was prompted by their impres- sion that the influence of visual field defects on spatial tasks has been little studied. The authors unfortunately seem not to have been aware of studies by Kerkhoff et al2 that not only replicated Barton and Black’s findings but indicate that such findings have been recorded as long ago as 1894. It would be useful if Barton and Black could indicate whether any aspects of their study are distinguished from the previous work.

1 Barton JJS, Black SE. Line bisection in hemi- anopia. J Neurol Neurosurg Psychiatry 1999;64:602–3
The authors reply: Mark is right in noting that Kerkhoff et al have mentioned this phenomenon of contralateral bisection bias in hemianopia. We will first examine the data provided by Kerkhoff et al, then peruse the literature he quotes, to answer Mark’s question.

Firstly, two of the three papers quoted by Mark do not provide data on hemianopic bisection bias. In one, the phenomenon is mentioned in a single sentence in the methods section; in the other, a German review contrasting hemianopia and hemineglect published last year, there is a short discussion about bisection without experimental data. The third paper studied bisection of vertical or horizontal lines on a series of six patients; however, all had bilateral cerebral damage and complex bilateral field defects, not hemianopia. Extrapolating from such unusual patients to those with more common unilateral hemianopia is not always appropriate.

Kerkhoff et al state that others have made this observation before.1,2 He quotes two works from the English literature. One is the paper by Schenkenberg et al.3 This report analyses the degree of brain damage without respect to hemianopia. Furthermore, hemianopic bias cannot be deduced from their tabulated data: there is bisection data for only six patients with hemifield defects without neurological or hemispheric lesions and the type of field defect is not stated. An informal test on the small sample of five patients with left hemispheric lesions shows no difference from the controls.

Another interpretation is a monograph by Teuber et al.4 This was also puzzling, because line bisection did not seem to be reported in the methods or results. We believe that Kerkhoff et al may have been referring to patients with lateral or vertical shift of fixation (“pseudofovea”, p 79) It does not necessarily follow that a shift of fixation leads to a shift in bisection judgments, though. Teuber et al do state that this might occur (p 82), and refer to another work of theirs,5 a case study of a man with a right occipital wound and left hemianopia. However, his bisection errors were biased rightward, as with neglect, rather than leftward, as we find with hemianopia.

Kerkhoff et al are more accurate in noting precedents in the older German literature. We became aware of these when we had a German monograph of Werth translated recently.6 It does seem to be the first to make this finding, in his report of a man with left hemianopia and leftward bisection errors. Apart from precedent, though, few conclusions are permitted from a single case study without controls.

The most important work we discovered is that by Liepmann and Kalms.7 These authors documented contralateral bisection bias in 10 patients with homonymous hemianopic lesions with intact hemifield vision. Interestingly, they noted that this bias was not present with very small or very large lines. They dismissed Axenberg’s hypothesis that the error was related to a coexistent gaze palsy but noted that it was related to increased numbers of saccades and fixations into the blind hemifield, an explanation which resembles the idea that the bias may be secondary to an adaptive shift of attention. (Note, though, that we and others have provided evidence that the spatial representation within normal hemifields can itself generate this type of bias.)8,9

We appreciate the opportunity to publicise the prior observations of Axenberg and Liepmann and Kalms,7 and the prescience of the latter’s speculations. Among our colleagues, “hemianopic bisection error” does not seem to be well known. We would also suggest that its scientific foundation in the German literature could be improved. We have provided better evidence in support of the existence of the bias and the conclusion that it is caused by hemianopia, with the use of statistical analysis, normal controls, and non-hemianopic brain-damaged patients which were used in the older German work. We hope that our brief initial report and this correspondence increase awareness of “hemianopic bisection error”.

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Myoclonus of peripheral origin

We read with interest the recently published article by Assal et al on “Post-traumatic stimulus suppressible myoclonus of peripheral origin.”1 We presented a patient with painful legs and moving toes associated with a peripheral sensory nerve lesion, and, although myokymia is often seen in peripheral nerve disorders, is abolished by local nerve blocks, and is thought to represent ectopically generated potentials in demyelinated nerve fibres. Myoclonic EMG pattern has also been described in PLMT due to hypertrophic mononeuropathy.2

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The authors reply: We thank Evidente and Cavinness for their interest in our paper. Their comments on the descriptive term we used to describe the abnormal movements of the second toe of our patient are well taken. We already partially considered this issue in our discussion. We do not agree that “myokymia” would adequately describe these movements for the following reasons. Clinically, there were no quivering or undulating movements of the skin overlying the muscle, and, although myokymia may sometimes lead to a movement of the fingers or toes, this movement is of very limited magnitude, in our experience not nearly as gross as that seen in our patient. Electrophysiologically, EMG discharges of myokymia discharge concern single motor units whereas many motor units fired in our patient. In some conditions myokymia may be related to the discharge of numerous single or grouped fasciculation potentials,1 but this activity does not manifest gross rhythmic muscle contractions either. Moreover, when myokymia have a peripheral origin, the ectopic activity originating from the motor nerve has been termed “contracting muscle”1 whereas we showed that this was not the case in our patient.

The particular condition in our patient was that the abnormal impulse generator seemed to be located on a sensory nerve (branch of
the deep peroneal nerve), distinct from the nerve (tibial nerve) supplying the contracting muscle. Therefore, a spinal cord relay had to accommodate, although neurologists in training, to manage the bigger movement and thus the treatment of young onset Parkinson's disease, the priming effect of levodopa in the development of dyskinesias and thus the treatment of young onset Parkinson's disease. Furthermore, the book rather overstates some of the trial data on various drug therapies which means that the best management of patients with Parkinson's disease can sometimes be hard to extract. Indeed the book is clearly written for the American physician, and so there’s essentially no mention of liuride, apomorphine, or cabergoline whereas pramipexole has a very high profile and selelgeline receives little adverse publicity. Furthermore, the classification of multisystem atrophy and the distinction between it and striatogniral degeneration, Shy-Drager and sporadic OPCA in the United States can be confusing. This emphasis obviously makes it harder to accommodate in the United Kingdom market, and the audience for which the book is attended, although neurologists in training, unable to afford the bigger movement disorder books, would probably find this book useful.

However, as an easy to read, rapid guide to Parkinson’s disease, this is a useful addition not least because of its length and cost and its attempt to deal with all aspects of the disorder equally.

ROGER BARKER


This book is much shorter than most that have recently appeared on sleep disorders and is all the more useful for this. The authors have been selective in the topics that they have covered and have emphasised the management of sleep disorders in primary care. Well recognised problems such as insomnia are included, and the book also deals with common but often overlooked conditions such as the restless legs syndrome and periodic limb movements in sleep. The chapter on pharmacological aspects of sleep deals mainly with hypnotic drugs and their use in insomnia and the place of stimulant drugs, especially amphetamines, in conditions causing excessive daytime sleepiness. One of the most original and most useful chapters is that which discusses sleep disorders in children. The overlap of medical, psychological, family, and social factors which are all important at this age are carefully dissected out. The book emphasises how patients present to medical attention, and how to assess the various symptoms of sleep disorders. The chapter on sleep disorders in preventive medicine is interesting and most of the chapters are enhanced by the inclusion of case studies. These are well chosen examples of clinical histories which exemplify how, and also how not, to manage sleep complaints. The tables, graphs and recordings from sleep monitoring records also make this book easier to read.

The authors cover each topic to a level which is sufficient for the non-specialist in sleep disorders, although some sections are rather superficial, and, as is almost inevitable in a multiauthor book, different chapters assume different levels of knowledge and understanding by the reader. The inclusion of a brief overview of the topic of some in the chapters would have been helpful. The discussion of excessive daytime sleepiness, for instance, concentrates on narcolepsy and gives little coverage of the many other disorders causing this symptom. Overall, however, this is a useful and comprehensive book which will be helpful to both medical and non-medical staff who are involved in handling sleep problems.

JOHN SHINEERSON


This book describes in detail the clinical features and latest research findings on congenital muscular dystrophy. The book follows on from an International Symposium on Congenital Muscular Dystrophy held in Tokyo in July 1994. The book has many positive features but also some less good features, so common in edited multiauthors texts.

On the positive side, the book gives a very comprehensive and authoritative review of the clinical features of the various types of congenital muscular dystrophies. I particularly enjoyed reading (and learning) about the different clinical phenotypes from clinicians who clearly have seen many cases. The fascinatory association of the muscular dystrophy with CNS abnormalities both clinical and radiological, are well described.

The chapters describing the morphological features in both muscle and brain are also comprehensive and helpful to those trying to establish a diagnosis. There is also an interesting chapter on the clinical management of patients, so important for those incurable diseases.

Where have the major advances in this area been. As with other muscular dystrophies, progress has been made by the finding of an absence of a structural gene, in this case merosin, in a subgroup of patients. The most common form of congenital muscular dystrophy in Japan, Fukuyarna congenital muscular dystrophy, has been mapped to a specific chromosomal location and a possible gene will be identified. Finally, on the positive side, there is a comprehensive bibliography which was updated to March 1997—helpful for those wishing to know more.

On the negative side is a feature I find concerning with many multiauthor books. Some chapters are repetitive, giving only a slightly different slant on the same problem. Also, towards the end of the book the chapters on pathogenesis I found less interesting than the chapters which tried to pull the clinical and pathological features together.

Who should buy this book? Well, it is a must for all paediatric neurologists who have an interest in neuromuscular disease, or aspiring to have an interest. For those with a general neurological interest it will be important to have access to a copy. It will also be helpful for adult neurologists, such as myself, who get a mental block at those clinical meetings where you are expected to remember the 85 varieties of floppiness. Be warned, I am now an expert and can confidently diagnose a case of Fukuyarna muscular dystrophy; shame that they are only seen in Japan!

DOUG TURBULL

It is not often that I buy a textbook but 10 years ago I bought the first edition of this book. Not only has it been a valuable source of reference but also a useful book to lend to those interested in a career in rehabilitation medicine. This second edition clearly delineates the broad field which now comprises the specialty of rehabilitation medicine. The comprehensive curriculum for trainees produced by the Royal College of Physicians puts this book at the top of the reading list.

An informal survey of six specialist registrars in rehabilitation medicine disclosed that all six had used this book at some time in their training. Enthusiasm for the book was greatest among trainees in their first two years of training with enthusiasm declining with increasing knowledge of the specialty, with the more experienced trainees preferring more specialised texts. Two of the six specialist registrars had already looked at the new edition and found it useful and the only criticism on content was the lack of information on sexuality in the disability which I agree is an omission.

My main criticism is that the presentation of the text seems old fashioned if it is compared with other newer text books such as those in rheumatology. I also noted in my copy at the end of the chapter on rarer neurological conditions that some of the references had obviously been missed out during the printing and an extra paper had had to be stuck in which rather spoilt the overall feel of the book.

Despite these minor criticisms I think this is an excellent book to be recommended to all trainees and those associated with the field of rehabilitation medicine. I would certainly have bought this addition too if I had not been kindly given this copy to review.

ROBIN STOTT


The authors state that this book is intended as a comprehensive, thorough, and practical textbook of psychiatry presented firmly within the medical model. It certainly is all of these things. It covers disorders which affect mental function from the cradle to the grave; it is thorough, producing a good amount of detail on each of the disorders, and practical, providing a brief but succinct treatment section for each disorder. The authors state that the book has its roots in the revolutionary changes that have occurred in psychiatry in the past 40 years highlighting the advances in psychopharmacology and the physiology of psychiatric and neurological conditions and that the book is designed to reflect this. It certainly does, having a very medical flavour, and in keeping with medical textbooks, is liberally illustrated by photographs of clinical signs of various disorders and relevant neuroimaging. Its overall layout is designed to complement the organisation of the diagnostic and statistical manual. Each topic is on the whole only a page or two long, written very succinctly and often almost in note form. In this way they are able to include a good deal of information. It is extremely well laid out and very attractive to the eye with a consistent organisation of each chapter throughout the text; tables are used where appropriate allowing for ease of access to the information.

The last part of the book contains a section on psychopharmacology with chapters both on groups of psychotropic agents and also commonly used or new single agents.

I enjoyed reading the book—it made me feel like a medical student again—where things can be easily compartmentalised, structured, and learnt and like all good text books, I found myself enjoying roaming around the chapters looking at unusual conditions not read about for a while! This book is a useful text for medical students and doctors training in psychiatry. It would also be an asset to those working in liaison psychiatry.

CAROL GREGORY


In recent years there has been a significant increase in the number of children who are investigated as potential candidates for epilepsy surgery, and in the number of centres which offer this service. Some of the reasons behind this are improved access to high resolution MRI and other techniques for the assessment of the origins of epilepsy, an increased level of awareness and expectation among parents, and acceptance by paediatricians and paediatric neurologists of the potential benefits of surgery in carefully selected cases.

Epilepsy surgery was developed to a large extent in adults. Traditionally it was offered to children only as a last resort, principally because of the understandable reluctance to remove or disconnect parts of the brain for anything other than life threatening pathology. The recent interest in childhood epilepsy has come about largely because of a better understanding of the natural history of epilepsy, declining surgical morbidity, and the potential which exists for improved cognitive and behavioural outcome from early control of seizures.

Against this background, Tuxhorn and colleagues have produced this book. Although generated as a result of the 6th International Bethel-Cleveland Epilepsy Symposium, this is not a book of the conference proceedings. Instead, many acknowledged experts in the field have combined to create a comprehensive and integrated account of the current status of paediatric epilepsy surgery. The book is divided into 11 sections which cover the range of clinical syndromes; cognitive and psychiatric aspects of childhood epilepsy; failure of medical treatment; presurgical evaluation; ethical issues; and finally the full range of surgical procedures, their outcomes, and complications. The chapters are succinct, well referenced, and consider contentious issues in a balanced manner.

This book will be invaluable to both expert and non-expert clinicians alike who are involved in the care of these patients. It will be of particular interest to paediatricians and neurologists referring patients to specialist centres, to those with ambitions to start up their own paediatric epilepsy programmes, and as a reference work for those already established in the field. It is to be commended.

ROBERT MACFARLANE

SHORT NOTICES

Readers may be interested in:


Continuous intrathecal baclofen infusion alleviates autonomic dysfunction in patients with severe supraspinal spasticity
R BECKER, U SURE, M PETERMEYER and H BERTALANFFY

J Neurol Neurosurg Psychiatry 1999 66: 114
doi: 10.1136/jnnp.66.1.114

Updated information and services can be found at: http://jnnp.bmj.com/content/66/1/114.1

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