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 antidiurenergic medication. Patients may present with severe arterial hypertension, tachycardia, hyperhidrosis, hypersalivation, and bronchial hypersecretion. Mortality is mainly influenced by these symptoms. In addition to antidiurenergic medication sedatives and analgesics 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 and autonomic dysfunction 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 tetraplegia from either hypoxic or traumatic brain injury were treated with CIBI. Before admission all patients were treated with maximum doses of various oral antispasitics.

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

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

The following conclusions can be drawn:

1. Improvement of autonomic dysfunctions with continuous intrathecal GABA-B agonist application was achieved in all patients treated and should be considered by others.
2. The use of CIBI offers a treatment of otherwise unresponsive autonomic instability in the acute medical setting.
3. The pharmacological background of these findings needs further investigation. Our preliminary findings should be confirmed in pharmacological testing set-up.

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#LETTERS TO THE EDITOR#

Continuous intrathecal baclofen infusion alleviates autonomic dysfunction in patients with severe supraspinal spasticity

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). 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 hypersecretion (5/6). Agitation was seen in five of six patients. In five patients the symptoms disappeared with intrathecal baclofen therapy. In one patient (No 5), 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 delayed cerebral and extraspinal 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 seen 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 (30% of the original communication). Tachycardia, arterial hypertension or hypotension, often associated with agitation, hyperhidrosis, hypersalivation, and tracheobronchial hyperssecretion 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.

Table 1: Autonomic dysfunctions during the course of treatment

<table>
<thead>
<tr>
<th>Patient No</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tachycardia</td>
<td>0</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>Arterial hypertension or hypotension</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>Hyperhidrosis</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hypersalivation</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Tracheobronchial hypersecretion</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Agitation</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

0=Not present, ±=improved, ±=unchanged.

The findings presented should be confirmed in different clinical situations—especially in patients with altered mental status. So far, we have restricted the indication for intrathecal baclofen therapy to the treatment of patients in a stable clinical condition. However, after these preliminary findings we now support an early treatment to achieve an autonomic stabilisation and early prevention of contractures in patients with supraspinal spasticity.

The following conclusions can be drawn:

1. Improvement of autonomic dysfunctions with continuous intrathecal GABA-B agonist application was achieved in all patients treated and should be considered by others.
2. The use of CIBI offers a treatment of otherwise unresponsive autonomic instability in the acute medical setting.
3. The pharmacological background of these findings needs further investigation. Our preliminary findings should be confirmed in pharmacological testing set-up.

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3 Amano M, Kubo T. Involvement of both GABA A and GABA B receptors in tonic inhibitory control of blood pressure at the rostral ventrolateral medulla of the rat. Naunyn-Schmiedeberg’s Arch Pharmacol 1993;348:1146–53.

A new case of late onset Lafora’s disease without generalised seizures

Lafora’s disease is clinically characterised by the triad of epilepsy, progressive dementia, and myoclonus as well as Lafora bodies in the brain and other tissues. The onset of this autosomal recessive disease is usually between the ages of 6 and 20 with a duration of 2–10 years. It can begin with generalised tonic-clonic seizures or focal seizures which are especially visual. Slight erratic myoclonus appears progressively with twicking movements of the fingers and facial

3 Amano M, Kubo T. Involvement of both GABA A and GABA B receptors in tonic inhibitory control of blood pressure at the rostral ventrolateral medulla of the rat. Naunyn-Schmiedeberg’s Arch Pharmacol 1993;348:1146–53.
The patient was a young woman who, at the age of 29, 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 four consecutive words or digits. In addition, she had dysarthria and pronounced the final words of a sentence in a musical tone. She also had slight myoclonus in her hands and face. Occasionally, after a sudden stimulus she displayed myoclonic jerks in her four limbs but at no time did she lose consciousness. Generalised hypreflexia with negative Babinski's sign was seen. The fundus of the eye was normal.

General analysis, routine laboratory analysis, serological tests, and CSF examinations were either normal or negative, as was an EEG study. The visual, auditory, and somatosensory evoked potentials were all normal. Brain CT and MRI all showed evidence of a slight degree of subcortical atrophy.

The first EEG record, carried out the day after admission, showed slightly slow background activity with bursts of slow waves and paroxysmal spikes and waves. Intermittent photic stimulation induced generalised discharges of slow and sharp waves with bursts of spike and polyspike wave complexes in occipital regions. Unexpectedly, on one occasion a massive generalised myoclonic seizure with a frequency of 15 flashes/second was seen but the patient did not lose consciousness. Six subsequent records were carried out with intervals of 4 days but there was no more evidence of photoparoxysmal discharges.

Background activity continued to be slow but there were persistent discharges of slow wave spike complexes of one cycle/second and 100–200 µV in the occipital regions. Treatment with valproic acid and clonazepam was initiated and the patient was discharged for observation and follow up.

At the age of 29 the patient moved to her parents' home as she had become unable to carry out any housework or other household tasks, 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 register 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 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 aminia, her deep reflexes are brisk, she has only slight erratic myoclonus but no other seizures. The EEG record 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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Abundance of left hemispheric embolic strokes complicating coronary angiography and PTCA

Stroke is a well known complication of coronary catheterisation but there are only a 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 and incidence as minor risk factors 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 discharge diagnoses 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 hemiparesis 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 the stroke event.

We further classified ischaemic events as embolic in patients with hyperacute presentation of stroke, whereas only one patient (6%) had a minor right hemispheric stroke, whereas only one patient (6%) had a minor right hemispheric stroke (p<0.0001, \( \chi^2 \)), and two patients (12.5%) had a vertebrobasilar event (p=0.0002, \( \chi^2 \)).

There were no strokes, 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 cosmetic heart failure). This difference was significant (p=0.001, \( \chi^2 \)).

The stroke rate of 0.23% after cardiac procedures found in our study is similar to other reports. Identification of patients who are prone to have a stroke as well as methods for its development might enable us to lower the risk of this serious complication. Brown and Topol reported that stroke patients had more risk factors for cerebrovascular disease than controls. Alternatively, our study suggests that performance of cardiac procedures 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 hypercoagulable state at the time of the procedure.

The clinical features of most stroke patients in our series are typical for an embolic event. Our finding that these emboli involve preferentially the left hemisphere, may provide a clue as to their source and mechanism of dislodgement. We cannot rule out completely a selection bias in this retrospective study as minor right hemispheric events may escape diagnosis more often than left hemispheric ones. However, such a bias does not hold for major neurological events. In addition, all patients were subject to close monitoring in an intensive care unit. 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 distal bend of the aortic arch, which is closer to the origin of the left common carotid artery, may cause dislodgement of atheromatous material preferentially into that artery. It was previously suggested that scraping the ascending aorta while searching for the origin of coronary arteriosclerosis was the cause of stroke. However, Tunick et al showed that most protruding aortic atheromas are located distal to the innominate artery, and are therefore distant from the left hemispheric strokes in most patients. This is in good agreement with our proposed mechanism 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 catheterisation was performed, atheroma may escape 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 the aortic arch, which is closer to the origin of the left common carotid artery, may cause dislodgement of atheromatous material preferentially into that artery. It was previously suggested that scraping the ascending aorta while searching for the origin of coronary arteriosclerosis was the cause of stroke. However, Tunick et al showed that most protruding aortic atheromas are located distal to the innominate artery, and are therefore distant from the left hemispheric strokes in most patients. This is in good agreement with our proposed mechanism 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.
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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Letters, Correspondence, Book reviews


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, surrounding 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 and second 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 spontaneously 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. The argument for surgery can also be made with lesions producing repetitive or progressive symptoms who there is significant neurological disability. Unfortunately, to date predictors of timing and size of hemorrhage are still unknown. In recent years lesion size and the number of previous hemorrhages has become a factor. 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.

The family tree. Males appear as squares and females as circles. Affected people are shaded and those who are dead appear with an oblique slash. The original reference case is marked with an asterisk.
The proband (figure; III.1) is currently 28 years of age, and has a 10 year history of insidious onset and progressive unsteadiness when walking. Upper limb ataxia and dysarthria had supervened around 7 years previously. He needs the support of a single person when walking. Upper limb ataxia and dysarthria had supervened around 7 years previously. He needs the support of a single person when walking. Upper limb ataxia and dysarthria had supervened around 7 years previously. He needs the support of a single person when walking.

The proband’s father (II.4) is currently 47 years of age. He sought help 4 years ago for partial and generalised seizures, and is currently taking lamotrigine. He admitted to heavy alcohol intake. Examination disclosed hepatomegaly and some memory impairment. There was sustained horizontal gaze nystagmus, limb incoordination worse on the left, diminished knee jerks, absent ankle jerks, and flexor plantar responses. He has marked truncal ataxia. Brain MRI was normal.

The proband’s daughter (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 perfuncted. She could not sit unsupported, was unable to reach out for toys, and was hypotonic. She had gaze dependent 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. II.4 and IV.2 were heterozygous for the GAA expansion, the expanded allele bearing a repeat of 840 triplets consistent with their carrier status. III.5 was also heterozygous for the expansion, and II.4 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 viral infections in IV.2). In a recent series of 56 pedigrees, at least two heterozygous parents (both fathers) manifesting ataxic features were identified.5 No data are available on possible environmental insults in these members. The current finding lends support to the conclusion of Lamont et al.,1 that a history of ataxia preceeding (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.

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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 supine 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 cranietomy. 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 cerebellar hemisphere and both occipital lobes appeared hypodense. There were also high density haemorrhagic lesions in the left temporal, right frontal and occipital lobes. She died the next day.

At postmortem the brain was swollen and markedly congested with patchy areas of subarachnoid haemorrhage scattered over the cerebral convolutions. The venous sinuses and cortical veins were patent. The vessels of the circle of Willis were patent and free of atheroma. Multiple haemorrhagic venous infarcts were present in the right anterior frontal (40×30×20 mm), right posterior frontal (30×20×20 mm), right parietal (15×10×10 mm), right temporal (20×20×10 mm), right occipital (25×10×20 mm) and left posterior frontal (30×45×40 mm) lobes. The overlying subarachnoid space and sulci in the affected areas were widely distended with blood (sulcal haematomas) and adjacent cerebral cortex and underlying subcortical white matter were disrupted by confluent small haemorrhages or haemorrhagic infarction of venous type. Microsurgical 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 supratentorial microvascular decompression all in the lateral decubitus position. There were two fatal intracerebral haemorrhages but there was no postmortem. Excessive cerebellar retraction 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 hypertension 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 important but the neck positioning.

A strong supposition of the hypothesis of mechanical venous obstruction, Gooding and Stimac have demonstrated in animal and infant cadaver models that turning the head to one side increased in 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 and Parker showed that jugular 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 cryogenic cirsiorhitis 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 the liver showed multi-focal 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 intracerebral 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 metasta.

Figure 1 Brain CT on day 10 showing multiple areas of cerebral haemorrhage.

Figure 2 Brain CT 1 month postdischarge showing complete resolution of the haemorrhagic cerebral lesions.
Contribution of in vivo 1H 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 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 perform a stereotactic biopsy even in elderly or severely deteriorated patients.

Recently, in vivo 1H 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.1

We report a case in which the diagnosis of an abscess was made on the basis of in vivo 1H MRS resulting in complete recovery.

A 50 year old woman with headache and increasing confusion over the previous 3 days 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/mm3 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 methylprednisolone resulted in clinical improvement and a stereotactic biopsy was planned after the performance of an in vivo 1H 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 x 20 mm x 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 contralateral 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 1H 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 1H NMR (figure C). The resonances were assigned by spiking samples with authentic standards. The main signals in the 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 1H MRS study.

The status of the patient improved rapidly with a 3 week intravenous antibiotic therapy with cefotaxime and metronidazole, then cefotaxime and metronidazole, then ceftriaxone and metronidazole for another 6 weeks. At the end of this period, in vivo 1H 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 1H 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.

Despite improvements in neuroimaging, CT guided stereotaxy,1 and newer antibiotics, this so called benign pathology kills one patient out of four, sometimes only because of delayed diagnosis. Radiologically, an abscess can mimic a brain tumour whether it be in the collected stage or, even more so, in the early stages of cerebritis.2 Misdiagnosis as a brain tumour can lead to inappropriate corticotherapy, resulting in a clinical worsening as happened in our patient, or in a dramatic intraventricular rupture with mortality as high as 80%.1 Thus diagnosis cannot rely only on clinical and radiological features; stereotactic aspiration is mandatory.

In vivo 1H 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 leukocytes against bacterial aggression. Lactate was also found but its bacterial origin was not certain. Moreover, a decrease of neuron density, energy metabolism failure, and phosphomembrane 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.4 High levels of succinate and acetate have been detected in the 1H NMR spectrum of aspirated abscess material.5

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.
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 difficult to compare the 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 expected 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 for 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 shown is about 8% (95% CI 1%-17%). 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.

P-H M VAN DER KUY
J H M LOHMAN
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Goadsby's editorial says many sensible things, but there are significant drawbacks not only conclusion can be drawn that the data supporting his statements that “the benefit of the active drug is added to that of placebo” or that “drug-combined therapeutic gain corrects for the effect of placebo.”

Consider the exceptional drug, megartripran, which is effective in almost all patients. In one study, in which the investiga- tors were very excited about the drug’s prospects and are also excellent caregivers, the placebo response is 45%, whereas the megartripran 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 megartripran 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 megartripran.

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 HC et al, IIIrd European Headache Federa- tion Meeting, Sardinia, 1996). However, in these and other studies the zolmitriptan response rate remained remarkably constant, between 62%-67%. 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 compare different drugs in different studies taking the different placebo responses into consideration. In my opinion it is inappropriate to use the placebo response as a qualitative measure and using it as a variable to compare drugs from different studies with different placebo responses. If there are different percentages of placebo response, the only conclusion can be drawn that the data from different studies cannot be compared.

Moreover, using the range of the SD of one drug to judge the other also seems to me to a methodological failure.

WILLEM J MEIJLER
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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, myocardi al 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.

KENNETH B KLEIN
Rainbird Island, WA, USA

Goadsby replies:

I thank van der Kuy and Lohman for their Goadsby replies: triptans. For truly valid comparisons among the trials remain the definitive approach. Hopefully, such trials will be forthcoming to allow for truly valid comparisons among the triptans.

KENNETH B KLEIN
Rainbird Island, WA, USA

Goadsby replies:

I thank van der Kuy and Lohman for their interest in the editorial and agree that treat ment choice is di

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KLEIN:1210–18.


The example of the zolmitriptan development programme, of which Klein was directly involved, is curious. The very large placebo response quoted by Klein has never been published (Diener, H-C et al. 3rd European Headache Federation Meeting Sardinia, 1996), may have resulted from a very skewed randomization. This problem, 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 hazardous 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 never 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 as to safety and tolerability it is in the mind of industry, not on 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 logDpH 7.4 which is \(0.2 \not= 0.02\), a typographical error pointed out to me by an earlier neurological colleague (Darryl 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.

PJ G GOADSBY
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8 Elferink AJA, Van Zwieten-Booij BJ. New antiepileptic drugs: analysis based on number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

9 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

10 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

11 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

12 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

13 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

14 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

15 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

16 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.

17 Rembold CM. A number needed to treat shows differences between drugs. Bmj 1997;314:603–5.
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 questions.

Firstly, two of the three papers quoted by Mark do not provide data on hemianopic bisection bias. In one,1 the phenomenon is mentioned in a single sentence in the methods section; in the other,2 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 contrast in 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.1,3 They quote two works from the English literature. One is the paper by Schenkenberg et al. This report analyzed bisection by 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 neural damage, 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 observation is a monograph by Teuber et al. This was also puzzling, because line drawings showed that the bisection field was leftward, as we find with hemianopia. Myoclonus of peripheral origin usually lasting less than 200 ms (often even in rhythmic myoclonus with long EMG duration will appear brief or shock-like. We agree that the EMG bursts resemble that of the peripheral nervous system. A variant of PLMT, however, may have an associated pain and is referred to as the painless legs and moving toes syndrome. The findings presented by Assal et al may very well represent myokymia. Myokymic discharges fire rhythmically at 1–20 Hz and in trains that last a few seconds followed by a short period of silence. Myokymia is often seen in peripheral nerve disorders, is abolished by local nerve blocks, and is thought to represent eptogenotypically generated potentials in demyelinated nerve fibres. A myokymic EMG pattern has also been described in PLMT due to hypertrophic mononeuropathy.3

VIRGILIO GERALD H EVIDENTE
JOHN N CAVINNESS
Parkinson’s Disease and Movement Disorders Center, Mayo Clinic, Scottsdale, AZ, USA

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.” They presented a patient with painless “myokymia” involving the dorsal interosseous muscle of the foot that was abolished by local anesthetic block. Electromyography showed bursts of rhythmic spontaneous grouping of motor units of 200 to 400 ms duration occurring at 1–6 Hz frequency. They take issue with the usage of the term “myokymia” for the abnormal movements described. Myoclonus is a brief, shock-like, involuntary movement with EMG activity usually lasting less than 200 ms (often even 50 ms or less). Myoclonic discharges are usually arrhythmic and irregular, with the exception of segmental (brachium or spinal) myoclonus which presents with rhythmic, long EMG bursts (for example, involving several or spinal or body segments. Descriptions of the electrophysiology of peripheral myoclonus are scarce. In our experience, the EMG bursts in peripheral myoclonus can be >100 ms in duration (often even 500 ms), but are not truly sinusoidal, are irregular in frequency and duration, and have a very variable interburst interval. By contrast, in the case presented by Assal et al, the EMG bursts appear sinusoidal, regular in frequency, and come in trains of EMG activity that are interrupted by periods of silence. Furthermore, even in rhythmic myoclonus with long EMG bursts, the movements still appear brief and shock-like. In contrast, the description by Assal et al of the “myoclonus” in their case was continuous and rhythmic. It is unlikely that EMG discharges of 400 ms duration will appear brief or shock-like. We agree that the EMG bursts resemble that of the painless legs and moving toes syndrome, which is often associated with peripheral nerve or nerve root lesions. A variant of PLMT, however, can have no associated pain and is referred to as the painless legs and moving toes syndrome.


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 nor 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. Electrophysiological myokymic discharges concern single motor units whereas many motor units fired synchronously in our patient. In some conditions myokymia may be related to the discharge of numerous single or grouped fasciculation potentials, but this activity does not manifest gross rhythmic muscle contractions either. Moreover, when myokymia have a peripheral origin, the ectopic activity originates from the motor nerve or the contracting muscle 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 be implicated. This might be an explanation for the rhythm and long duration of the myo- clonic bursts seen since, as mentioned by Evi- dentie and Caviness, spinal myoclonus may present such characteristics. This last hypoth- esis reinforces our impression that myoclonus, already used by others in similar conditions, was the most adequate term to describe the single painless moving toe of our patient.

MICHEL R MAGISTRIS  FREDERIC ASSAL FRANCOIS JG VINGERHOETS 
Clinique et Polyclinique de Neurologie, H C U G, Geneva, Switzerland


BOOK REVIEWS


This is a new pocket size handbook on Parkinson’s disease with 11 chapters covering all aspects of this disorder ranging from defi- nition and classification to newer surgical therapies. It is well written and easy to read, although the format means that some of the pictures and figures are so small that they are difficult to interpret. However, the simple chapter layout and the marking out of them on the margins of the page does make it easy to use. The book makes good use of graphs and tables, although some of the tables are so overinclusive that it is hard to know their value in terms of what is rare and what is not.

The text is concise and clear although there are controversial statements that I would take issue with—for example, the existence of paraneoplastic parkinsonism and the notion that the intralaminar nuclei of the thalamus are part of the basal ganglia. Conversely other controversial issues are not discussed and these include the cognitive aspects of early 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 Parkin- son’s disease can sometimes be hard to extract. Indeed the book is clearly written for the American physician, and so there is essentially no mention of liuride, apomor- phine, or cabergoline whereas pramipexole has a very high profile and selegiline receives little adverse publicity. Furthermore, the classification of multisystem atrophy and the distinction between it and striatogniral de- generation, Shy-Drager and sporadic OPDCA in the United States can be confusing. This emphasis obviously makes it harder to accommodate in the United Kingdom mar- ket, and the audience for which the book is intended, 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 been published on sleep disor- ders 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, in conditions such as the restless legs syndrome, are well described.


This book gives little coverage of the many other chapters would have been helpful. The book follows on from an International Symposium on Congenital Muscular Dystrophies which was held in Tokyo in July 1994. The book has many positive features but also some less good fea- tures, so common in edited multi-author 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 particu- larly enjoyed reading (and learning) about the different clinical phenotypes from clinicians who clearly have seen many cases. The fasci- nating association of the muscular dystrophy with CNS abnormalities both clinical and radiological, are well described.

The chapters describing the morphological features in both muscular dystrophy and brain are also comprehensive and helpful to those trying to establish a diagnosis. There is also an interesting chapter on the clinical manage- ment 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 protein, in this case merosin, in a subgroup of patients. The most common form of congenital muscular dystro- phy in Japan, Fukuyarna congenital muscular dystrophy, has been mapped to a specific chromosomal location and a disease causative 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 con- cerning with many multi-author 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 impor- tant 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 remem- ber the 85 varieties of floppiness. Be warned, I am now an expert and can confidently diag- nose a case of Fukuyama muscular dystro- phy; shame that they are only seen in Japan!

DOUG TURNBULL


This book describes in detail the clinical fea- tures and latest research findings on congeni- tal muscular dystrophy. The book follows on from an International Symposium on Congenital Muscular Dystrophies which was held in Tokyo in July 1994. The book has many positive features but also some less good fea- tures, so common in edited multi-author 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 particu- larly enjoyed reading (and learning) about the different clinical phenotypes from clinicians who clearly have seen many cases. The fasci-
A new mantra in the health service is the
rehabilitation of all its problems; the dominance
of the health professionals (mainly doctors)
and thus the dominance of the medical model
of ill health; the subordination of manage-
ment to expertise; the lack of citizen/user
input; the lack of clear financial and clinical
information; and the fudging of the rationing
issue. Adding to these structural problems are
the pressures of demography, technological
innovation and dysfunctional consumer choice.

Oliver Morgan’s book is a worthy but not
exciting attempt to clarify the elements of
each of these problems. The answers,
threaded through the text with a slightly irrit-
ating presumptuation of radicalism, are
accountability, transparency, and high quality
information for all.

All this is to be achieved by creating an
informed and participatory citizenry through
the imaginative use of information technol-
ogy, coupled with diffusion through society of
the many exciting new ways of engaging the
public.

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 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.

JOHN JENNEN

**Handbook of Medical Psychiatry.** David P
MOORE and James W JEFFERSON. (Pp 545).
Published by Mosby, St Louis, Missouri,

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 is an excellent book, covering
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 disor-
der. 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 importance of neuropsychophar-
macology and the physiology of psychiatric
and neurological conditions and that the book
is designed to reflect this. It certainly does, hav-
ing a very medical flavour, and in keeping with
textbook norms, it 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

**Paediatric Epilepsy Syndromes and
their Surgical Treatment.** Edited by
Tuxhorn, H Holthausen, and H Boenigh.
(Pp 928, £85.00). Published by John Libby

In recent years there has been a significant
increase in the number of children who are
investigated as potential candidates for epi-
lepsy surgery, and in the number of centres
which offer this service. Some of the reasons
behind this are improved access to high reso-
lution MRI and other techniques for the
assessment of the origins of epilepsy, an
increased level of awareness and expectation
among parents, and acceptance by paedatri-
cians 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 pathol-
gy. 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 psychia-
tric aspects of childhood epilepsy; failure of
medical treatment; presurgical evaluation;
ethical issues; and finally the full range of sur-
gical procedures, their outcomes, and compli-
cations. The chapters are succinct, well
referred, 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 estab-
lished in the field. It is to be commended.

ROBERT MACFARLANE

**Healthcare Litigation.** Edited by
Michael Barnes, Bill Brathwaite, and
Anthony B Ward. (Pp 432, £49.50). Pub-
lished by Blackwell Science, Oxford,

**Psychiatric Disorders with a Bio-
chemical Basis.** Edited by
David Donaldson. (Pp 242, £28.00).
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