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

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 antiadrenergic medication. Patients may present with severe arterial hypertension, tachycardia, hyperhidrosis, hypersalivation, and bronchial hyperssecretion. Mortality is mainly influenced by these symptoms. In addition to antiadrenergic 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 physiotherapeutic 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 effect on autonomic symptoms originating from severe hypoxic or traumatic brain damage. Eighteen patients with severe tetraspasticity 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 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.

To confirm 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 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.

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 (3/6), hypersalivation (5/6), tracheobronchial hyperssecretion (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 decerebrate extrapyramidal and extensor 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 appear 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 application of 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 might enhance the positive effect of CIBI on severe autonomic dysfunctions.

GABA-B receptors are widely distributed throughout the CNS. The action of GABA-B receptors on autonomic regulation, especially in pathological conditions, is unknown. Sympathetic and cardiovascular depressant effects of baclofen have been shown with local application of GABA-A and GABA-B receptor agonists and antagonists at the rostral ventrolateral medulla (RVLM) of the rat, in the cat ventromedial and posterior hypothalamus, and in isolated spinal cord preparations. It was possible to increase blood pressure with injection into the anterior hypothalamus and nucleus tractus solitarius.

Autonomic nervous system regulation, involving different regions of the CNS, is complex. Concerning the positive effect of intrathecal baclofen on autonomic dysfunction the findings presented should be confirmed in different clinical situations—for instance, in patients with acute brain injury. 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 test settings.

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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>±</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hypersalivation</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Tracheobronchial hyperssecretion</td>
<td>+</td>
<td>±</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Agitation</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

0=Not present; +=improved; ±=unchanged.
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 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 had dysphasia, her deep reflexes are brisk, she clonus is present accompanied or not by slight myoclonus. We use 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 parent’s home as she had become unable to carry out any housework or other household tasks, including the care of her child. A Ben-ton 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 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 paroxysmatic 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 was 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.7 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 rates in 2359 patients as minor as right atrial afebrile ones. 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 events. We further classified ischaemic events as 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 hyperacuteness presentation strongly suggest that the neurological events were embolic in nature.

Stroke locations were distributed unevenly between the main vascular territories. Thirteen 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 vertebrobasilar event (p<0.0002, χ²).

There was left-handedness 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 differed from the control group in 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 difference was significant (p=0.01, χ²).

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 mechanisms 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 generalised hypercoaguable 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 this embolism involves 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 and the 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 scrapping the ascending aorta while searching for the ostia of coronary arteries 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 not a 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 the catheter was left 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 thin strenght arteries that subclavian preferentially into the vertebral system, sparing 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 vertebrobasilar event.

In conclusion, we suggest that cardiac catheterisation still carries a measurable risk for stroke, especially in patients with acute angina pectoris or congestive heart failure.

### Table 1: Clinical Characteristics of Stroke and Control Patients

<table>
<thead>
<tr>
<th></th>
<th>Stroke (%)</th>
<th>Control (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>16</td>
<td>48</td>
</tr>
<tr>
<td>Male (%)</td>
<td>10 (62)</td>
<td>25 (52)</td>
</tr>
<tr>
<td>Mean age (range)</td>
<td>61 (35–81)</td>
<td>61 (40–80)</td>
</tr>
<tr>
<td>Hypertension (%)</td>
<td>6 (35)</td>
<td>24 (50)</td>
</tr>
<tr>
<td>Hyperlipidaemia (%)</td>
<td>9 (55)</td>
<td>23 (40)</td>
</tr>
<tr>
<td>Diabetes (%)</td>
<td>5 (29)</td>
<td>15 (31)</td>
</tr>
<tr>
<td>Smoking (%)</td>
<td>8 (47)</td>
<td>31 (64)</td>
</tr>
<tr>
<td>Atrial fibrillation (%)</td>
<td>2 (12)</td>
<td>2 (4)</td>
</tr>
<tr>
<td>CoAng/PTCA (%)</td>
<td>10 (62)</td>
<td>24 (47)</td>
</tr>
<tr>
<td>Indication for CoAng/PTCA:</td>
<td></td>
<td>0.01*</td>
</tr>
<tr>
<td>Urgent</td>
<td>12 (75)</td>
<td>18 (37)</td>
</tr>
<tr>
<td>Unstable AP</td>
<td>10 (62)</td>
<td>13 (31)</td>
</tr>
<tr>
<td>Acute MI</td>
<td>2 (13)</td>
<td>5 (10)</td>
</tr>
<tr>
<td>Non-urgent</td>
<td>4 (25)</td>
<td>30 (63)</td>
</tr>
<tr>
<td>Stable AP</td>
<td>1 (6)</td>
<td>1 (2)</td>
</tr>
<tr>
<td>CHF</td>
<td>1</td>
<td>NS†</td>
</tr>
</tbody>
</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.
Males appear as squares and females as circles. A Cavernomas have a characteristic


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 seen.

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 in the family tree (figure) it 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 intracranial 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 reversibility of hemiparesis when aged 20. He was noted to have the cutaneous angiomatosus 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 where there is significant neurological disability. Typically, the patient is younger and the lesion appears to be growing. However, in a recent study involving 145 patients the authors suggest that risk factors for aggressive behaviour include pregnancy, familial or multiple form, previous uncontrolled bleed, or stereotactic radiotherapy, incompletely, 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. Furthermore, 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

Presented by 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 had marked truncal ataxia. Brain MRI was normal.

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 evoked 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 perfused. 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. II.5 was also heterozygous for the expansion, and II.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.1 No data are available on possible environmental insults in these members. The current finding lends support to the conclusion of Lamont et al.,3 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.

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 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 athrombosis. Multiple haemorrhagic venous infarcts were present in the right anterior frontal (40×30×20 mm), right posterior frontal (30×20×20 mm), right parietal (15×15×10 mm), right temporal (20×20×10 mm), right occipital (25×10×20 mm) and left posterior frontal (35×20×40 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. Postmortem examination disclosed venous and capillary congestion and coalescence of multiple microhaemorrhages. The cause remained elusive.

In 1978 Jannetta et al reviewed 825 cases of posterior fossa surgery. 5 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 Hanakta and Kondo reviewed 278 patients with supratentorial microvascular decompression surgery all in the lateral decubitus position. There were two fatal intracerebral haemorrhages but there was no postmortem. Excessive cerebellar retraction and 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 obstruction. Venous congestion 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.

I am 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 results in tension 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 venous occlusion on the side of the dominant venous drainage can result in severely limited cerebral venous drainage. Emerson 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 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 and venous infarction caused by bilateral internal jugular vein 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 extraocular venous obstruction are two possible mechanisms with important management implications.

We thank the Department of Radiology and Department of Media and Illustration, Flinders Medical Centre, for their assistance in the preparation of radiology and pathology illustrations. We also thank Vicky Frangoulis for manuscript preparation.

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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 this pathology is to consider an abscess when neuroradiological imaging may be confusing and with neurological signs similar to tumours, permitting a non-specific without evidence of infection.

Contribution of 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 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 gadoxinum 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 cares 2 weeks earlier. Intravenous methylprednisolone 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 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 visualized 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 meronadazole, 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 returned to a normal metabolism. Six months later, 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 indication of infection; (4) known extracranial infection. Despite improvements in neuroimaging, CT guided stereotaxy, 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. 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%. 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 polymicrobial 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 phospholipid 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 a 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 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 useful to compare the ratio 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 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. For sumatriptan 100 mg the therapeutic penalty is 17% (95% CI 10%–24%) according to the text, but in figure 3 the penalty shown is about 8% (95% CI 4%–12%). 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, but considers that it is an urgent 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 “a therapeutic gain corrects for the effect of placebo.”

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

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 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

Therapeutic gain on the x axis is defined as headache response (moderate/severe to nil/mild) at 2 hours–headache response to placebo. Therapeutic penalty on the y axis is defined as adverse event rate on active drug – adverse event rate on placebo. Data are given as 95% CIs. The dashed line indicates the rTG/TP for sumatriptan 100, which acts as a reference. The following triptans are included: sumatriptan 50 mg (s 50), sumatriptan 100 mg (s 100), naratriptan 2.5 mg (n 2.5), zolmitriptan 2.5 mg (z 2.5), and zolmitriptan 5 mg (z 5).
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.

KENNETH B KLEIN
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Goadsby replies:

I thank van der Kuy and Lohman for their interest in the editorial9 and agree that treatment choice 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 end points as they are translated into clinical practice. Perhaps we will only be able to sort these compounds out when more direct head to head comparisons are made. In the meantime the trial data must be mixed with clinical experience. My experience has been that whereas the differences between the triptans are not huge there are differences that matter in practice, and fitting the drug to the clinical problem can be very rewarding in terms of satisfying a patient’s unmet therapeutic needs. Since the editorial a further compound has been approved in Europe, rizatriptan at a dose of 10 mg orally, bringing to four (including sumatriptan, naratriptan, and zolmitriptan), the choices we have for non-ergotamine used specific antimigraine treatments. The data indicate that rizatriptan will also have an important place in the treatment of acute migraine attacks.

In regard to the numbers that have been used. Unfortunately, the summary data, indeed the field, represent a moving target. The data quoted in the text were from a single large study and this data 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 (Tiftel-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. Certainly in the editorial the point was made that direct comparative studies are necessary and stated this were from a meta-analysis that in-"
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 lines 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.3,4 He quotes two works from the English literature. One is the paper by Schenkenberg et al.5 This report analyzed bisection 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 inform- al test on the small sample of five patients with left hemispheric lesions shows no difference from the controls.

Another paper is a monograph by Teuber et al.6 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,7 a case study of a man with a right occipital wound and left hemi- anopia. 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.8 We do seem to be the first to make this finding, in his report of a man with left hemianopia and leftward bisection errors. Apart from precedence, though, few conclusions are permitted from a single case study without controls.

The most important work we discovered is that by Liepmann and Kalmus.9 We appreciate the opportunity to publicise the prior observations of Axenfeld10 and Liepmann and Kalmus,11 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 concerning with the persistence of the bias and the conclusion that it is caused by hemianopia, with the use of statistical analy- sis, normal controls, and non-hemianopic brain-damaged controls, none of which were used in the older German work. We hope that our brief initial report and this correspond- ence 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.” They presented a patient with painless “myokymic” movements of the second toe of our patient. The authors reply:

Mark’s question. Their comments on the

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 “myokymic” movements of the second toe of our patient. Our brief initial report and this correspondence increase awareness of “hemianopic bisection error.”


The authors reply: We thank Vingerhoets and Carniss 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 move- ment 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 studies in our patient did not show evidence of myokymia discharges concern single motor units whereas many motor units fired synchronously in our patient. In some condi- tions myokymia may be related to the discharge of numerous single or grouped fas- ciculation potentials,1 but this activity does not manifest gross rhythmic muscle contrac- tions either. Moreover, when myokymia have a peripheral origin, the ectopic activity origi- nates from the motor nerve supplying the muscle1 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
BOOK REVIEWS


This is a new pocket size handbook on Parkinson's disease with 11 chapters covering all aspects of this disorder ranging from definition 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 proximate effect of levodopa in the development of dyskinesias and thus the treatment of young onset Parkinson's disease. Furthermore, the book rather omits 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 is essentially no mention of liuride, apomorphine, or cabergoline whereas pramipexole has a very high profile and selegeline receives little adverse publicity. Furthermore, the classification of multisystem atrophy and the distinction between it and striatonigral degeneration, Shy-Drager and sporadic OPCA in the United States can be confusing. This emphasis obviously makes it easier to accommodate in the United Kingdom market, and the audience for which the book is aimed, 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 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. The book deals mainly with hypnotic drugs and their use in insomnia and the place of stimulant drugs, especially amphetamines, in conditions causing excessive daytime sleepiness. The book 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 dissectioned 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 as preventive medicine is interesting and most of the chapters are enhanced by the inclusion of case studies. These are well chosen examples 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 fascinating association of the muscular dystrophy with CNS abnormalities both clinical and radiological, are well described.

The chapters describing the morphological features in both muscular dystrophies 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 protein, in this case merosin, in a subgroup of patients. The most common form of congenital muscular dystrophy in Japan, Fukuyama congenital muscular dystrophy, has been mapped to a specific chromosomal location and the 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 concerning with many multiauthour 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 that they 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 Fukuyama muscular dystrophy; shame that they are only seen in Japan!

DOUG TURNBULL

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 Dystrophies held in Tokyo in July 1994. The book has many positive features but also some less good features, 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 particularly enjoyed reading (and learning) about the different clinical phenotypes from clinicians who clearly have seen many cases. The fascinating association of the muscular dystrophy with CNS abnormalities both clinical and radiological, are well described.

The chapters describing the morphological features in both muscular dystrophies 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.

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A new mantra in the health service is the re-emphasis 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 management to profession; 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 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 irritating presumption 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 technology, coupled with diffusion through society of the many exciting new ways of engaging the public.

The book does, therefore, offer useful thoughts on the depressingly familiar question of what we should do to rescue our health system. But its style, half way between a scholarly review and a call to arms, makes for dreary reading. I wish it were more feisty. As it happens I agree with Morgan’s central thesis, that public engagement contributing to all aspects of health care has the best chance of provoking sensible reform. If only the text had done more to persuasively recommend that this radical option was not only right, but also worth striving for, I would have fewer reservations about the book and be clearer about who should read it.

ROBIN STOTT


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 sexual and 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 resource indeed 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 JENNBER


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 a delightful key to understanding the 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 role of 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 an extensive 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 an 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

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