Stiff person syndrome with eye movement abnormality, myasthenia gravis, and thymoma

Stiff person syndrome (SPS) is a rare disorder of the central nervous system characterised by progressive fluctuating rigidity and painful spasms of the body musculature. We describe a patient with SPS with positive glutamic acid decarboxylase (GAD) antibodies who developed diplopia. Thymoma was detected by computed tomography (CT), and after thymectomy his symptoms improved. One month after thymectomy, he tested positive for anti-decarboxylase receptor (AChR) antibodies.

Case report
A 45 year old man presented with a four week history of back pain and stiffness of his trunk causing difficulty in bending forward and turning over while lying down, which he attributed to a minor injury sustained while playing squash. He later developed asymmetrical stiffness of the legs and difficulty walking. His past medical history was notable for an episode of dysphagia (two weeks' duration) associated with heartburn six months ago; a gastroenterological evaluation and an endoscopy at that time were normal. He recovered spontaneously and there was no recurrence.

On examination his mental status, speech, and cranial nerves were normal. He had exaggerated lumbar lordosis. Neurological examination showed normal bulk with increased tone of the flexors and extensors of the knee and ankles. Power and coordination were normal, deep tendon reflexes were brisk, but he had flexor plantar responses. There was no evidence of fatigable muscle weakness. Sensory examination was normal.

A chest radiograph and magnetic resonance imaging (MRI) of the brain and the spinal cord were normal. He was anti-GAD antibody positive at 3.4 U/ml (radioimmunoassay; normal 0–1 U/ml). Full blood count, vitamin B12, folate, thyroid function tests, liver function tests, urea, electrolytes, glucose, cortisol, immunoglobulins, and electrocardiography (EKG) were normal. Stiffness resolved six months after thymectomy as evidenced by eye movement recordings that showed less variability of horizontal and vertical saccades with a tendency to be slow, and slight endpoint nystagmus. There was no ptosis or weakness after sustained upgaze for one minute. Eye movement recordings, obtained with a high resolution video pupil tracker (Eylux, Sensomotoric Instruments, Berlin, Germany; sample rate 250 Hz) confirmed the clinical findings (fig 1, top panel). Anti-AChR antibodies were negative. The neurological findings were unchanged.

Motor and sensory nerve conduction studies and ulnar and radial repetitive nerve stimulation were normal. Concentric needle electromyography (EMG) showed sporadic fasciculation potentials in the tibialis anterior. Single fibre EMG from 34 potential pairs from the orbiculalis oculi revealed only one site with definitely abnormal jitter. A chest CT scan revealed a thymic mass. Histological examination confirmed thymoma with minimal involvement of the perithymic fat. His symptoms improved over a month after thymectomy.

One year from the onset of symptoms, one month after thymectomy, he tested positive for anti-AChR antibodies (44×10^10 M/l) (radioimmunoassay in the same laboratory, normal 0–5×10^10 M/l) and remained positive for anti-GAD antibodies (2.0 U/ml). His eye movements improved significantly after thymectomy as evidenced by eye movement recordings that showed less variability of saccadic velocity (fig 1, bottom panel). Eighteen months after the onset of symptoms he is off medications and back to his normal routine. He has mild intermittent stiffness of his back, precipitated by anxiety. Occasional mild diplopia at far distance persists.

Discussion
SPS was first described by Moersch and Wolffman in 1956 and was subsequently shown to be associated with anti-GAD antibodies in 40–60% of cases and anti-amphiphysin antibodies in some paraneoplastic cases.

In 1990, Piccolo et al reported a case of generalised myasthenia in a patient with SPS. This patient had radiological evidence of thymoma. A patient in the series of Vincent et al had SPS with anti-GAD antibodies, neuromyotonia and myasthenia with anti-AChR antibodies. Nicholas et al reported a case of SPS associated with histologically proved thymoma, who developed ocular myasthenia after thymectomy.

Hagiwara et al described a patient with SPS associated with invasive thymoma but not with myasthenia or anti-AChR antibodies. However, since the patient reported by Piccolo et al developed myasthenia six years after spontaneous resolution of SPS, and our patient's anti-AChR antibodies turned positive after one year, it is possible that the patient reported by Hagiwara et al will develop myasthenia in the future. Saravanan et al described a patient with SPS associated with ocular myasthenia. Neither anti-AChR nor anti-GAD antibodies were detected.

At the time of initial presentation, our patient did not have any clear signs of generalised myasthenia, although the transient dysphagia he experienced prior to the development of symptoms of SPS may have represented symptoms of bulbar myasthenia.

Figure 1 Horizontal and vertical eye movement recordings during saccades; (top panel) before thymectomy and (bottom panel) after thymectomy.
Notably, Hagiwara et al’s patient also reported dysarthria, which could have been due to myasthenia. The diplopia, variable velocity of saccades and endpoint nystagmus was not exclusively due to ocular myasthenia. This patient became seropositive after 12 months, even though his myasthenic symptoms improved after thymectomy.

Five cases of SPS associated with myasthenia gravis have been reported. This is the first report of abnormalities on eye movement recordings strongly suggesting myasthenia gravis in SPS before the patient became seropositive for anti-AChR antibodies. Our patient is probably the third patient with SPS and myasthenia with historically proven thymoma and the second such patient with positive anti-GAD and anti-AChR antibodies. Our report suggests that patients with SPS can develop other autoantibody mediated disorders even after many months and should be followed up over a long period even if they are asymptomatic. In addition, when patients with SPS have eye movement abnormalities or bulbar symptoms, myasthenia gravis should be suspected even if they are negative for anti-AChR antibodies at presentation. Thymoma should be investigated for, as thymectomy may improve both SPS and myasthenia.

S Thomas
Department of Ophthalmology, University Hospitals Leicester, Leicester, UK

P Critchley, M Lawden
Department of Neurology, University Hospitals Leicester

S Farooq
Ophthalmology, University of Leicester, Leicester, UK

A Thomas
Department of Neurology, University Hospitals Leicester

F A Proudlock
Ophthalmology, University of Leicester

C S Constantinescu
Department of Neurology, University of Nottingham, Nottingham, UK

I Gottlob
Ophthalmology, University of Leicester

Correspondence to: Professor I Gottlob, Ophthalmology, University of Leicester, RKCSB, PO Box 65, Leicester, LE2 7UX, igottlob@leicester.ac.uk
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References
Cerebrospinal fluid was clear without pleocytosis, but the cerebrospinal fluid pressure was 350 mm H₂O.

Magnetic resonance imaging (MRI) of the neck with and without enhancement revealed thrombosis of the right IJV, starting from the junction with the right subclavian vein (see fig 1). However, there were no structural abnormalities adjacent to the right IJV, and the carotid arteries were normal. Digital subtraction venous angiography confirmed extensive thrombosis in the right IJV, the right sigmoid sinus, and the superior sagittal sinus (see fig 1). The rest of the intracranial dural sinuses were patent, and no vascular malformation was detected.

Phenytoin and valproic acid were promptly administered resulting in improvement in the patient's focal motor seizures. He was also given heparin and warfarin and the intra-arterial infusion of tissue plasminogen activator resulted in thrombosis at this unusual site. The rest of the examination was normal. Abdominal examination revealed a palpable mass in the right upper quadrant just lateral to the midline. Sonography of the abdomen revealed a unilateral retroperitoneal mass. A spinal MRI showed extensive spinal cord compression from T12 to L4 (fig 1). Biopsy of the abdominal retroperitoneal mass revealed neuroblastoma. The neonate was therefore surgically decompressed through an approach from T12 to L4 and a large intra-abdominal retroperitoneal mass was excised.

**Case 1**

A 4 month old baby boy with a diagnosis of unilateral leg palsy due to birth trauma, despite normal vaginal delivery, was admitted because of a palpable abdominal mass. The infant’s left leg lacked spontaneous movement, was flaccid, and deep tendon reflexes were absent. He had poor rectal tone and dribbling of urine. The levels of urinary catecholamine derivatives were increased. Spinal magnetic resonance imaging (MRI) demonstrated a large retroperitoneal mass with thoracolumbar cord involvement. A diagnosis of neuroblastoma was made following biopsy of the abdominal mass. Multimagent chemotherapy proved effective in reducing the size of neuroblastoma. His left leg function returned after several months’ chemotherapy. At present, after two years he is free of disease, he can stand and walk with a brace, and his neurogenic bladder is managed with clean intermittent catheterisation.

**Case 2**

A 10 day old baby boy was seen for evaluation of right lower limb weakness. He had not moved this leg since birth. He was born at full term via a normal vaginal delivery with vertex presentation. He had a hyper-extended thigh and decreased tone in the remainder of the leg. Other limbs were normal. Abdominal examination revealed a palpable mass in the right upper quadrant just lateral to the midline. Sonography of the abdomen revealed a unilateral retroperitoneal tumour adjacent to the right kidney with spinal cord involvement. A spinal MRI showed extensive spinal cord compression from T12 to L4 (fig 1). Biopsy of the paravertebral mass revealed neuroblastoma. The neonate was treated with multimagent chemotherapy. However, he developed paraplegia of the left leg within two weeks of starting chemotherapy. The spinal cord was therefore surgically decompressed through an osteoplastic laminotomy and the extradural mass was fully resected. Although there was partial recovery of left leg function the right limb remained plegic.

**Discussion**

Birth trauma causing brachial plexus injury is relatively common where obstetric services are limited, but lumbosacral plexopathy after a normal vaginal delivery is extremely rare. Unilateral lower extremity palsy in a neonate must lead the primary care provider to consider other diseases. The combination of neurological deficits and an abdominal mass should alert the physicians to consider neuroblastoma. Early diagnosis can improve outcome, and neuroblastoma diagnosed even in the prenatal period has been reported to have excellent prognosis.

**References**

Anorexia nervosa remission following left thalamic stroke

Anorexia nervosa is an intense fear of weight gain, inaccurate perception of body size, weight or shape, amenorrhoea, and a body weight <85% of expected weight (or mass index (BMI) ≤17.5). We report a patient who, following a left thalamic stroke demonstrated a remarkable recovery from a 7 year history of anorexia nervosa.

The patient grew up in a family with both parents and two older brothers. When she was 14 years old, a young cousin died of a “brain haemorrhage.” Six months later the patient started a “healthy eating” regimen. She was first admitted to hospital for her eating disorder in April 1995, aged 15 years, and was prescribed antidepressant medication. The problem continued despite psychiatric and psychological treatment (usual weight 43 kg, BMI 17).

In May 2002, aged 22 years, she experienced a sudden onset of right arm and leg weakness with a sensory disturbance of the right face, arm, and leg. There was no history of diabetes, cigarette smoking, illicit drugs, or oral contraceptive use. She was admitted to hospital. She was told that a computed tomogram (CT) showed that she either had a brain tumour or had suffered a stroke. She was transferred to the regional neurology unit. There she was alert, but had a slight decrease in sensation on the right side of the face; there was no visual field defect. She had a right pronator drift. She had grade 4 strength throughout the right upper limb. Leg strength was normal. The right arm and leg were mildly hyperaesthetic and there was impaired proprioception in the right fingers. There was right sided ataxia. On the right she had brisk reflexes and an extensor plantar response.

Her brain CT demonstrated left thalamic hypoattenuation, which on magnetic resonance imaging showed involvement of the left posterolateral thalamus and posterior temporal lobe (fig 1). The infarct area involved the left inferolateral artery territory. Magnetic resonance angiography was normal. Other investigations (chest x ray, electrocardiogram, thoracic echocardiogram, full blood profile, thrombophilia screen, glucose, liver function tests, and thyroid function tests) were normal. The patient was extremely anxious and thought frequently about her cousin’s death. However, she gradually improved and reported to a neuropsychologist that she no longer had an eating disorder. Her realisation came quite suddenly 3 days after her transfer to the neurology service. She chose cauliflower cheese for her evening meal and asked a visitor for a chocolate chip biscuit; neither of these foods would have been acceptable as part of her anorexic diet.

Within 6 months the patient gained 4 kg in weight (41 kg to 45 kg, BMI 18.7). Regular menstruses returned after two years of amenorrhoea. Eight months after the stroke she wrote the following descriptions of her feelings before and after her stroke: Pre-stroke: “Anorexia controlled my life and there were many hypotheses to account for her anorexia

She continued with antidepressant medication and her mood remained stable. The patient completed the Eating Disorders Inventory-2 from the perspectives of pre-stroke and 13 months post-stroke. The bulimia and interpersonal trust scales were at the mean for non-patient college females both pre- and post-stroke. Drive for thinness and body dissatisfaction were high pre-stroke, even in comparison with the eating disorder group. The scores on drive for thinness, body dissatisfaction, ineffectiveness, and interpersonal awareness all fell dramatically post-stroke, and her post-stroke scores were close to the mean for non-patient college females.

Discussion

Our patient demonstrated sustained remission from anorexia nervosa for a 13 month period following a left posterolateral thalamic stroke. She reported significantly changed attitudes to food. Clearly the pre-stroke assessment, completed retrospectively, has to be interpreted cautiously. Nevertheless, the findings strongly suggest important shifts in her attitudes. There are two possible hypotheses to account for her anorexia.
remission: (a) the cerebral infarct switched off her anorexia; or (b) the personal trauma of the stroke, including being told that she might have a brain tumour or had a stroke.

Thalamic pathways have been implicated in the control of normal eating. As part of Papez circuit, the anterior thalamus projects to the cingulate gyrus and the dorsomedial thalamus; the thalamus plays an important role in anorexia nervosa. Hyperphagia has been reported with a variety of lesions in the thalamus, hypotalamus and frontal lobe. Lesions in these areas have also been implicated in the onset of anorexia nervosa. In contrast to our patient’s remission from anorexia with a left posterolateral thalamic infarct, anorexia has been associated with dorsomedial thalamic infarction. Stereotactic thalamotomy has been used as a treatment of anorexia nervosa. The right dorsomedial and intralaminar thalamic nuclei were lesioned in one patient, while a bilateral procedure was performed in two; all three made a sustained improvement. There are other reports of improvements following an encephalitic illness and a right thalamic haemorrhage.

Trauma may contribute to the development of anorexia nervosa. However, there are no reports, to our knowledge, of a traumatic event leading directly to the cessation of an eating disorder. Our patient was traumatised by the sudden death of a 21 year old cousin from a “brain haemorrhage” when she was aged 13 years. A change in her eating pattern developed into anorexia nervosa over subsequent months. Whether this sudden and sustained recovery from an eating disorder was due to a psychosomatic traumatic event or to the direct effect of the left thalamic stroke is not certain. The abruptness of the change, and the unusual thalamic abnormalities in anorectics that reverse in remission, lends weight to the latter hypothesis.

Is the rapid assessment stroke clinic rapid enough in assessing transient ischaemic attack and minor stroke?

Our rapid assessment stroke clinic (RASC) was established as part of a single point of access for general practitioners to refer patients with suspected transient cerebral or ocular ischaemic attacks (TIA) or recovered non-hospitalised stroke in response to the publication of the National Clinical Guidelines for Stroke and the National Service Framework for Older People.5 Similar rapid access neurovascular clinics have been set up throughout the United Kingdom to provide readily available access to primary care for the management of similar patients. These clinics have significant revenue costs for the NHS, and hence the importance of reviewing their process and outcome. We now report the fate of non-attendees to highlight the risk of early stroke.

Between October 2000 and December 2002, 1460 patients were referred to the RASC. When a referral (usually by phone or fax) is received, the patient is contacted by phone to arrange a convenient appointment, or by post if not contactable by phone. Those who fail to attend the clinic on the first appointment are given a second appointment to attend, and all patients were prospectively registered in the single point of attendance. The medical notes of the non-attendees were reviewed to determine the reason for the non-attendance. If there was no relevant record in the medical notes, the general practitioners or the patients themselves were contacted by phone. Death certificates were reviewed where appropriate. Any relevant imaging, including computed tomography of the head or carotid Doppler ultrasound, was also reviewed.

In all, 1460 patients were referred during the 27 months study period and 121 failed to attend in spite of being sent two appointment. The median waiting time from referral to appointment was 17 days (range 0 to 96); 47.6% of patients were seen within two weeks of referral. The mean age of the non-attendees was 71 years (29 to 93); 44 were male and 77 female. Risk factors for TIA or minor stroke included cerebral infarction (5.7%), hypertension (14%), diabetes (5.7%), and hypercholesterolaemia (11.5%). Reasons for non-attendance included 39 (32%) who had a stroke requiring admission to hospital, of which 27 (69%) occurred during the first three days after referral (fig 1). Thirteen of the 39 strokes (33%) were fatal. CT showed evidence of infarction in 31 (80%), intracranial haemorrhage in 2 (5%), and was normal or not undertaken in 6 (15%). Seventeen patients (44%) had carotid Doppler ultrasound, and of these, four had stenosis exceeding 50%.

Patients with TIA are at significant short term risk of stroke, previously reported as ranging from 4% to 8% in the first month.1 Therefore the National Service Framework and National Clinical Guidelines for Stroke recommended the setting up of rapid access neurovascular clinics in which patients should be seen within 14 days of referral. In a study of patients presenting to an emergency department—almost all of whom were enrolled within 24 hours of the TIA—the reported stroke risk was 5% in the first two days.4 The Oxfordshire community stroke project (OCSP) prospectively followed a population of patients presenting to their primary care provider with a TIA or completed stroke reported a 4.4% risk of stroke in the first month following a recent TIA.1 In the subsequent OCSP study, which redefined the index event, the authors suggested that the risk was much higher than the initial estimate, lying at 8.6% and 12.0% at seven and 30 days, respectively.4 In our present review, 32% of patients who failed to attend the clinic did so because they had a stroke requiring hospital admission in the interval between seeing their general practitioners and the clinic appointment; 27 occurred in the first three days after referral, suggesting that the recommendation in the National Clinical Guidelines for Stroke about the timing of referrals to neurovascular clinics may need revision.

Our study may be criticised on the grounds that information about the timing of the index event was not included; however, we regard this report as being a pragmatic view of what is happening in reality. A rapid access neurovascular service is unlikely to be effective in preventing stroke unless patients can be seen and treated on the same day that they present. This study highlights the need for urgent evaluation and treatment of those at risk of stroke, ideally on the same day as the index event. Studies are required to determine the most effective intervention. The challenge for stroke physicians is to test the effects of combination treatment comprising combined antiplatelet therapy, a cholesterol lowering drug, and blood pressure lowering.

References


Figure 1 Time interval between referral and stroke occurrence.
An unusual cause of dysphagia

We report a case of a patient with Lambert Eaton Myasthenic Syndrome (LEMS) associated with small cell lung cancer (SCLC) presenting with a 2 year history of dysphagia. This presentation has not been described in previous literature.

Case

A 71 year old right handed deck worker initially presented with a 6 month history of dysphagia, weight loss, nausea, and fatigue. He was cachectic with no other clinical signs to be found on examination. Baseline blood tests were unremarkable. Upper gastrointestinal (GI) endoscopy revealed a small hiatal hernia only. Subsequent investigations included a barium swallow, computer tomography (CT) scans of the chest and abdomen, oesophageal manometry, small bowel follow through, and laparoscopy, which were all normal. He was subsequently found to have LEMS.

LEMS usually precedes the cancer diagnosis by many years and is more common in men than women. The mean age at diagnosis is 50 years, with presentations ranging from 15-70 years old. The median age at diagnosis of the underlying malignancy is 60 years. It is estimated that 10-15% of patients with LEMS have a neoplasm.

Dysphagia as the sole presenting symptom of LEMS is extremely rare however. This usually develops late in the course of the disease and is not present at the onset. Dysphagia as the sole presenting symptom of LEMS is extremely rare however. Proximal lower limb girdle weakness is the most frequent presentation. In a case series of 50 consecutive patients, leg weakness was the presenting complaint in 62% of patients.

Further investigations revealed adenocarcinoma of the lung with metastases. The dysphagia resolved completely. Repeat CT staging after completion of his treatment demonstrated a complete response.

Discussion

Dysphagia occurs in 24–34% of patients with LEMS. This usually develops late in the course of the disease and is not present at the onset. Dysphagia as the sole presenting symptom of LEMS is extremely rare however. Proximal lower limb girdle weakness is the most frequent presentation. In a case series of 50 consecutive patients, leg weakness was the presenting complaint in 62% of patients. Less frequent presentations are generalised weakness, achin and stiffness, autonomic symptoms (impotence, dry mouth, constipation), arm weakness, diplia, and dysarthria.

Guruprakash et al. reported a case of a 59 year old man presenting with dysphagia who was subsequently found to have LEMS. Further investigation revealed adenocarcinoma of the lung with metastases. The dysphagia resolved completely with a combination of preoperative guanidine hydrochloride, followed by bilateral orchiectomy and chemotherapy. This resulted in resolution of the dysphagia and some improvement in the chorea. The role of IVIg in treating LEMS is established. It is also recognised that treatment of the underlying tumour can result in improvement or remission of symptoms relating to the paraneoplastic syndrome.

This case illustrates that LEMS is an unusual but important cause of swallowing difficulties. In the present patient, establishing the cause of dysphagia led to the diagnosis and treatment of the underlying SCLC.

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References


Traumatic brain injury and haemorrhagic complications after intracranial pressure monitoring

Intracranial pressure (ICP) monitoring is now a widely accepted tool in the management of patients with head injuries. However, intracranial haemorrhage is recognised as a possible complication following placement of an ICP device. The purpose of this study was to investigate the incidence of haemorrhage after ICP monitor insertion through a thorough review of post-insertion computed tomography scans, and to classify them in a clinically relevant manner.

Materials and methods

Patients

Over 5 months, the Neurosurgery Service at Harborside Medical Center treated 314 patients with traumatic head injury. There were 247 males and 67 females with a median (SD) age of 35.16 (22.9) years (range 0.4 to 102 years), and all were admitted to the hospital. Placement of an ICP monitor (CaminoTM, intraparenchymal) was undertaken in 130 of these patients. We retrospectively analysed the patient’s hospital charts and all available radiological studies, with particular attention paid to our own interpretation of CT scans before and after ICP monitor insertion. The final numbers in the study were 101 males and 29 females with a median (SD) age of 36.6 (21.9) years (range 1.8 to 102 years).

ICP monitoring

Indications for ICP monitoring followed the head injuries (ICP monitoring) guidelines: (a) patients with severe head injury, GCS ≤ 8 with an abnormal head CT; (b) patients with severe head injury, GCS ≤ 8 with a normal head CT, and having two or more of the following: age ≥ 40 years, systolic blood pressure < 90; or posturing; (c) patients with GCS 9–12 and abnormal head CT, if undergoing therapies for other injuries with possible deleterious effects on ICP; and (d) subsequent to removal of intracranial mass.

The fibre optic device was placed at the bedside (intensive care unit, emergency room) or at completion of the surgery in the operating theatre. Some patients needed replacement of an ICP device because of technical problems with the device. The right side was preferred for the insertion of the ICP monitor.

CT scanning

The institutional protocol was to perform CT scanning during the first 24 hours after the insertion of ICP device. We were not able to obtain CT in this time frame in four cases. A grading system for haemorrhages after ICP monitor insertion from our institution was used (fig 1). Grade 0 was used to report patients with no complications on post-placement studies. Grade 1 is a small punctuate haemorrhage or localised subarachnoid haemorrhage (SAH). Grade 2 haemorrhage is an intracranial bleed, diffuse SAH or extra-axial haematoma without a new neurological deficit and does not require operative intervention. In a case of grade 3 complication, revision craniotomy is required or there is a new neurological deficit, even a death.

Results

Of the 314 patients with traumatic head injury, ICP monitor insertion was performed in 130 (41%). Nineteen patients had more than one ICP monitor inserted; altogether, 155 procedures were carried out. Right sided procedures prevailed (n = 102, 66%). The majority of the patients in this study were admitted with the diagnosis of a closed head injury (n = 116, 89.2%), 10 patients had open head injury, and four suffered a gunshot wound to the head. One hundred and six procedures (68%) were performed at the bedside, and 49 insertions (32%) took place in the operating theatre.

We retrospectively analysed the patient’s hospital charts and all available radiological studies. There were 140 procedures performed without any haemorrhagic complications on follow up radiological studies (grade 0). After 10 insertions (6.5%), a small punctuate haemorrhage or localised subarachnoid haemorrhage occurred. These complications were classified as grade 1 haemorrhages. Five patients (3.2%) sustained an intracerebral haematoma that did not necessitate evacuation or manifest as a new neurological deficit (grade 2). There were no haemorrhagic complications requiring evacuation or resulting in a noticeable change in the patient’s clinical condition (grade 3). Altogether, the complication rate was 9.7% for this study. More haemorrhagic complications occurred after ICP monitor placement in the operating theatre (n = 8/49, 16.3%), compare the bedside procedures (n = 7/106, 6.6%). This distribution did not reach statistical significance (p = 0.057).

Conclusions

There is a wide range (0–15.3%) in the literature of reported incidences of intracranial haemorrhages following placement of an ICP monitoring device. However, most studies have multiple targets such as outcome, different treatment options or indication criteria and these published reports failed to distinguish between large haematomas requiring surgical evacuation and small punctuate haemorrhages picked up incidentally only on imaging. In head trauma, there are multiple lesions on radiological examinations, and without detailed knowledge of the patient’s surgical procedures, a punctuate haemorrhage can be counted as an evolving contusion or go unnoticed. Due to previous metal artefacts from tip of the ICP monitor catheter, some small lesions were detected only after its removal.

In our traumatic brain injury group, we found a complication rate of 9.7% with no grade 3 haemorrhage. Although the most common grade 1 haemorrhage seems to be unimportant, we do not know its long term consequences and it may cause a false reading of a high ICP with subsequent unnecessary therapeutic interventions. The incidence of grade 3 haemorrhage was 0.15% (1 in 684 procedures) in our institution for the general neurosurgical population (trauma, tumors, cerebrovascular), and there is a similar complication rate and stratification for the paediatric subpopulation.

Although intracranial pressure monitoring plays an indispensable role in the management of head injuries, the indications for this invasive neurosurgical procedure should always be carefully considered. Even with the utmost precautions, haemorrhagic complications may occur. Classification of the complications in the clinically relevant scheme may help to compare results of future studies.

M Blaha, D Lazar

Department of Neurological Surgery, University of Washington School of Medicine, Seattle, Washington, USA

Correspondence to: Dr M Blaha, Neurosurgery Department, Central Military Hospital, U Vojenské nemocnice 1200, Prague, Stetovice 169 02, Czech Republic; drmartinblaha@yahoo.com

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Critical care neurology and neurosurgery

Edited by Jose I Suarez. Totowa: Published by Humana Press, 2004, $145.00 (hardback), pp 611. ISBN 1-58829-089-1

This is a worthy attempt to produce a comprehensive multi-author text of neurological and neurosurgical ITU. The volume is extensive with more than 60 authors contributing 34 chapters in over 600 pages. The scope is wide ranging and covers a broad sweep of topics relating to critical illness due to primary neurological and neurosurgical conditions. It has little or nothing to say about the neurological complications of general medical intensive care.

There are many excellent individual chapters—I learnt a great deal from the neurological contributions concerning raised intracranial pressure and monitoring, and also the surgical section on vascular disease. However, there are surprising omissions; for example, as general neurologists we are probably asked to consult about ischaemic-hypoxic brain injury more than any other single condition and yet this book has little concerning this important topic. There is a relatively little about the practical aspects of management although there are honourable exceptions and I particularly enjoyed the section on ventilation and tracheostomy. A more up-to-date description of central respiratory abnormalities due to neurological disorders would have been preferable. Does anybody really see the patterns of central herniation described by Plum? In the modern world ventilation is introduced much earlier and these descriptions are generally of historic interest only. For such a comprehensive text I would have preferred a little more about the history and philosophy of neurological intensive care—particularly a recognition of the different sorts of units related to stroke and long term ventilatory management.

This book attempts to present an overview of the subject, including chapters on most aspects of neurological critical care, but unfortunately the structure is rather loose and the content is organised without an obvious overall strategy. This is a disappointment and rather dilutes the value of this book as a textbook. It is relatively expensive and I was disappointed by the poor quality of the illustrations. It is surely essential in the modern world of neuro-imaging within the ITU to be presented with high quality reproduction of functional imaging in addition to more conventional modalities. Similarly, the lack of structure demands a more coherent presentation of individual chapters making better use of tables and figures.

There are several excellent new texts of critical care neurology and neurosurgery against which this book must be measured. The lack of structure and organisation means that it falls short of the more coherent books written by single authors or small groups. The book will remain an asset to specialist neurocritical care units but is unlikely to be of value to general neurologists or trainees.

R Howard

Neurology of the arts: painting, music, literature

Edited by F Clifford Rose. Published by Imperial College Press, 2004, £65.00 (hardback), pp 432. ISBN 1-86094-368-3

Neurology of the arts: painting, music, literature is a multi-authored book that explores the intersection between neurology and the arts. The topics in the book are wide ranging, moving from discussions of Dostoyevsky and epilepsy, to amusia, back to Samuel Johnson and Mozart’s movement disorders. Neurology is the underlying glue that binds the book. The chapters are really quite diverse and touch on the use of literature or painting to portray neurological disorders, include descriptions of the neurological conditions of famous artists, writers or musicians, or delineate the neurological basis for music and painting. Many of the authors have a background in neurology or neuroscience, but there are fascinating contributions from Professors of music, literature, and art.

The editor offers an erudite chapter on the representation of neurology in art, beginning with an ancient tablet from Egypt illustrating a person with an atrophic leg, suggestive of polio. Also, he explores migraine as a possible source for artistic creativity in Hildegard of Bingen and outlines the influence of neuroanatomy upon artists like Leonardo da Vinci, Theodore Gericault, and Rembrandt van Rijn. Finally, he summarises the panoply of diseases from which Van Gogh may have suffered. It is a book beautifully illustrated to the various artists who have suffered from epilepsy. In other chapters the art of Sir Charles Bell and the poetry of Henry Head are described.

There are two exceptionally strong chapters on the cerebral localisation of music. In one the neuroanatomy of music perception and musical memory is described while another summarises research into the neural basis for music in musicians and non-musicians. In this vein, another chapter describes amusia—a rare but intensely studied cognitive disorder. The effect of Mozart on epilepsy (protective), and the relationship of music and madness provides interesting contrasts on music’s effects on behaviour.

For readers with background in neurology with a special interest in literature there is much to enjoy. Christopher Goetz—a leading medical historian—notes the influence of Shakespeare on Charcot’s teaching. The astute observations by Shakespeare on various neurological conditions once used by Charcot as a teaching tool offer remarkable insights into both Shakespeare and Charcot. Joyce’s use of medical metaphors in Ulysses and the neurological basis for music and painting. Many of the authors have a background in neurology or neuroscience, but there are fascinating contributions from Professors of music, literature, and art.

Fifty neurologic cases from mayo clinic


A book of 218 pages, which starts with semantic dementia and ends with mild cognitive impairment of amnestic type via Tangier disease, necessarily lays the emphasis on the esoteric rather than the mundane. Whether the subject matter will be of interest to “surgeons...and of particular help to medical students” is a matter for others to judge. However, it seems to this reviewer, that the average surgeon will not have any particular desire to be entertained with Whipple’s disease, Angelman’s syndrome, or Erdheim-Chester disease. The agendas of publishers and clinicians do not always coincide and it would be churlish to shoot the messenger when Professor Noseworthy’s work contains much else to savour—especially the preface and acknowledgements!

Neurologists tend to be competitive individuals and this book certainly lays down the gauntlet. The format is tried and tested, with the history, examination findings, and results of investigations inviting the reader to predict the denouement, which is presented overleaf, together with a commentary by an expert in the field. The quality of the illustrations is first class. The range of cases presented is mind boggling and the commentaries extremely well researched and up to date.

I have only a few minor criticisms. Many of the commentaries contain little mention of the differential diagnosis. It seems churlish to present a case of facioscapulohumeral muscular dystrophy (FSH) without facial involvement or mention of Beevor’s sign and expect the average reader to hit the nail on the head. I am unconvinced by the argument that asking our patients to wiggle their ears is ever likely to lead to a fruitful outcome. It might have been helpful to include the normal ranges alongside the results of tests.

At £24.95 this seems good value for money and in my opinion will enhance any departmental or personal library.

A J Wills

STROKE—pathophysiology, diagnosis, and management, 4th edition


This is a major update—by a new editorial team—a of major reference book in cerebrovascular diseases. Updating such a big reference work is a herculean task, but the large the editors have succeeded in their task. They have assembled many very distinguished authors and put together a pretty...
introduction:
The editors themselves recognise this in their
have long gestations and I suspect that many
fruition. Unfortunately such large beasts
must be congratulated in bringing it to
huge challenge and the authors and editors
methodical reader who wishes to assess the
detail what the overall message should be.
So, this book will appeal to the thoughtful,
detail, mentioning relevant systematic
fits—comes from randomised controlled
treatment—balancing its harms and bene-
I mean that the best evidence on the choice of
have more arrows and annotations to point
out exactly what the abnormality was. It
would be preferable to replace the unsub-
tracted angiograms with subtracted films.
The chapters on management and treat-
ment are comprehensive, and cover all of the
options that are currently in use. Thus, one
can find useful material on everything from the
treatment of hyperacute ischaemic stroke,
to the use of neuro-interventional treatments
for arteriovenous malformations (AVMs),
arterial dissections, and vascular stenosis in the
blood supply to the brain. Thus there is
good coverage of what can be done but it is
not always clear what should be done. By this
I mean that the best evidence on the choice of
treatment—balancing its harms and bene-
fits—comes from randomised controlled trials and systematic reviews of such trials.
In general, the authors in the book have
tended to discuss each trial in considerable
detail, mentioning relevant systematic reviews relatively infrequently and it is often
doing difficult to extract from this mass of
detail what the overall message should be.
So, this book will appeal to the thoughtful,
methodical reader who wishes to assess the
evidence in very great detail but the “reader
in a hurry” may find it difficult to locate the
text that gives the final verdict on a particular treatment.
Producing reference works of this size is a
huge challenge and the authors and editors
must be congratulated in bringing it to
fruition. Unfortunately such large beasts have
long gestations and I suspect that many of
the manuscripts were submitted some time
in 2002, to achieve a 2004 publication date.
The editors themselves recognise this in their
introduction:

“We even hope that the information
contained in this edition makes for
its rapid obsolence, so great are
our aspirations for continued rapid
development in our field”.

In many ways it is good news that the
authors’ predictions have been proven, since
for example, the International Subarach-
noid Aneurysm Trial (ISAT)—published in
2002—has clearly shown that for patients
with ruptured intracranial aneurysms, coiling
with detachable platinum coils leads to a
substantial reduction in death or disability,
compared with conventional neurosurgical
clipping. It appeared too late to be included in
the book. The pace of change in change in
stroke is indeed rapid!
So, this fourth edition is a welcome update
to a well known reference book. The dense
text and heavy referencing (e.g. the 50 page
chapter on intracerebral haemorrhage has
390 references), mean that it is a mine of
information, but in the very nature of such
books, not always light reading.

P Sandercock

The treatment of epilepsy

Edited by Simon Sharvon, David Fish, W Edwin
Dodson, Emilio Perucca. Published by Blackwell
Publishing Ltd, 2004, £150.00 (hardcover),
pp 952. ISBN 0-63206-046-8

This is a text that ought to be read by all physicians who treat people with epilepsy. It
may appear dauntingly large on first acquain-
tance, but it is well written, full of practical
advice, and gives the reader helpful details about the drugs that most of us use on a daily
basis.
The first section contains chapters on the
clinical and epidemiological aspects of epi-
lepsy and the clinical pharmacology of anti-
epileptic drugs. The second section is on the
management of epilepsy, including: newly
diagnosed epilepsy, status epilepticus, epi-
lepsy in remission, reproductive aspects of
epilepsy, and the management of special
groups such as learning disabled people. The
third section is devoted to individual anti-
epileptic drugs introduced by a thoughtful
assessment of the evidence upon which we
have to make choices of antiepileptic therapy and practical advice on the changing of
antiepileptic drugs. The final section is about
epilepsy surgery with details of the neces-
sary investigations, assessments, and surgical
procedures.

If this all sounds like too much detail for
the general neurologist I would beg to
disagree. You may not want to read the
introductory chapter on historical aspects of
the treatment of epilepsy (but you will miss out
on a fascinating account of drug develop-
ment if you don’t) or you may feel you do not
need to read the chapter on mechanisms of
antiepileptic drug action (though you would
be wise to do so for this is one of the best
chapters on the subject that you could hope
to find), but the sections on the principles of
medical treatment and antiepileptic drugs
should be of interest to all who have patients
with epilepsy.
Chapters such as that on the treatment of
epilepsy in general medical conditions will be
particularly useful to neurologists working in
hospitals with renal and liver units. The section on the individual drugs contains all
details that you need but can’t remember when you are rung up and asked about those
side effects and drug interactions (which you
should know and can’t find in the BNF). For
anyone unfamiliar with epilepsy surgery this
section is an excellent summary of the
subject.
There are only one or two weak points: as
in most multi-author texts there is some
overlap, e.g. in the description of seizure models in two adjacent chapters, and I was
puzzled why there was no chapter on the
management of the idiopathic generalised epi-
lepsies as there was in the first edition. It
would be useful to have had more practical
advice for special circumstances such as
foreign travel and the management of
patients unable to take oral medication, but
these are minor quibbles. My advice would
be: get a copy and keep it by your desk at
work, you won’t regret it.

M Jackson

Disorders of the brain and mind 2

Edited by Maria A Ron, Trevor W Robbins.
Cambridge: Published by Cambridge University

This is the second book in the series “Disorders of brain and mind” edited by neuropsychiatrist
Maria Ron. Her co-editor for this book is the
neuroscientist and cognitive psychologist
Trevor Robbins. Together they have produced
a formidable compilation of articles written by
leaders in their respective fields, them-
selves included. These describe our current
understanding of the neural basis of com-
monly encountered psychiatric disorders
including schizophrenia, mood disorder,
dementia, personality disorder, and addic-
tion. The structure of the book is a particular
strength. First, the format of grouping chapters thematically is retained so that both
basic and clinical scientific aspects of partic-
ular disorders are covered. Examples include
a chapter on how mutations in the tau gene
are central to the development of a range of
dementias, a chapter that describes how
advances in neuropsychological and neuro-
imaging research has improved early diagno-
sis and differential diagnosis of the
dementias, and a chapter on the scientific
study of consciousness coupled with one on
how this applies to an understanding of
avolition in schizophrenia. Second, by includ-
ing more generic groupings devoted to
neurodevelopment, genetics, and neuro-
imaging, the methodologies or concepts that
are currently proving to be of fundamental
importance to the advancement of know-
ledge in neuropsychiatry are also addressed.
Thus this excellent book should provide
something of interest to research workers in
the clinical neurosciences as well as clinicians
who wish to catch up or learn about aspects of
neuropsychiatry de novo.

E Joyce