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-acetylcholine 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 (radioimmunooassay in the same laboratory, normal 0–5 U/ml) and remained positive after thymectomy. 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 orbicularis 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 × 10^9 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 Woltman in 1956 and was subsequently shown to be associated with anti-GAD antibodies in 40–60% of cases and anti-ambiphysin 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.

This patient had radiological evidence 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 × 10^9 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.

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 were likely 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 histologically 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 autoimmune 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 presentation. Thymoma should be investigated for.

Internal jugular vein thrombosis associated with shiatsu massage of the neck

Thrombosis of the internal jugular vein is a relatively rare condition that can be induced by a variety of mechanical injuries,1,2 Acupressure, or “shiatsu”, is an oriental massage technique and many acupoints on the body surface, known as “tsubos”, are used for shiatsu. Shiatsu of tsubos in the nape of the neck is known to improve tension headache due to neck and shoulder aches. However, we recently came across a case of internal jugular vein (IJV) and cerebral sinus thrombosis after shiatsu massage of the neck.

Case report

A 35 year old man, a non-smoker, was suffering from a stiff neck. He consulted a shiatsu masseur, who performed shiatsu massage on the right side of his neck and right shoulder for 30 minutes. Immediately after the shiatsu massage, the patient noticed pain and swelling of the right side of the neck, both of which subsided within seven days. Two days after the shiatsu massage, he developed a severe, constant right occipital headache and consulted his attending physician. His cervical radiograph was normal. The patient continued to have severe headache, however, and on the seventh day after the massage, he developed blurred vision. On the twentieth day, he developed weakness and paraesthesia of his right arm and leg, and mild agraphia for kanji characters. When he also developed focal motor seizure, he was admitted to our hospital. He underwent a neurological examination on the twenty third day after the shiatsu massage.

The patient did not have any history of recent trauma, dental procedures, or upper respiratory infection. There was no history of any other relevant medication including homeopathic or herbal medicines, or pathologic conditions. There was no family history of premature stroke or thrombotic events.

Physical examination was normal and no neck mass was detected. On neurological examination, he showed normal consciousness and orientation. Funduscopic examination revealed bilateral papilloedema without haemorrhage, but the remaining cranial nerves were intact. He had mild muscle weakness and sensory deficit in the right arm and leg. Ataxia was not detected in any of the limbs and trunk. Mild agraphia for kanji characters was observed.

Laboratory analysis showed prothrombin time, partial thromboplastin time, antithrombin III, protein C, and protein S were normal, but values for anticardiolipin antibody IgG and lupus anticoagulant were negative. Plasma homocysteine was within normal limits. Autoantibodies and cryoglobulins were absent. No evidence of any systemic disease was found on investigation.

References


Figure 1 Top panel: post enhancement T1-weighted magnetic resonance (MR) image of the head (A) axial, (B) coronal, and (C) sagittal. (A) and (B) show the left parietal haemorrhagic infarct. The superior sagittal sinus and right transverse sinus show high intensity signal within the lumen instead of the normal “flow void”, indicating thrombosis. Middle panel: MR image of the neck (A) T1-weighted, (B) T2-weighted, (C) post enhancement T1-weighted, and (D) coronal T2-weighted showing right internal jugular vein thrombosis without other structural abnormalities (arrows). Bottom panel: digital subtraction angiogram (A) lateral view of the head during the early venous phase of right carotid digital subtraction angiography confirms the non-opacification of the superior sagittal sinus, the deep cerebral venous system and the transverse sinuses. The predominant venous drainage is via the sphenoparietal sinus (arrow). (B) Anteroposterior view of the neck—the right jugular vein had an area of obstruction at its junction with the right subclavian vein.
Cerebrospinal fluid was clear without pleocytosis, but the cerebrospinal fluid pressure was 350 mm H₂O.

Magnetic resonance imaging (MRI) scan of the brain showed infarction with haemorrhage in the left parietal lobe and an area of increased signal intensity in the area of the right transverse and superior sagittal sinuses (fig 1). In addition, 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 (fig 1). The rest of the intracranial 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 intracranial hypertension was treated with a lumboperitoneal shunt. The headache and papilloedema slowly improved over the next three weeks, after which the patient was discharged. Neurological examinations over the past several months have revealed only mild clumsiness and paraesthesia of his right hand and leg.

Discussion

Our patient started complaining of a swelling and pain in the right side of the neck immediately after the shiatsu massage of the neck. Subsequently, over a period of about a month, he developed progressive headache, right extremity paralysis, papilloedema, and decreased pain in his neck, and the temporal relation was also given heparin and warfarin and the intracranial hypertension was treated with a lumboperitoneal shunt. The headache and papilloedema slowly improved over the next three weeks, after which the patient was discharged. Neurological examinations over the past several months have revealed only mild clumsiness and paraesthesia of his right hand and leg.

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Although congenital dumbbell neuroblastosomas are rare, spinal cord compression and involvement of the peripheral nerves or the nerve plexus are not uncommon with abdominal neuroblastoma. The incidence of intraspinal involvement of neuroblastoma varies between 6% and 24%. Intraspinal neuroblastoma is a direct extension of a peripheral tumour. Dumbbell neuroblastoma is the commonest malignant cause of spinal or nerve root compression in young children and is regarded as an unresectable tumour. Chemotherapy should be considered for patients with partial deficits and surgical decompression should be reserved for children with recent onset of severe neurological dysfunction or deterioration in a 24–72 hour period. We treated case 1 with chemotherapy only, because his neurological deficit had been observed four months ago and was not progressive. After disappearance of the retroperitoneal and intraspinal masses with chemotherapy the deficit improved partially. The second neonate's condition deteriorated during chemotherapy, and surgical decompression resulted in recovery of one limb although the fixed deficit from birth, in the lower right extremity, did not change.

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 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 computerised 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 planter response.

Her brain CT demonstrated left thalamic hypopituation, 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. No other investigations (chest x ray, electrocardiogram, thoracic echocardiogram, full blood profile, thrombophilia screen, glucose, liver function tests, and thyroid function tests) were normal.

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 had a stroke.

Thalamic pathways have been implicated in the control of normal eating. As part of Papez circuit, the anterior thalamic projects to the cingulate gyrus and the dorsomedial thalamic projects to the basal nuclei of Meynert. The thalamus may have an integrative role with higher order somatosensory and visuospatial function. Neuroimaging techniques have demonstrated several abnormalities in the anorectic state, some of which are reversible with treatment. The smaller size of the thalamus and thalamic perfusion changes in anorexics suggest that the thalamus plays an important role in anorexia nervosa. Our patient demonstrated the usual clinical features of lateral thalamic infarction: hemiataxia, and hemisensory and motor deficit.

Hyperphagia has been reported with a variety of lesions in the thalamus, hypothalamus 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 posterior-lateral 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 psychologically traumatic event or to the direct effect of the left thalamic stroke is not certain. The abruptness of the change, and the post-traumatic thalamic abnormalities in anorectics that reverse in remission, lends weight to the latter hypothesis.

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Figure 1 Time interval between referral and stroke occurrence.

Table 1

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<th>Interval between referral and stroke (days)</th>
<th>No of patients</th>
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<tr>
<td>&lt;1</td>
<td>8</td>
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<td>1–7</td>
<td>3</td>
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<td>8–14</td>
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(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. 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. 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. 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.

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 thalamus plays an important role in the control of normal eating. As part of Papez circuit, the anterior thalamic projects to the cingulate gyrus and the dorsomedial thalamic projects to the basal nuclei of Meynert. The thalamus may have an integrative role with higher order somatosensory and visuospatial function. Neuroimaging techniques have demonstrated several abnormalities in the anorectic state, some of which are reversible with treatment. The smaller size of the thalamus and thalamic perfusion changes in anorexics suggest that the thalamus plays an important role in anorexia nervosa. Our patient demonstrated the usual clinical features of lateral thalamic infarction: hemiataxia, and hemisensory and motor deficit.
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 cachetic 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 empirically started on cisapride and managed on an appropriate pureed diet.

Eight months after his initial presentation the patient complained of a gradual deterioration in his speech and problems with proximal muscle weakness. Clinical examination demonstrated an obvious dysarthria and broad based gait. There was no limb weakness or sensory neuropathy and reflexes were preserved with no post-tetanic potentiation. There was no disturbance of extraocular eye movements or ptosis. The patient did not display any cognitive impairment. He subsequently developed involuntary choreoathetoid movements.

Baseline blood tests and chest x ray (CXR) were again normal. Magnetic resonance imaging of the brain demonstrated evidence of small vessel disease. Electromyography and nerve conduction studies were performed. Sensory conduction velocities were within normal limits. Motor conduction velocities demonstrated a clear and reproducible decremental response to repetitive stimulation in the right ulnar nerve (32%) and in the right median nerve (25%), maximal at 3 Hz stimulation. Compound action potentials in the muscles tested increased by more than 50% following a period of exercise (right ulnar 0.9 to 2.8 mV, right median 1.7 to 2.8 mV). Electromyogram (EMG) of the right biceps and 1st dorsal interosseous muscles were normal. Overall the results were felt to be consistent with a diagnosis of Lambert- Eaton Myasthenic Syndrome (LEMS). The presence of anti-voltage gated calcium channel antibodies confirmed the diagnosis.

Repeat CT scanning in combination with positron emission tomography (PET) scanning revealed enlarged subcarinal lymph nodes only. A transthoracic lymph node biopsy confirmed the diagnosis of small cell lung cancer. Anti-Hu antibodies were found to be positive.

The patient initially received a course of intravenous immunoglobulins (IVIg) (1 g/kg for 5 days), which resulted in an improvement in speech, swallowing, and gait. He then proceeded to start treatment with cisplatin/etoposide chemotherapy and concurrent radiotherapy. His movement disorder improved slightly during this time but his 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, but may be 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, aching and stiffness, autonomic symptoms (impotence, dry mouth, constipation), arm weakness, diplopia, and dysarthria.

Guruprakash et al reported a case of a 59 year old man presenting with dysphagia who was subsequently found to have LEMS. This usually develops late in the course of the disease, but may be 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, aching and stiffness, autonomic symptoms (impotence, dry mouth, constipation), arm weakness, diplopia, 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 rectum with bony metastases. The dysphagia resolved completely with a combination of preoperative guanidine (1 g/kg for 5 days), which resulted in an improvement in speech, swallowing, and gait. He then proceeded to start treatment with cisplatin/etoposide chemotherapy and concurrent radiotherapy. His movement disorder improved slightly during this time but his dysphagia resolved completely. Repeat CT staging after completion of his treatment demonstrated a complete response.

References


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Traumatic brain injury and hemorrhagic 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 a recognised as a possible complication following placement of an ICP device.1,2 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 Harborview Medical Center treated 314 patients with traumatic head injury. There were 247 males and 67 females with a median (SD) age of 31.6 (22.9) years (range 0.4 to 102 years), and all were admitted to the hospital. Placement of an ICP monitor (Camino™, intraparenchymal) was undertaken in 160 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:3 (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.1,2 However, most studies had 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 paediatric neurosurgical population (trauma, tumors, cerebrovascular),4 and there is a similar complication rate and stratification for the paediatric population.

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.

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References


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 neurological 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 against which this book must be measured. The lack of structure and organisation means that it falls short of the more coherent books against which this book must be measured.

The editor offers an erudite chapter on the representation of neurological history in art, beginning with a tablet from ancient Egypt illustrating a person with an atrophic leg, suggestive of polio. Also, he explores migraine as a possible source for artistic creativity in Hildegarde de Bingens and outlines the influence of neuroanatomy upon artists like Leonardo da Vinci, Theodore Gericault, and Rembrandt van Rijn. Finally, he summarises the neuropathology of diseases from which Van Gogh may have suffered. In a full chapter dedicated 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 muscle. In one the neuroanatomy of muscle 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 provide 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 other works elucidates a unique perspective on this author’s work. Two chapters address Dostoyevsky; one depicts his use of epilepsy in writing, the other discusses the potential aetiology for his epilepsy. A full chapter is dedicated 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.

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 exposed to 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 overhead, 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 analyst to hit the nail on the head. I am convinced 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.

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.

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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—of a major reference book in cerebrovascular diseases. Updating such a big reference book is a huge task, and by and large the editors have succeeded in their task. They have assembled many very distinguished authors and put together a pretty number of individuals with a strong interest in both science and the arts. This book understands the desire of our community for this information.

B L Miller
comprehensive reference work. The early chapters on epidemiology, clinical manifesta-
tions, and underlying causes provide a very broad 
and detailed coverage. There is even a 
chapter devoted solely to infarcts of the 
anterior choroidal artery! The chapters are well 
written and heavily referenced. There are 
plenty of illustrations (the chapter by Bousser 
on cerebral venous disease has particularly 
good illustrations that are clearly annotated).

However, in some of the other early 
chapters the selected scans and angiograms 
have not been done on state-of-the-art technology. I accept that of course to find a 
scan to illustrate a particular and unusual 
problem is not always easy (particularly not a 
recent one!). In view of this, it would have 
been preferable for some of the figures to 
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), 
artrial 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 
quite 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 obsolescence, 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, colling 
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 Shorvon, David Fish, W Edwin 
Dodson, Emilco 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 
modes in two adjacent chapters, and I was 
puzzled why there was no chapter on the 
treatment of the idiopathic generalised epi-
lepsies as there was in the first edition. It 
would be useful to have had more practical 
advise 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 addiction. 
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 parti-
cular 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 diagnos-
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