Treatment of paroxysmal sympathetic storm with labetalol

First described by Penfield in 1929, paroxysmal sympathetic storm is characterised by episodic hyperhidrosis, hypertension, hyperthermia, tachypnoea, tachycardia, and posturing. It has commonly been associated with closed head traumatic brain injury, agenesis of the corpus callosum, hydrocephalus, and suprasellar or diencephalic tumours. Penfield hypothesised that these sympathetic spells were caused by epileptiform discharges in thalamic nuclei irritated by increased intracranial pressure, thereby leading him to name this entity “diencephalic autonomic seizures”. Electroencephalograms obtained on patients during these autonomic attacks, however, have not shown epileptic activity, and anticonvulsant therapies have not proved useful in their treatment. Bromocriptine and morphine have been the standard treatments for paroxysmal sympathetic storm, and propranolol has been shown to reduce the hyperpyrexia seen during autonomic spells. In this case report, we describe a patient treated successfully with labetalol, but not metoprolol, suggesting that β1 antagonism alone is not sufficient to suppress paroxysmal sympathetic storm. A 21 year old white man was an unrestrained passenger in a motor vehicle accident and developed a closed head shear injury. He was admitted to a hospital where a head CT showed hydrocephalus necessitating a ventriculoperitoneal shunt placement. A head MRI showed abnormal T2 signal in the corpus callosum and the dorsal midbrain consistent with shear injury. Although initially comatose, he improved to near baseline over the next few months. He was admitted to our hospital 15 months later with a shunt infection, necessitating treatment with vancomycin, shunt externalisation, and, eventually, replacement. During and after resolution of the shunt complications, he developed episodes of sympathetic hyperactivity while under continuous monitoring in an intensive care unit. These attacks were characterised by (1) diaphoresis throughout the body, (2) tachycardia (heart rate 140–160 bpm) measured by automated pulse oximetry or by ECG, (3) hypertension (blood pressure 170–180/100 mm Hg) measured by arterial line pressure transducers or by sphygmomanometer, (4) fevers to 39.1 °C orally, and (5) flexor posturing. He was alert during these episodes, and responded to questions appropriately with denial of any acute onset of discomfort or pain. Furthermore, these attacks were not correlated with periods of bladder distension (Foley catheter in place) or impaction (radiograph not suggestive of retained stool). Individual episodes lasted 5–10 minutes and recurred at 5–10 minute intervals. Clusters of these spells would last 1 to 2 hours with more than three clusters a day. Multiple CSF and blood cultures were negative. Serial head CT showed marked reduction of hydrocephalus and no brain stem abnormalities after shunt correction. No other intracranial pathology was noted. Plain films and MRI of the spine showed no myelopathic findings suggesting autonomic dysreflexia. Abdominal and pelvic CT did not show any hidden masses or lesions. Toxicology screen at onset of symptoms was negative. Electroencephalograms obtained during these episodes of dysautonomia disclosed theta and delta slowing with some sharply contoured waves, but no definite ictal or interictal epileptiform activity (figure). Although initially treated successfully with bromocriptine (5 mg twice daily) and morphine (15 mg every 6 hours), he was withdrawn from morphine with a methadone taper at the request of his parents secondary to concerns over addiction. He was then started on metoprolol (25 mg thrice daily) with little effect on the frequency or severity of these spells. EEG obtained during episodes of paroxysmal sympathetic storm (tachycardia with heart rate of 120 bpm) shows predominant delta and theta waves (greater on the right than on the left) with no clear epileptiform activity, indicating that these attacks are not of seizure origin.
of the hyperautonomic episodes. Replacement with 100 mg labetalol twice daily led to reduction in the frequency of events to about one a day. Subsequent increase of the medication to 200 mg twice daily resulted in a marked decrease to less than one paroxysmal sympathetic storm over several days. At the time of discharge, the patient had returned to his prediagnosis baseline.

The current observations lend support to the prevailing view that paroxysmal sympathetic storm may represent disruption of autonomic function in the diencephalon and brainstem. Bullard has proposed that the clinical syndrome may be the result of a release phenomenon within the brainstem and a paradoxical response from loss of overlying cortical or subcortical inhibition. Recent more current studies suggest localisation to the central sympathoexcitatory regions including the paraventricular hypothalamic nucleus, lateral periaqueductal grey matter, lateral parabrachial nucleus, or rostral ventrolateral medulla. Compromised autonomic neuronal integrity centrally is not surprising in the setting of infection after traumatic brain injury. These observations can potentially be used in managing central sympathetic storm. Imidazoline agonists and specific $\alpha_2$ adrenoceptor antagonists, such as clonidine and methyldopa, have recently been shown to have inhibitory actions centrally, but not within the rostral ventrolateral medulla. These agents have so far been used in the treatment of essential hypertension, tetanus, or autonomic dysreflexia. $\beta$ Blockers such as propranolol, however, have long been the mainstay of treatment of the hypertension, tachycardia, and hyperpyrexia associated with paroxysmal sympathetic storm. This non-selective $\beta$ adrenergic antagonist acts through inhibition of peripheral catecholamine activity, and being highly lipophilic, may also exert central effects through membrane stabilisation or receptor blockade. Moreover, propranolol may reduce sustained muscle contraction. Taken together, these findings suggest that non-selective $\beta$ receptor antagonism is sufficient to inhibit the clinical manifestations of diencephalic seizures.

The present case suggests that $\beta_1$ receptor antagonism is not sufficient to treat hyperautonomia during paroxysmal sympathetic storm. This patient was initially placed on starting doses of metoprolol, a selective $\beta_1$ adrenergic antagonist, yet clinical evidence of increased cerebral vascular reserve using technetium-99m HMPAO brain SPECT side challenge. This demonstrated a decreased vascular reserve in both frontal and temporal lobes, as well as in the basal ganglia. Magnetic resonance angiography and subcortical four vessel angiography showed nearly complete obstruction of the terminal portion of each internal carotid artery and the outline of a moyamoya network (fig C and D). Staged enccephaloduroarteriovenousanagiosis was performed on the left and right sides, 1 week apart, resulting in an evident amelioration of the patient's involuntary movements.

Chorea is one of the rarer, although acknowledged, presenting features of moyamoya disease; chorea is usually observed in children. It is suggested that about 6% of patients with moyamoya disease have chorea. Other types of involuntary movements have been described in patients with moyamoya disease. Valsalva related seizures,6 episodes of carpopedal spasm,7 recurrent episodes of brief involuntary movements, and is usually caused by some asymmetric, focal brain lesion. The clinical presentation of our patient was associated with opera type singing. Singing requires both hyperventilation and the breath holding Valsalva's manoeuvre. Hyperventilation causes an increase in arterial oxygen tension, which subsequently causes vasodilatation, which, in turn, reduces blood flow. In addition, Valsalva's manoeuvre increases cerebral venous pressure, which then increases intracranial blood volume and intracranial pressure, thereby reducing the arterial perfusion pressure. Thus, in those regions of the brain and cortex that are already critically perfused, hyperventilation and Valsalva's manoeuvre can easily lead to transient ischemic attack, which is characterised by unilateral or bilateral stenosis or occlusion of the proximal portion of the carotid arteries, together with an abnormal vascular network at the base of the brain. Most childhood cases manifest with the signs and symptoms of cerebral ischaemia or infarction, whereas intracerebral haemorrhage prevails in adults.5 7 We describe here a case of moyamoya disease in a 29 year old multipa-
Late recurrence of glossopharyngeal neuralgia after IXth and partial Xth nerve rhizotomy: treatment by microvascular decompression

Glossopharyngeal neuralgia, or vagoglossopharyngeal neuralgia as some would prefer, is a rare condition, occurring with a frequency of about 1% of that of trigeminal neuralgia. Medical treatment, particularly with carbamazepine, is usually effective. A significant number of patients do, however, become refractory and go on to surgical treatment. The best established surgical treatment is rhizotomy of the glossopharyngeal and upper vagal nerve roots, which seems to be invariably effective if the diagnosis is correct although it is not without morbidity and even mortality.1 Late recurrence after such treatment, as described below, has not previously been reported and raises interesting issues of mechanism and method of treatment which are considered in this brief report.

The patient initially presented in 1988 as a 23 year old woman with typical glossopharyngeal neuralgia, experiencing severe intermittent pain in the left side of the throat, the back of the tongue, and the ear. The pain was aggravated by talking and swallowing and relieved, to some degree, by pressure on the left side of the neck. At first there was a good response to carbamazepine. When medication was stopped after several months the pain returned and was less well controlled with a further course of the drug. Neurological examination, CT, and MRI were normal. In 1988 she underwent posterior fossa cranial neurosurgery and exploration of the IXth and Xth cranial nerve roots. No lesion, in particular no vascular compression, was identified. The left IXth nerve root and the two uppermost Xth nerve rootlets were divided adjacent to the brain stem. Her postoperative course was uncomplicated and she remained entirely symptom free for over 9 years.

In 1998, now aged 33 years, she developed recurrence of her original pain which she described as essentially identical to that at the initial presentation. Again the pain responded to carbamazepine but required a high dose (1500 mg daily) which was accompanied by troubling side effects (drowsiness and dizziness). In addition she was not completely pain free. Neurological examination and further MRI were normal. In October 1998 exploration of the right IXth and Xth cranial nerve roots was carried out. The previously divided nerve roots were identified and the completeness of the initial section confirmed. There was now, however, a large, ectatic vertebral artery to which the proximal ends of the previously sectioned roots were adherent and which was distorting the remaining Xth nerve rootlets and the Xth nerve. A microvascular decompression was carried out with a Teflon patch being placed between the ecstatic artery and the normal and previously sectioned nerve roots. The procedure was without complication and the patient has remained well and entirely pain free since that time (18 months).

The first description of glossopharyngeal neuralgia is credited to Weisenberg in 1910, in a patient in whom the pain was secondary to a cerebellopontine angle tumour. The pain is characteristic although two variants have been described; an otic form with pain predominantly deep in the ear, in the external acoustic meatus, and the mastoid region and an oropharyngeal form in which the pain is experienced in the pharynx, the tonsillar area, the palate, tongue and the back of the throat. Vagal neuralgia has been described primarily as a post-traumatic problem (dental, surgical, etc.). An entity often described as glossopharyngeal neuralgia is paroxysmal palatal pain, a condition characterized by severe and sharp, painless, attacks of pain in the palate, which may spread to the pharynx. The cause of this condition is unclear although it has been noted to improve following dental treatment in some cases.

In the case we are reporting the glossopharyngeal neuralgia recurred 10 years after initial section of the IXth and Xth cranial nerve roots. The symptom was essentially identical to that before surgery and was relieved by microvascular decompression. This report describes unusual recurrence of glossopharyngeal neuralgia and raises the potential role of microvascular decompression in the treatment of glossopharyngeal neuralgia.
the soft palate, and the posterior third of the tongue. For patients refractory to medical treatment several surgical options are available including extracranial avulsion, intracranial preganglionic root section, trigeminal tractotomy, either open or percutaneous CT guided, and microvascular decompression. As mentioned at the outset, intracranial root section has been the most often employed and is generally regarded as curative. It was, however, realised early that section of the upper vagal roots is important in that some cases without the additional section were either not relieved or experienced early recurrence. More recently microvascular decompression has been employed, particularly by Jannetta with complete relief of pain in 76% and substantial improvement in a further 16% in the largest series, with a mean follow up of 48 months. As with trigeminal neuralgia, the actual incidence of presumed causative “neurovascular conflict” and, indeed, the exact mechanism of causation are as yet unresolved questions.

The particular dilemma posed by the present case had both a diagnostic and a therapeutic arm, referable in each case ultimately to mechanism. The only reported cases of recurrence after preganglionic section of the trigeminal nerve were reported by Jannetta and in a smaller series with long follow up1 there were no recurrences after preganglionic section of the IXth and upper Xth roots. Likewise, after total sensory root section via the posterior fossa (Dandy procedure) trigeminal neuralgia does not recur.

In our case, assuming completeness of the initial section, there seemed to be, essentially, three possible explanations for the recurrent pain. Firstly, that the pain was due to involvement of the remaining non-trigeminal somatic sensory neurons. However, this seemed an unlikely denominator as in the initial section of both the trigeminal nerves (in the VIth and the remainder of the Xth cranial nerves); secondly, that it was some form of postdenervation pain akin to the anaesthesia dolorosa described after spinal nerve section; and, thirdly, it was a form of trigeminal neuralgia, there being a reported coincidence of the two forms of neuralgia in a few cases. The close similarity of the recurrence to the initial pain, both in nature and site, the long period since the initial pain, and the response to carbamazepine all favoured the first possibility. On the basis of this diagnosis, coupled with the patient’s relative youth and, it might be said, her strong insistence, re-exploration was undertaken as described above. Whatever one’s position on the vascular compression theory of the cause of cranial nerve neuralgias this is an impressive and, in conjunction with the undesirability of further nerve section, encouraged treatment by microvascular decompression alone. The immediacy of pain relief, sustained now for 18 months, supports this decision. It might be argued that the presumed vascular compression was overlooked at the first procedure but there are several points against this. Firstly, both procedures were performed by the same surgeon, experienced in posterior fossa surgery, and the area of compression was in the same place as the initial section. Secondly, the young age at first presentation is against a vascular pathology, particularly where the vertebral artery is causative; and thirdly, there is a reported incidence of new vascular compression in re-exploration for recurrent trigeminal neuralgia.

In conclusion, the salient points to emerge from this brief report are that vagoglossopharyngeal neuralgia can recur after IXth and partial Xth nerve section and that this patient provides evidence for a pure vagal neuralgia. This supposition is supported by the finding that even the most caudal vagal rootlets may carry general somatic afferent fibres to the spinal trigeminal tract. In addition, the two separate episodes with differing pathologies raise the interesting question of whether there is a particular propensity for neuralgia which may or may not require a vascular trigger. This bears on the point raised by Adams et al as to why there are so many possibly causative vessels and so few neuralgias.

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Immunochemistry distinguishes between Pick’s disease and corticobasal degeneration

The clinical syndrome of frontotemporal dementia is associated with several neurodegenerative disorders. Pick’s disease, corticobasal degeneration, motor neuron disease associated dementia (MND-dementia), frontotemporal dementia and parkinsonism linked to chromosome 17 (FTDP-17), and frontal lobe degeneration (FLD). These disorders, although they do not match the frequency of Alzheimer’s disease, are far from uncommon, and present clinicians and neurologists with formidable, if not insurmountable, clinical challenges. However, recent advances in cellular and molecular pathology, biochemistry, and molecular genetics have been instrumental in their nosological definition. The discovery of a mutation in the tau protein gene on chromosome 17 in 1998 has established that several phenotypically heterogeneous familial dementias with a confusing variety of names all belong to FTDP-17. Most but not all frontotemporal dementias are characterised by intracranial inclusions formed by abnormal cytoskeletal components, both in neurons and glia. Pick’s disease, corticobasal degeneration, and FTDP-17 belong to the larger group of tauopathies, as their hallmark lesions contain tau protein, distinguishing them from MND/dementia and FLD, two disorders without tau pathology. Of the three tauopathies, FTDP-17 can be defined by its genetic abnormality, whereas the differential diagnosis of Pick’s disease and corticobasal degeneration remains difficult.

Clinically Pick’s disease is characterised by frontal and anterior temporal lobe dysfunction and progressive dementia, whereas neuropathologically the underlying frontotemporal atrophy is complemented histologically by the presence of large, swollen, achromatic neurons, the Pick cells, and by tau positive intraneuronal inclusions, the Pick bodies. The clinical features of corticobasal degeneration include the asymmetric atrophy of extrapyramidal parkinsonism, and the “alien limb” phenomenon (apparent purposeful movements which are not under voluntary control) followed by cognitive impairment. Histologically there is neuronal loss, astrogliosis, neuronal and glial inclusions, but the most prominent feature is the presence of large, swollen neurons, morphologically indistinguishable from Pick cells. It is the occurrence of the Pick cells in both disorders which causes most of the differential diagnostic problems.

In the human central CNS six tau isoforms are generated by alternative splicing and these are then posttranslationally phosphorylated. At molecular level the electrophoretic profiles of aggregated tau proteins in these neurodegenerative disorders are disease specific. For example, although both Pick’s disease and corticobasal degeneration are tauopathies, characterised by tau positive cellular inclusions, their tau profile is apparently different: the tau doublet is 64 and 69 kDa in corticobasal degeneration, but 55 and 64 kDa in Pick’s disease. HPLC analysis of 12 tau antibodies which, exploring the different phosphorylation patterns of these two disorders, has made a more accurate neuropathological diagnosis possible.

Sections from the temporal lobe and the temporal lobe (including the hippocampus) from 10 cases of corticobasal degeneration and 10 cases of Pick’s disease were examined by immunohistochemistry, using phosphorylation independent (SMI15, TP007, TP70, 304, 189) and phosphorylation dependent (AT180, AT270, AT8, 12E8) anti-tau antibodies. All sections were immunostained with all the anti-tau antibodies in conjunction with appropriate positive and negative controls. All the swollen neurons and neuronal and glial inclusions were positively immunostained with all the anti-tau antibodies in corticobasal degeneration. However, Pick’s disease all the antibodies but one immunostained the Pick bodies and the large swollen Pick cells: antibody 12E8 gave negative results (figure A and B). Phosphorylation at the site of Ser262/Ser263 is thought to be one of the most prominent factors affecting the biological activity of tau. In corticobasal degeneration antibody 12E8 detected the phosphorylated epitope Ser262 and/or Ser356 whereas in
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Pick's disease sites remain unphosphorylated and consequently this antibody did not label any of the Pick bodies or Pick cells. These findings are important both from a practical and from a theoretical point of view. The immediate practical implication is a more accurate neuropathological diagnosis, enabling Pick’s disease to be distinguished from corticobasal degeneration; this, in turn, is essential for improving clinical investigations. In addition, new insights into the molecular pathology of these disorders contribute to the nosological definition and better understanding of a complex group of neurodegenerative diseases.

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Neurological emergencies: acute stroke

We greatly enjoyed reading the recently published review on acute stroke by Davenport and Dennis. Their didactic approach and strict compromise to distinguishing between evidence based approaches and their personal beliefs deserve compliment. We would like to contribute by clarifying some information on two topics.

Firstly, the authors make a brief comment on the use of anticoagulants in the treatment of cerebral venous thrombosis (CVT) indicating that a recent randomised trial showed a non-significant favourable effect. This may lead to a misinterpretation. Most experts currently agree that anticoagulation should be used in the acute treatment of CVT based on the available evidence. In 1991, Einhaupl et al published the results of their randomised double-blind prospective study comparing heparin and placebo. This study was prematurely interrupted after enrolling 20 patients because of the dramatic differences noted in outcome in favour of the heparin group (basically eight patients recovered fully and no deaths occurred in the heparin arm whereas only one patient recovered completely and three others died in the placebo group). Concerns about possible methodological flaws in this German trial fueled some controversy about its results. This led to a new trial to the Cerebral Venous Sinus Study Group (1) which randomised 60 patients with CVT to receive nadroparin or placebo for 3 weeks. Poor outcome was defined as death, Barthel index of less than 15 at 3 weeks or Oxford handicap score equal or greater than 3 at 12 weeks. The results showed poor outcome in six of 30 patients (20%) in the nadroparin arm and seven of 29 (24%) in the placebo group at 3 weeks and four of 30 (13%) in the nadroparin treated patients versus six of 29 (21%) in the placebo group (absolute risk reduction of 7% and relative risk reduction of 38% for a non-statistically significant difference). But also very remarkably, there were no new symptomatic cerebral haemorrhages even among the 15 patients treated with anticoagulation who had haemorrhagic lesions on the initial CT. Finally, the same authors performed a meta-analysis combining the results of the two available trials which showed a “modest but clinically important” (although not statistically significant) benefit in the use of anticoagulation (absolute risk reduction of 14% in mortality and 15% in the nadroparin treated patients) with relative risk reductions of 70% and 56% respectively. When combined with the proved safety of anticoagulation even in the setting of pre-existent haemorrhagic infarct and the highly unpredictable course of patients with CVT, these results should encourage the use of anticoagulation in the treatment of CVT.

Secondly, Davenport and Dennis mentioned the use of decompressive craniectomy as one of the interventions that may be used in patients with stroke who are rapidly deteriorating from raised intracranial pressure. However, they questioned whether this aggressive approach was associated with “improved survival with acceptable quality of life.” Growing experience with our patients as well as the available literature seems to indicate that it does, especially when decompressive surgery is performed early. Schwab et al studied 63 patients with complete middle cerebral artery (MCA) infarctions and evidence of increased intracranial pressure treated with either early (within 24 hours of symptom onset) or late craniectomy. Mortality was 27% (compared with 78% in historical controls) and all survivors were reported to be able to walk short distances without assistance and none were left with global aphasia. Mean Barthel index scores were 68.8 in the early hemicraniectomy group, 62.6 in the late hemicraniectomy group, and 60 in historical controls (but the very high mortality in this last group may account for a less dramatic difference in functional outcome). Similar favourable results were reported by Carter et al in their retrospective analysis of 14 patients treated with decompressive surgery after massive non-dominant hemispheric infarctions. Eight of their 11 surviving patients were able to function with minimal to moderate assistance (Barthel index >60) 1 year after the surgery. Depression and failure to reintegrate socially were often found in this group of patients’ as opposed to the experience reported by Schwab et al.

In conclusion, decompressive craniectomy seems a valuable treatment option in cases of malignant MCA infarction, especially when involving the non-dominant hemisphere. This surgery is potentially lifesaving and reported functional outcomes are encouraging. Therefore, it needs to be considered early in every patient with complete MCA strokes showing incipient signs of increased intracranial pressure.

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patients, who have a greater capacity for functional recovery than older people (who make up the bulk of our case load), in whom we doubt whether such good functional outcomes are achievable.

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Illusory movements of the paralysed upper limb in stroke

Feinberg et al have recently reported the association between anosognosia for hemiplegia and the illusion of movement of the paralysed upper limb. They considered the illusion a form of confabulation that is distinct from other phantom phenomena. This view is not supported by my findings in a patient with a stroke who experienced transient purposeful movements of his paretic hand.

The patient was a 66 year old right handed man who presented with acute onset weakness of his right arm and leg and slurring of his speech. He was known to be hypertensive and a non-insulin dependent diabetic patient. Neurological examination confirmed the presence of right hemiplegia with facial involvement and mild to moderate sensory dysphasia. Muscle power, as measured by the Medical Research Council (MRC) scale, was 1/5 and 2/5 in the upper and lower limbs respectively. Proprioceptive, and posterior column sensations were intact. No visual field defects were found on examination using the confrontation method. There was no astereognosis or sensory extinction of tactile or visual stimuli. The patient was alert and cooperative. His comprehension of spoken and written language was good but there was evidence of moderately severe nominal dysphasia. The rest of the physical examination was normal. Brain CT confirmed the presence of a non-haemorrhagic infarct in the left corona radiata. The patient scored 19 on the mini mental state examination. There was no evidence of right hemispatial neglect as assessed clinically and with the line bisection test. The patient was correct in 8/10 items of the anosognosia for hemiplegia questionnaire.

Six weeks after his stroke the patient developed an itchy skin condition, probably a drug hypersensitivity reaction. When he scratched his skin with his left (good) hand to relieve the itching he thought that his right hand was scratching the same or a different area. He considered itching he thought that his right hand was active. When he scratched his skin with his left (good) hand to relieve the itching he thought that his right hand was scratching the same or a different area. He considered itching he thought that his right hand was active. Finally, it should be further noted that our study had left hemispatial neglect and left hemisensory defect. The patient described by Bakheit had a right hemiplegia, and had no neglect or sensory defects. Therefore, Bakheit’s patient would not have qualified for our study and cannot be fairly compared with our study population. Additionally, the factitious movements described in Bakheit’s patient differed from those experienced by our patients in two significant respects. Firstly, Bakheit’s patient experienced a “mirroring” phantom movement of the plegic right limb only when the normal hand was active. In our study, to minimise the potentially confounding role of completion, we specifically excluded from the main analysis those patients who only experienced illusory limb movements when the non-plegic limb was active. Secondly, the phantom movements experienced by Bakheit’s patient were restricted to a particular idiosyncratic action—namely, scratching—as opposed to those experienced by our patients who experienced illusory limb movements when simply asked to raise the left arm, an action which previously failed to elicit factitious movements in Bakheit’s patient. Therefore, according to the criteria set out in our investigation, the movements experienced by the patient of Bakheit could not be categorised as illusory limb movements in our study. Finally, it should be further noted that our patients were examined within a week of onset (some within a day) of acute hemiplegia, before significant “reorganisation of cortical maps and neural plasticity” is likely to have occurred. The patient of Bakheit is reported to have had phantom movements at 6 weeks after onset of hemiplegia when cortical reorganisation and neural plastic effects are more likely to have occurred.

In our opinion, Bakheit has committed the same error that we have previously cautioned against. He has failed to distinguish “phantom limb movements” from patients with true phantom limbs that occur in association with right hemispheric damage and hemineglect. Patients with true phantom limbs, as in Bakheit’s case, do not deny the identity of the actual arm and recognise the phantom movements as illusory. By contrast, the patients with illusory limb movements in our study all denied ownership of the plegic arm and believed in the reality of the factitious movements. Our group in which we found illusory limb movements and which bears a relation to anosognosia and represents a variety of confabulation. Finally, we point out that confabulation is not confined to amnesic patients, and occurs in other conditions such as Anton’s syndrome.

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Social deprivation and prevalence of epilepsy and associated health usage

I read the study of Morgan et al on social deprivation and prevalence of epilepsy and associated health usage with great interest. In my opinion, Bakheit has committed the same error that we have previously cautioned against. He has failed to distinguish “phantom limb movements” from patients with true phantom limbs that occur in association with right hemispheric damage and hemineglect. Patients with true phantom limbs, as in Bakheit’s case, do not deny the identity of the actual arm and recognise the phantom movements as illusory. By contrast, the patients with illusory limb movements in our study all denied ownership of the plegic arm and believed in the reality of the factitious movements. Our group in which we found illusory limb movements and which bears a relation to anosognosia and represents a variety of confabulation. Finally, we point out that confabulation is not confined to amnesic patients, and occurs in other conditions such as Anton’s syndrome.

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motor asymptomatic first degree relatives (mean age 52.6 (SD 10.6) years) of families with at least two members affected by Parkinson’s disease to 29 relatives (mean age 52.1 (SD 4.1) years) of patients with sporadic Parkinson’s disease and to 32 healthy controls (mean age 51.9 (SD 4.6) years). To account for a possible “low dopamine syndrome”, we studied memory, frontal lobe function, mood, personality traits, somatic complaints, and fine motor abilities. Tests used were the short form of the Wechsler adult intelligence scale, the auditory verbal learning test, the controlled oral word associ- ation test, the Wisconsin card sorting test (Nelson version), the paradoxed depression scale, the revised version of the Freiburg personality inventory, a list of complaints, and a standardized finger tapping test. We found that first degree relatives of both patients with familial Parkinson’s disease and those with sporadic Parkinson’s disease differed significantly from controls in several tests. They had lower scores in total fluency and fewer categories in the Wisconsin card sorting test. Relatives of both patients with familial Parkinson’s disease and with sporadic disease expressed more impulsiveness, more strain, and less extraversion on personality assessment. In addition, relatives of patients with familial Parkinson’s disease had more errors than controls in the Wisconsin card sorting test. Relatives of patients with sporadic Parkinson’s disease showed more depression, more somatic complaints, and inhibitedness than controls and also less extraversion, less emotionality, and a lower tapping rate of the right hand. Our results, both motor and non- motor, were comparable with those of patients with familial Parkinson’s disease and are in keeping with some of the findings of Dujardin et al.

On average, our proband sample was 14 years older than that of Dujardin et al, and by contrast with them we included assessment of depression as a possible confounder of the neuropsychological test results. Depression may have a substantial impact on cognitive function, and a history of depression is thought to be a risk factor for developing Parkinson’s disease.1 In our study, there were no correlations between cognitive impairment and depression. We therefore considered only lobe dysfunction and depression as independent signs of the “low dopamine syndrome” in our samples. Another important result of our investigation was that, apart from one personality trait (“aggressiveness”), we could not establish differences between relatives of patients with familial Parkinson’s disease and those of patients with sporadic Parkinson’s disease in any test item, nor were there item clusters in subsets of probands. Thus, according to our data, frontal lobe dysfunction and depression can be found to a variable degree in some relatives of patients with both the familial and the sporadic form of Parkinson’s disease. It should be kept in mind that the finding of such neuropsychological abnormalities does not prove that their origin is genetic.

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BOOK REVIEW


Have you ever had that feeling that something is just on the tip of your tongue but you can’t quite get at it or that if only you had one more piece of the jigsaw, you would be able to see the whole picture? Welcome to juvenile myoclonic epilepsy. It is one of the most rewarding conditions in epilepsy to diagnose and treat. Indeed juvenile myoclonic epilepsy has the unusual, dual virtues of being both common and treatable. But what is it? This book introduces the condition—prevalence 3%-11% of all epilepsy, easily diagnosed if you think to ask for early morning twitchiness or clumsiness, characteristic EEG appearance etc. But then come all the tantalising clues that leave one on the brink of understanding. It is obviously genetic and a linkage to chromosome 6 has been suggested for years, now honed down to near the HLA gene. But a recent analysis has tried to subdivide juvenile myoclonic epilepsy according to electroclinical criteria to obtain more homogeneous groups for genetic analysis and this has suggested genetic heterogeneity. Why are there so many focal elements in this generalised epilepsy syndrome? These include focal clinical seizure manifestations, focal EEG changes, focal imaging changes such as thickening of the grey matter detectable by mathematical techniques. What is the overlap with other syndromes such as childhood absence epilepsy and why are seizures triggered by reading or praxis in some cases? At least all can agree that it usually gets better with valproate but comes back if you stop the drug. Unfortunately this text does not discuss other newer medications. Experience with them is largely anecdotal except the treatment of the myoclonus with benzodi- azepines and piracetam.

This book summarises our knowledge of juvenile myoclonic epilepsy in a readable and concise but comprehensive text. The trouble is that we are on a threshold between descriptive knowledge and understanding so juvenile myoclonic epilepsy remains one jigsaw piece short of a picture. It will be of interest primarily to those in the epilepsy and genetics fields.

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