LETTERS TO THE EDITOR

Complex partial seizures provoked by photic stimulation

In patients with known or suspected epileptic seizures, non-specific activation methods such as hyperventilation or intermittent photic stimulation (IPS) are used to provoke epileptic potentials, which may prove the epileptic nature and specify epileptic syndromes. A photoconvulsive reaction with generalised spike wave activity may be provoked by IPS and is almost confined to patients with generalised epilepsy. There are, however, some reports on patients with partial epilepsy and photoconvulsive reaction.1 We report on two patients with known photoconvulsive reaction, who developed these with focal epileptic discharges consequent to IPS and discuss possible mechanisms.

Patient 1, a 44 year old woman presented with a 33 year history of complex partial seizures starting with behavioural arrest followed by oroalimentary automatisms, which were sometimes followed by secondary generalisation. She was treated with carbamazepine and reported 1–2 seizures a month. Brain MRI failed to disclose any focal abnormality. Except for mild generalised slowing (7.5%), probably due to or accentuated by carbamazepine, focal slowing (5–4/s) with phase inversion over F8, was seen in two EEG recordings (average of 1 spike in 7 minutes). During hyperventilation (3 minutes) the number of spikes increased to an average of 1 spike in 1 minute. During IPS (started with 1 s duration), the number of single spikes increased to 6 in 3 minutes or 2 per minute (figure). During the second recording, the spike activity in the anterotemporal region finally became rhythmic with subsequent generalisation. This was accompanied by a complex partial seizure typical for this patient (behavioural arrest followed by oroalimentary automatisms) finally running into a generalised tonic-clonic seizure.

Patient 2 was a 19 year old woman who had complex partial seizures with secondary generalisation for 2 years. The seizures started with fear (“indescribable terror”) accompanied by a fearful expression. This was followed by a repetitive ictal speech which was sometimes followed by secondary generalised tonic-clonic seizure. She reported one of these seizures as a consequence of flashing lights in a discotheque and avoided flashing lights since then. She was treated with valproate and reported 4–5 seizures per year. MRI was normal. EEG disclosed focal slowing (4 cps) with phase inversion over T6, which corresponded to decreased perfusion of the right midtemporal and parietotemporal regions established by HMPAO-SPECT. Intermittent photic stimulation (12/s) evoked a photoconvulsive reaction with bifrontal accentuated generalised spike-wave activity associated with myoclonic eyelid jerks. Independent of photoconvulsive reaction, 8 seconds later on single sharp-wave activity with phase inversion over T6, occurred consequent to IPS and became rhythmic. This was associated with complex partial seizures starting with fear accompanied by a terrifying fearful expression, which were followed by ictal speech (repetition of single words) finally running into a generalised tonic-clonic seizure (documented by a simultaneous EEG/Video recording).

Both patients developed complex partial seizures with secondary generalisation resulting from IPS and one of them reported a complex partial seizure provoked by flash-lights in a discotheque. To our knowledge, neither complex partial seizures nor activation of temporal epileptic activity consequent to IPS have previously been reported. Specific stimuli like rubbing, cold wind, or tactile stimuli may evoke spike activity in the contralateral cerebral regions and provoke partial seizures. Even patients with myoclonic epilepsy may develop contralateral spikes after electrical peripheral nerve stimulation. In all these patients, spike potentials were evoked in primary cortical representation areas of the respective stimuli. Our patients showed provocation of anterotemporal (Fp1, patient 1) and postero-temporal (T6, patient 2) epileptic activity resulting from IPS, which may have been adjacent to the visual cortex in patient 2 but was distinctly apart from the primary visual cortex in patient 1. Complex partial seizure symptomatology in the first patient included oroalimentary automatisms, indicating a seizure origin in the amygdalo-hippocampal complex. Visual hallucinations, which are likely with epileptic discharges in the visual cortex or visual association areas, however, were missed. This indicates that provoked complex partial seizures during IPS in our patients occurred without epileptic activity in the visual cortex. Temporal epileptic activity as a consequence of IPS was probably mediated via occipitotemporal connections such as the fasciculus longitudinalis inferior.

Provocation of sharp waves with phase inversion over Fp1, and the occurrence of a photoconvulsive reaction in patient 2 raises the question whether both phenomena were
interrelated. Similar constellations were previously reported in individual patients with photoconvulsive reaction who had partial epilepsy and occipital epileptic foci. Cortical and subcortical recordings in monkeys during IPS showed paroxysmal discharges predominantly in prerolandic areas, which were followed by bursts in the pontine and mesencephalic reticular formation and, finally, by generalised discharges. These findings have been interpreted in favour of a cortical origin of the photoconvulsive reaction, which is supported by the studies of Ricci et al using neuromagnetic methods in humans with photoconvulsive reaction to identify the location of the photoconvulsive reaction generator: They found a regional sensitivity involving frontal, occipital, and temporal areas, but the cortical excitability was extremely unstable, which was attributed to a deficient GABAergic system. This suggests that photoconvulsive reaction is a generalised phenomenon and not due to polyfocal generation. The occurrence of focal epileptic discharges associated with focal seizures and secondary generalisation in patient 2 does not indicate a relation between focal epileptic discharges and the photoconvulsive reaction as the second appeared in only one of the patients.

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Cortical discharges and the photoconvulsive reaction in Creutzfeldt-Jakob-like syndrome

Creutzfeldt-Jakob-like syndrome induced by lithium, levomepromazine, and phenobarbitone

Creutzfeldt-Jakob-like syndrome was first reported by Smith and Kocen in 1988. Its symptoms resemble Creutzfeldt-Jakob disease but it is induced by drugs, particularly lithium, and most patients recover without sequel after discontinuation of drugs. It also displays a characteristic EEG similar to Creutzfeldt-Jakob disease, but this returns to normal when the patient recovers.

There have been some case reports of Creutzfeldt-Jakob-like syndrome after that of Smith et al (table), but no paper seems to have described the detailed course of EEG changes. This paper presents a case of Creutzfeldt-Jakob-like syndrome possibly induced by lithium, levomepromazine, and phenobarbitone, in which we succeeded in recording the course of EEG changes.

A 65 year old woman was admitted to a hospital with coma and myoclonus. She had a history of manic and depressive disease for 8 years and had been treated with 200 mg lithium carbonate, 25 mg chlorpromazine, and 10 mg levomepromazine daily. Her first symptom was forgetfulness from 20 May, then she complained of appetite loss from 27 May, diarrhoea from 1 June, myoclonus from 3 June, and gait disturbance from 4 June. At the same time she complained of visual disturbance. Gradually her conscious level declined. When she was admitted to the hospital on 4 June, she had convulsions. At that time, she was injected with 200 mg phenobarbitone intramuscularly and this was continued for 2 more days at the same dose. Physical examination disclosed no abnormality. Neurologically there was general hypotonia and hypeflexia without Babinski’s sign. Serum glutamic oxaloacetic transaminase, glutamic pyruvic transaminase alkaline phosphatase, and creatine kinase were increased slightly, and serum ammonia was 64 µmol/l (normal range 30–59 µmol/l). Plasma sodium and potassium concentrations were normal. Her creatinine clearance was 46 ml/min and thyroid function was normal. Examination of CSF gave normal results. Chest radiography, brain CT, and brain MRI showed no abnormality. ECG showed T wave inversion from V1 to V3. The EEG showed slow basic activity but no periodic discharge on 4 June, but showed PSD on 7 June (figure). Its periodicity decreased on 10 June and had returned to her previous EEG on 19 June. Her ECG had also returned to normal by 14 June. Her myoclonus disappeared on 6 June, and her conscious level gradually improved from 9 June; she could open her eyes on 10 June, then could answer our questions regarding place and time and could walk without help from 13 June. She was discharged on 25 June fully recovered.

She was diagnosed as having Creutzfeldt-Jakob-like syndrome induced by lithium,
chlopromazine, levomepromazine, and phenobarbital. Her CSF lithium concentration was 0.82 mmol/l on 4 June. According to Taguchi et al,^4^ lithium concentration in CSF is about one fourth of the serum concentration after taking lithium for more than 1 week, and it has been shown that a serum lithium concentration is toxic above 1.5 mmol/l, so her serum concentration is likely to be high enough to be toxic. Her symptoms such as forgetfulness, diarrhea, coma, myoclonus, and visual disturbance were all compatible with lithium intoxication. The cause of her high lithium concentration was clear with the discovery that she took three times as much as prescribed when she could not sleep well. Periodic EEGs on April 22, 2022, and June 16, 2022, showed no epileptic activity, nor were any new EEG changes noted. The drugs were discontinued and was displayed for about 3 days. There are some case reports of Creutzfeldt-Jakob-like syndrome induced by chlopromazine and levomepromazine, but there are apparently no reports of its induction by phenobarbital. We could not identify the role of phenobarbital injection from 4 to 6 June, but it was possible it might have some part in the induction of PSD, and her hypotonicity and hyporeflexia are uncommon compared with previous reports.5,6 In conclusion, this drug induced Creutzfeldt-Jakob-like syndrome showed us the importance of taking a drug history, as previously pointed out by Smith et al.7

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Central nervous system involvement in a novel connexin 32 mutation affecting identical twins

Connexin 32 (Cx32) is a gap junction protein expressed in the peripheral nervous system (PNS), central nervous system (CNS), and in many other tissues.1 Mutations in the Cx32 gene are associated with X-linked Charcot-Marie-Tooth disease (CMTX), and account for about 10% of the patients with hereditary motor and sensory neuropathy (HMSN).

At least 130 different mutations have been reported in the Cx32 gene causing peripheral neuropathy. Classically, distal weakness and atrophy initially involving the lower limbs, as well as sensory abnormalities, depressed tendon reflexes, and pes cavus are usually found in males by the second decade, whereas carrier females clinical manifestations, if present, are in most instances milder than in affected males. Nerve conduction studies in affected males are usually, but not always, suggestive of a demyelinating process, although they are not quite as slow as in patients with CMT1A. In females, conduction velocities (CVs) may be in the normal range or only mildly reduced, as seen in axonal neurodysfunction.

We describe a new Cx32 point mutation (A^64_ to Val) in genetically established identical twins with similar CMT phenotype and extensor plantar reflexes. The probands were first seen at the age of 20. Their principal complaint was cramps in the legs, “going over” on the ankles, and mild weakness in the hands. On examination, Twin 1 could not stand on his heels and had a mild intrinsic hand muscle weakness. There was a mild distal atrophy in both upper and lower limbs. Pinprick and tactile sensations were diminished up to the knees and vibration was impaired distally in the lower limbs. Tendon reflexes were not assessed, but both plantar responses were extensor. His median, ulnar, and peroneal motor CVs were 33.0 m/s, 33.0 m/s, and 31.0 m/s, respectively, and the distal amplitudes were 0.7 mV, 5.0 mV, and 3.3 mV. The sensory potentials were all absent. Twin 2 had identical clinical manifestations, except that the left plantar reflex was flexor whereas the right was clearly extensor. His motor CVs and amplitudes of the same nerves described above were 32.0 m/s and 1.7 mV, 34.0 m/s and 6.0 mV, and 33.0 m/s and 4.0 mV, respectively. No sensory response was obtained. Their mother had minimal neuropathic features and both plantar reflexes were extensor. Her median and peroneal motor CVs were 43.0 m/s and 37.0 m/s, and the median sensory CV was 40.0 m/s. Their sister and the mother’s brother were clinically and electrophysiologically normal. The maternal grandfather was not examined, but had a long history of a slowly progressive neuropathy.

The presence of the 17p11.2-p12 duplication was excluded by fluorescent quantitation of the chromosomes and segregates with the disease. Another member of the family carrying the mutation had normal MRI and other non-related patients with the same mutation did not show any clinical signs of CNS involvement, raising the possibility of a casual association. Bell et al presented a family with a mutation on code 93 whose clinical manifestations included tremor, brisk reflexes and spasticity. On MRI there was atrophy of the cerebral cortex and cerebellum. The presence of a Babin’s sign in our family strongly suggests that in this novel mutation there is involvement of the corticospinal tract. Unfortunately no imaging or evoked potential studies were possible. Cx32 is a gap junction protein expressed in the parietal region and Schmidt-Lantermann incisures in the PNS, and in cell bodies and oligodendrocytes processes in the CNS. Why mutations in Cx32 usually lead only to PNS dysfunction is still an open question. Presumably, there is a unique relation between Cx32 and the structural organisation of the PNS. Another possibility is that other connexin proteins might compensate for Cx32 dysfunction in the CNS and other tissues, but not in the PNS.

Although the clinical manifestations are extremely similar in most of the Cx32 neuropathies suggesting that different mutations do not cause different phenotypes, different degrees of severity and the presence of unusual signs, like the one we present here,
have already been described to occur with some mutations.

There are only two previous reports relating to three pairs of identical twins with CMT and known genetic defects. In the two pairs with the 1p11.2 duplication there was remarkable clinical variability. We have also seen a pair of identical twins with a P0 mutation in whom there was marked variability in early ages (unpublished data). Apart from the asymmetry of toe responses in one of the probands, the genetically identical twins described here are phenotypically very similar, suggesting that the expression of this mutation was not influenced by other non-genetic factors.

Codon 39 seems to be of particular importance to Cx32 protein function as changing of the wild type amino acid has caused CNS dysfunction in addition to the peripheral neuropathy. Moreover its expression does not seem to depend on non-genetic factors, as might be expected in a hemizygous condition.

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**Letters, Correspondence, Book reviews**

**Sagittal T2 weighted MRI of the cervicodorsal cord : high linear signal extending from C4 to T5 vertebral levels.**

A predominantly right sided tetraparesis and urinary retention. There was no history of neck trauma. Cranial nerve examination was normal. There was a right sided hemiplegia and a moderate left sided hemiparesis. Deep tendon reflexes were normal, right plantar response was extensor. There were bilateral spinohalamic problems below T4 with loss of touch sense in the right leg. Thus the examination was consistent with atypical, right cervical Brown-Séquard's syndrome.

**Biological investigations were normal.** CSF protein was 0.54 g/l, glucose 2.54 mmol/l (2.10–4.20 mmol/l). There were 7 white cells and 25 red cells/mm³.

There were no oligoclonal bands. The ECG was normal. There was no aortic dissection shown on CT or MRI. Visual evoked potentials were normal. Somatosensory evoked potentials were abnormal for the right lower limb at the cervical level. A sagittal T2 weighted MRI showed linear high signal from C4 to T5 vertebral levels consistent with an ischaemic lesion (figure). On corresponding axial cuts, this was shown to involve the region of the anterior horns at cervical level and to prevail on the right half of the spinal cord at dorsal level. MRI of the cerebellum and brain stem was normal. Cerebral angiography showed an irregular stenosis of the right and left cervical vertebral artery typical of a dissection. The patient was treated with oral anticoagulants. One year later, the sequelae were a spastic paraparesis with right sided central pain and mild urinary retention. MRI and MRA showed the resolution of the cord signal and normal right and left vertebral artery.

The cervical cord is mainly supplied by radicular arteries rising from the vertebral artery. Thus, vertebral artery dissection can lead to an ischaemia limited to the cervical cord. Extensive ischaemia to the dorsal cord (T5) is uncommon. Our results suggest that this area is sometimes supplied from the vertebral artery. Some authors state that this region could be a critical zone and its vascularisation could be provided from the arterial cervicodorsal cord region. The bilateral ischaemic lesions extending through several cervical and dorsal segments are in favour of water-shed infarcts caused by hypoperfusion due to bilateral vertebral artery dissection.

**Autonomic dysfunction and orthostatic hypotension caused by vitamin B12 deficiency**

Orthostatic hypotension sometimes is a reversible neurological complication of vitamin B12 deficiency.1,2 Eisenhofer detected deficient sympathetic chromaffin release in insulin tolerance testing,3 but the mechanism of orthostatic hypotension in vitamin B12 deficiency remains unclear. We report a patient with vitamin B12 deficiency and reversible orthostatic hypotension and discuss the mechanism of this symptom.

A 77 year old man admitted to our hospital had had unstable gait and urinary urgency for 6 months, clumsiness of the hands and tingling sensations in the legs for 3 months, and, for a month, occasional dizziness on standing. The dizziness was mild without any attack of syncope. He had no other symptoms or signs of autonomic dysfunction but impotence and erectile failure were noted 10 years before the onset of neurological symptoms. He had not taken any medicine which would affect the autonomic nervous system. He did not have a habit of drinking.

Physical examination on admission detected no signs of anaemia, heart failure, or dehydration. Neurological examination showed dysaesthesia and decreased sensation of all modalities in the legs for 3 months, and, for a month, occasional dizziness on standing. The dizziness was mild without any attack of syncope. He had no other symptoms or signs of autonomic dysfunction but impotence and erectile failure were noted 10 years before the onset of neurological symptoms. He had not taken any medicine which would affect the autonomic nervous system. He did not have a habit of drinking.

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over 30 mm Hg with normal heart rate response. His serum noradrenaline concentration was reduced at rest, and its increase after tilting up was minimal. Sudomotor function was evaluated by sympathetic skin response (SSR) \(^1\) and local sweat response to acetylcholine (Ach). \(^2\)

Before treatment, the SSR amplitude was decreased, and the number and area of sweat droplets were decreased in responses to intermittent Ach injection. The myelinated fibre density of biopsied sural nerve was 5927/\(\mu m^2\). Some thin myelinated fibres were present, as were a few myelin ovoids. Examination of the teased fibres showed evidence of demyelination (about 20\%) and axonal degeneration (about 10\%). Electron microscopy showed a normal unmyelinated fibre density (30 945/\(\mu m^2\)). Collagen packets (15 000/\(\mu m^2\)), and denervated Schwann cell subunits (12 000/\(\mu m^2\)) were present, but their densities were within the normal range for his age. \(^3\)

A highly sensitive acetylcholinesterase (AChE) histochemical test (modified Tago’s method) \(^4\) of the sural nerve detected a slightly reduced density of sudomotor sympathetic unmyelinated fibres (3500/\(\mu m^2\); normal 3700–6500/\(\mu m^2\)).

Daily intramuscularly administered 1 mg vitamin B12 for a week then 1 mg once a month increased its serum concentration rapidly to normal, resulting in the gradual amelioration of orthostatic dizziness, and his neurological symptoms except for erectile failure, after a month. The abnormalities seen in the autonomic nervous system tests also disappeared when vitamin B12 was given for 6 months (table). The serum of the baroreflex responsible for his orthostatic hypotension is considered to be in the efferent pathway because of the preserved heart rate response in head up tilt test.

The low serum noradrenaline concentration in particular can be explained by disturbance of the sympathetic postganglionic fibres. These findings are supported by the decreased SSR amplitude and the reduced local sweat response to Ach. By contrast, the density of the unmyelinated fibres and AChE positive fibres were relatively well preserved when his age was considered. Furthermore, there was the rapid recovery of serum noradrenaline concentration, the SSR size, and the sweat response to Ach after giving the vitamin B12 supplement. These results suggest dysfunc-

nal cord induce the dysfunction of postganglionic fibres by a trans-synaptic effect. \(^5\) Vitamin B12 is related to the methylation reaction regulated by S-adenosylhomocysteine and S-adenosylmethionine. \(^6\) This reaction has a crucial role in the myelin formation associated with neurological deficits in patients with vitamin B12 deficiency. Dysfunc-

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Letters, Correspondence, Book reviews

CORRESPONDENCE

Sandifer’s syndrome and gastro-esophageal reflux disease

Perkin and Murray-Lyon’s Neurology and the gastrointestinal system reviews gastrointestinal disorders with neurological features. \(^7\) The authors do not mention Sandifer’s syndrome, a disorder of the upper gastrointestinal tract with neurological manifestations occurring in children and adolescents. Sandifer’s syndrome is the association of gastro-esophageal reflux disease with spastic torticollis and dystonic body movements. Nodding and rotation of the head, neck extension, gurgling sounds, writhing movements of the limbs, and severe hypo-

1 Kalbfleish JM, Woods AH. Orthostatic hypotension associated with pernicious anemia: report of a case with complete recovery following vita-

min B12 therapy. \(JAMA\) 1962;182:198–200.


9 Metz J. Cobalamin deficiency and the pathogenesis of nervous system disease. \(Anna\) \(Neurol\) \(Nerv\) \(Syst\) 1992;42:59–79.

Results of autonomic nervous system tests before and after vitamin B12 treatment

<table>
<thead>
<tr>
<th>Tests</th>
<th>Before treatment</th>
<th>After treatment</th>
<th>Age matched normal control</th>
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<tbody>
<tr>
<td>Head-up tilting test</td>
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<tr>
<td>Systolic blood pressure (mm Hg)</td>
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<tr>
<td>Supine</td>
<td>104</td>
<td>106</td>
<td>112 – 135</td>
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<tr>
<td>5 min after tilting</td>
<td>71</td>
<td>93</td>
<td>118 – 140</td>
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<tr>
<td>15 min after tilting</td>
<td>76</td>
<td>106</td>
<td></td>
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<tr>
<td>Heart rate (bpm)</td>
<td></td>
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<tr>
<td>Supine</td>
<td>58</td>
<td>66</td>
<td>59 – 83</td>
</tr>
<tr>
<td>5 min after tilting</td>
<td>70</td>
<td>73</td>
<td>65 – 95</td>
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<tr>
<td>15 min after tilting</td>
<td>73</td>
<td>76</td>
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<tr>
<td>Noradrenaline (pg/ml)</td>
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<tr>
<td>Supine</td>
<td>94</td>
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<td>128 – 512</td>
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<tr>
<td>5 min after tilting</td>
<td>129</td>
<td>288</td>
<td>258 – 752</td>
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<tr>
<td>Sympathetic skin response</td>
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<tr>
<td>Amplitude (mV)</td>
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<tr>
<td>L palm</td>
<td>0.33</td>
<td>0.9</td>
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<tr>
<td>R palm</td>
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<tr>
<td>L Sole</td>
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<td>1.9</td>
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<tr>
<td>R Sole</td>
<td>0.61</td>
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<td>Local sweat response to acetylcholine</td>
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<tr>
<td>Number of sweat droplets ((\mu m^2))</td>
<td>24</td>
<td>69</td>
<td>42 – 72</td>
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<tr>
<td>Total area of sweat droplets ((mm^2/cm^2))</td>
<td>0.27</td>
<td>2.86</td>
<td>0.75 – 11.5</td>
</tr>
</tbody>
</table>

1 Perkin GD, Murray-Lyon I. Neurology and the gastrointestinal system. \(J\) \(Neurol\) \(Neurosurg\) \(Psychiatry\) 1998;66:291–300.


3 Sutcliffe J. Torsion spasm and abnormal postures in children with hiatus hernia Sandifer’s syndrome. \(Prog\) \(Paediatr\) \(Pae\) 1969;2:190–7.

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DEMETRIOS S THEODOROPOULOS
Is inherited thrombophilia a risk factor for arterial stroke?

The paper of Ganesan et al adds the factor V Leiden mutation to the list of inherited thrombophilias which has not been shown to be significantly increased in consecutive series of children and young adults with arterial stroke. 1-3 In their commentary on this paper, Brown and Bevan 4 admit ignorance as to whether the finding of inherited thrombophilia in a patient with stroke indicates an increased risk of recurrent stroke but nevertheless recommend consideration of lifelong anticoagulation. No evidence in support of this recommendation is cited.

Brown and Bevan recommend repeating measurements of protein C, protein S, and antithrombin III for at least 3 months after the acute event but depressed concentrations returning to normal between 12 and 24 months after childhood stroke have previously been reported. 2, 5 It would therefore seem prudent to follow concentrations of protein C and protein S for at least this time period before concluding that they can be attributed to an inherited thrombophilia, particularly if the presence of such a disorder is to be managed by “lifelong anticoagulation”.

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Lyme borreliosis and intracranial aneurysm

We read the article by Oksi et al describing three patients with Borrelia burgdorferi infection and intracranial aneurysms with great interest. We encountered a patient with neuroborreliosis and an aneurysm of the basilar artery, whom we describe.

A previously healthy 33 year old man presented with headache and progressive right hemiparesis. On neurological examination there was right facial weakness, moderate weakness of the right arm and leg (3/5), and brisk deep tendon reflexes. A right Babinski’s sign was present. Cerebral CT and MRI showed left anterior infarction, without enhancement with contrast. Examination of CSF disclosed a leucocyte of 3/mm3; the protein content was 3.49 g/l. The IgG index was raised to 1.35. The CSF was xanthochromic, because of bilirubin. IgG antibodies against Borrelia burgdorferi in the CSF were detected. A cerebral angiogram showed narrowing of the left anterior cerebral artery and an aneurysm of the basilar artery. Serum IgG antibodies against Borrelia burgdorferi were positive. A diagnosis of Borrelia burgdorferi infection was considered. However, we postulate that the presence of the aneurysm in our patient was a coincidence. There are two other explanations for the xanthochromia: bilirubin in the CSF. The first is the raised protein content of the CSF (in a patient with subarachnoid haemorrhage due to neuroborreliosis). Our patient had an aneurysm of the basilar artery. If one of the above mechanisms in neuroborreliosis, it can also lead to formation of aneurysms or vascular infarction.

Let us consider a causal relation between neuroborreliosis and the aneurysm. In our patient, only one of the three patients had neuroborreliosis. In the other two patients there was no evidence of involvement of the central (or peripheral) nervous system. The authors state that an increased cell count or increased protein content is a necessity for neuroborreliosis, referring to an article by Garcia-Monco and Benach. However, this is not always the case as shown, for example, in the studies by Lu and Coyle in which they describe patients with neuroborreliosis having DNA or antigens of Borrelia burgdorferi in CSF despite normal results in the authors’ own laboratory. The authors think that for some reason, could not be associated with the focal narrowing of the left anterior cerebral artery with Borrelia burgdorferi but could not do the same with aneurysms of the basilar artery. They do not have any evidence that excludes the possibility that the aneurysm could also have been caused by Borrelia burgdorferi. As we refer to in our article, high numbers of inflammatory cells have been found in aneurysms. We consider that it is possible that the aetiology of inflammatory changes in both instances may be the same—although probably seen at a different time point. We, and others, have found that disseminated Lyme borreliosis is not associated with cerebral vasculitis. 6

Polet and Weinstein present a case of a patient with an intracranial aneurysm and suspected neuroborreliosis. They write that antibodies against Borrelia burgdorferi were detected in the CSF. However, the total IgG was increased, and they do not state whether the antibodies were intracellular or extracellular. We do not have any evidence that they are intracellular or extracellular. If vasculitis is one of the primary pathological mechanisms in neuroborreliosis, it can also lead to formation of aneurysms or vascular infarction. However, we are unable to explain what happened in our patient by using their concept. Our patient had an aneurysm of the basilar artery. They do not have any evidence that excludes the possibility that the aneurysm could also have been caused by Borrelia burgdorferi. As we refer to in our article, we have found DNA or antigens of Borrelia burgdorferi in CSF despite normal results in the authors’ own laboratory. The authors think that for some reason, could not be associated with the focal narrowing of the left anterior cerebral artery with Borrelia burgdorferi but could not do the same with aneurysms of the basilar artery. They do not have any evidence that excludes the possibility that the aneurysm could also have been caused by Borrelia burgdorferi.


Letters, Correspondence, Book reviews

BOOK REVIEWS


These three books and CD-ROM form part of a six book series for the Open University course on Biology: Brain and Behaviour. Book 2, Neurobiology, covers the biophysical properties of the neuron, before dealing with neural networks and the functions of the CNS to the immune system and behaviour. Book 3 runs through the senses, with the discussion concentrating on the audition, vision, and somatosensory systems. Book 4 and 5 in the series deals with disease processes of the brain and mind. The books not reviewed comprise the first book in the series entitled Behaviour and Evolution and books 4 and 5 on Development and Flexibility and Control of Behaviour. The six books are therefore written for a specific audience, which is clearly reflected in the format of the text and figures. However, this having been said, these books are easily accessible to other students not primarily interested in neurosciences, although the way the books are packaged and presented would make them hard to commend all the details raised in these books, although the accompanying CD-ROM is a helpful innovation in this respect. Overall these books and CD-ROM make an attractive package which is lost to most students and teachers as a result of it being used for a specific Open University course. However, there is much of value in this series for those interested in the education of medical students.

JERRY BROWN


This is a book of 172 pages dedicated to the memory of Frank Morrell. It is a multiauthored text, originating largely from North America (with a notable United Kingdom contribution from the Maudsley Hospital). After a historical review including stimulation and recording techniques, novel approaches to using electrocorticography to predict surgical outcome after temporal lobe resection are presented convincingly and then followed by another chapter showing how parallel approaches can be applied in tailored resections. Electrocorticography findings in extratemporal epilepsy are then dealt with, confirming that restricted frontal lobe abnormalities predict a favourable outcome, particularly when combined with a well defined structural lesion. The technique of chronic electrocorticography is also reviewed, including the demonstration of how to define the limits of interictal epileptiform activity and the ictal onset zone if a complete resection of the structural lesion is not possible.

The disparate results in clinical studies using pharmacological activation are then considered, but sensible conclusions are drawn about the relatively minor role of this approach in determining the limits of a potential cortical excision. It must be emphasised that this book keeps its feet on the ground where necessary. A comprehensive multidisciplinary contribution follows, describing the findings in cortical dysplasia, and the way this probably affects the surgical outcome when compared with patients with other structural lesions. There are then three chapters on studies in the mesial temporal region, involving patient selection, prognosis, volume, and the combination of acute and chronic electrocorticographic techniques. Some of this is then applied to a chapter on hemispherectomy. The book finishes with chapters on the applications of image guided surgery to...

The complex relations between intracranial and inner ear fluids are fascinating for both the scientist and the clinician. This volume represents the Proceedings of the Second International Conference on Intracranial and Inner Ear Fluids, which was held in Bath, UK in June 1997, and accurately reflects the sense of enthusiasm and collaboration at that meeting. The contributors include neurosurgeons, audiologists, otologists, neurologists, epidemiologists and basic scientists, and the scope of the material is very impressive.

The book comprises four sections. The first, Intracranial physiology, contains four chapters including a very clear review of the anatomy and physiology of intracranial fluids by Segal, and then three examples of experimental work on cats, guinea pigs, and humans. The second section, Intracranial pathophysiology, opens with a review of “Pathophysiology of the cerebrospinal and cerebrovascular circulations” by Pickard et al, and then eight chapters considering related topics. The tympanic membrane displacement (TMD) test procedure is discussed, representing a non-invasive method of assessing intracranial fluid pressure, and particularly useful in the assessment of shunt malfunction. The third section, Inner ear physiology, contains 10 chapters, and considers the inner ear fluids, perilymph, and endolymph in very considerable detail. The final section, Inner ear pathophysiology, is perhaps the least consistent in the volume and at times strays from the fluid remit of the book. It does, however, contain a very useful chapter considering the Tullio phenomenon (by O’Mahoney and Luxon) that deserves careful study.

For anyone interested in the areas described above this book will be interesting and useful. Collaboration and indeed communication between those interested in the intracranial fluids and inner ear fluid is in its infancy, and whereas this book does contain exciting material there is little that is of clinical relevance yet, although some of the techniques and concepts described hold great promise. Many departmental libraries would benefit from the inclusion of this volume, although only those directly involved in this area would be able to justify a private purchase.

DAVID BAGULEY


No one can doubt the increasing importance, to affected families and the healthcare system, of Alzheimer’s disease, Parkinson’s disease, and the other degenerative conditions of the nervous system. Furthermore, study of the degenerating brain can provide fundamental insights into brain function. Although there are authoritative books on memory, disorders of memory, and on the neurological diseases covered in this book, the strength of the book is in the accounts of different views of memory in neurodegenerative disease. These differing perspectives mean that this book will be of interest to neurorologists, neuropsychologists, psychiatrists, and researchers in the neurosciences.

The book is divided into three broad sections with summary chapters at the end of each. The first section deals with the biological aspects of neurodegenerative disease, with reviews on neuropathology, animal models, neurochemistry, and neuroimaging.

The two chapters on neuroimaging are particularly valuable, being clear and well referenced. Although the genetic advances in this area are mentioned in several chapters, it is not a major topic in this work.

The last section of this book will be particularly useful for clinicians, as there are admirable summaries of the assessment of memory, including very interesting accounts of cross cultural issues in neuropsychological assessment and the reliability of psychometric instruments. The important clinical issues of early detection and of differentiating dementias and memory disorders are well presented. This section ends with an exploration of drug and surgical treatments for neurodegenerative disease.

There is particular consideration of the possible cognitive sequelae of neurosurgery for akinetic-rigid syndromes and tremor. I would recommend this book to anyone who wants a clear and authoritative account of the role of neuropsychology, experimental psychology, and theories of memory structure and organisation in relation to the neuropsychology of the dementias and other neurodegenerative conditions.

CLARE GALTON