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.

We report on two patients with known photovconvulsive 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/s), probably due to or accentuated by carbamazepine, focal slowing (5–4/s) with phase inversion over F 8, 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 seizures. 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.

Intermittent photic stimulation (12/s) evoked a photovconvulsive reaction with bifrontal accentuated generalised spike-wave activity associated with myoclonic eyelid jerks. Independent of photovconvulsive reaction, 8 seconds later on single sharp-wave activity with phase inversion over T 6, 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.

Both patients developed complex partial seizures with secondary generalisation resulting from IPS and one of them reported a complex partial seizure provoked by flash-light 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 (F 7, patient 1) and posterotemporal (T 6, 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 F 8, and the occurrence of a photovconvulsive reaction in patient 2 raises the question whether both phenomena were...
interrelated. Similar constellations were previously reported in individual patients with photoco
vulsive reaction who had partial epilepsy and occipital epileptic focus. 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 photoco
vulsive reaction, which is supported by the studies of Ricci et al using neuromagnetic methods in humans with photoco
vulsive reaction to identify the location of the photoco
vulsive re
action 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 photoco
vulsive 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 photoco
vulsive reaction as the second appeared in only one of the patients.

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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 phenobarbione, 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 phenobarbione intramuscularly and this was continued for 2 more days at the same dose. Physical examination disclosed no abnormality. Neurologically there was general hypotonia and hyporeflexia without Babinski’s sign. Serum glutamic oxaloacetic transami
nase, glutamic pyruvic transaminase alkaline phosphatase, and creatine kinase was increased slightly, and serum ammonia was 64 \( \mu \text{mol/l} \) (normal range 30–59 \( \mu \text{mol/l} \)). 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,
cholorpromazine, levomepromazine, and phe- 
nobarbitone. Her CSF lithium concentration was 0.82 mmol/l on 4 June. According to 
Taguchi et al., lithium concentration in CSF is about one fourth of the serum concentra-
tion after taking lithium for more than 1 week, and it was thought that a serum lithium con-
centration 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, diarrhoea, 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 merged 3 days after all 
the drugs were discontinued and was displayed for about 3 days. There are some case reports of 
Creutzfeldt-Jakob-like syndrome induced by chlorpromazine and levomepromazine, 
but there are apparently no reports of its induction by phenobarbitone. We could not 
identify the role of phenobarbitone injected 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 uncom-
mon compared to previous reports.4–6

In conclusion, this drug induced Creutzfeldt-
Jakob-like syndrome showed us the importance of taking a drug history, as previ-
ously pointed out by Smith et al.

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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 
neuropathy 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 ten-
don reflexes, and pes cavus are usually found 
in males by the second decade, whereas in 
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, al-
though they are not quite as slow as in 
patients with CMT1A. In females, conduc-
tion velocities (CVs) may be in the normal 
range or only mildly reduced, as seen in 
axonal neuropathies.

We describe a novel Cx32 point mutation 
(Ala 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 
intrinsics muscle weakness. There was a 
distal atrophy in both upper and lower 
limb. Pinprick and tactile sensations were 
diminished up to the knees and vibration was 
damaged distally in the lower limbs. Tendon 
reflexes were reduced, but both 
plantar responses were extensor. His median, 
ulnar, and peroneal motor CVs were 33.0 
ms, 33.0 ms, and 31.0 ms, respectively, and 
the distal amplitudes were 0.7 mV, 5.0 mV, 
and 3.5 mV. The sensory potentials were all 
absent. Twin 2 had identical clinical manifes-
tations, 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 
ms, 34.0 ms and 6.0 mV, and 33.0 ms and 
4.0 mV, respectively. No sensory response 
was obtained. Their mother had minimal 
neuropathic features and both plantar re-
flexes were extensor. Her median and pero-
neal motor CVs were 43.0 ms and 37.0 ms, 
and the median sensory CV was 40.0 ms. 
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 a high resolution demonstrative 
quantitative polymerase chain reaction with five mic-
rosatellite markers contained within the 
involved segment.

Sequencing Cx32 with the ABI™ 
Dye Primer Cycle Sequencing Ready Reaction 
Detected a C to T transition (figure) at 
amino acid 39 causing an alanine to valine 
substitution in the first extracellular loop. 
This mutation abolishes a restriction site for 
the enzyme BssHII and oligotyping 200 control 
cromosomes and the father’s DNA, no 
mutation was found. The mother was shown 
to harbour the mutation.

The monozygosity status of the twins was 
confirmed by the observation of the same 
alleles at each of the 13 highly polymorphic 
microsatellite markers tested. The possibility 
of this occurring by chance is >0.01%.

Although the clinical manifestations 
are extremely similar in most of the Cx32 
neuropathies suggesting that different muta-
tions do not cause different phenotypes, 
different degrees of severity and the presence of 
unusual signs, like the one we present here,

Sense electropherogram of a segment of Cx32 
exon 2 showing the C to T mutation at base pair 
39 (open arrow) and the corresponding control 
segment (lower trace).

Val amino acid substitution are clearly on the 
right side of the classic CMT phenotype 
spectrum. This amino acid is conserved in 
other species, not found in 200 control chro-
mosomes and segregates with the disease.

A second notable feature in this family is 
the presence of extensor plantar responses in 
all three people shown to carry the mutation. 
Involvement of the CNS in patients with 
Cx32 mutations have been demonstrated by 
slowly of the central conduction time in 
their brainstem auditory evoked potentials,7 
but clinical manifestations secondary to cen-
tral dysfunction does not seem to be a 
common feature. Paulson et al.8 reported a 
patient who developed dystarhy and incor-
rodaptation after high altitude skiing. His MRI 
showed conluent, symmetric, white matter 
changes. Another member of the family 
carrying the mutation had normal MRI9 
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 re-
flectance and spasticity. On MRI there was atro-
phy of the cerebral cortex and cerebellum. 
The presence of a Babinski’s sign in our family 
strongly suggests that in this novel mutation 
there is involvement of the cortico-
principal tract. Unfortunately no imaging or 
evoked potential studies were possible. 

Cx32 is a gap junction protein expressed in 
the paranodal region and Schmidt-Lanterm 
incus in the PNS, and in cell bodies and oligodendrocytes processes in the 
CNS. Why mutations in Cx32 usually lead 
only to CNS dysfunction is still an open 
question. Presumably, there is a unique rela-
tion between Cx32 and the structural organ-
isation or metabolic requirements of the 
CNS. Another possibility is that other conn-
exin proteins might compensate for Cx32 
dysfunction in the CNS and other tissues, but 
not in the PNS.
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 17p11.2 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 nerve 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

A 77 year old man admitted to our hospital had unsteady 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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Results of the autonomic nervous system investigations are given in the table. When the patient was tilted 45° on October 28, 2023 by guest. Protected by copyright.http://jnnp.bmj.com/ J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.66.6.804 on 1 June 1999. Downloaded from http://jnnp.bmj.com/ on October 28, 2023 by guest. Protected by copyright.

Isolated ischaemia of the spinal cord due to bilateral vertebral artery dissection

Clinical features in vertebral artery dissection are rarely associated with an ischaemic lesion of the spinal cord. The few cases related and studied with MRI strictly involve the cervical cord region. Isolated ischaemia of the spinal cord due to bilateral vertebral artery dissection can be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory could be provided from the arterial territory 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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. Sudo motor function was evaluated by sympathetic skin response (SSR) and local sweat response to acetyl-eholine (ACh). Before treatment, the SSR amplitude was decreased, and the number and area of sweat droplets were decreased in responses to intradermal ACh injection. The myelinated fibre density of biopsied sural nerve was 5927/mm². 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/mm²). Collagen pockets (15 000/mm²), and denervated Schwann cell subunits (12 000/mm²) were present, but their densities were within the normal range for his age. A highly sensitive acetylcholinesterase histochemical test (modified Tago's method) of the sural nerve detected a slightly reduced density of sudomotor sympathetic unmyelinated fibres (3500/mm²; normal 3700–6500/mm²). 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 lesion 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-

tion of the sympathetic postganglionic fibres without marked morphological change, although we cannot exclude the possibility that sympathetic neurons in the brainstem or spinal cord induce the dysfunction of postganglionic fibres by a trans-synaptic effect. Vitamin B12 is related to the methylation reaction regulated by S-adenosylhomocysteine and S-adenosylmethionine. This reaction has a crucial role in the myelin formation associated with neurological deficits in patients with vitamin B12 deficiency. Dysfunction in unmyelinated sympathetic neurons, however, has not been shown. Our findings suggest that vitamin B12 is required for the physiological function of sympathetic postganglionic fibres.

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

<table>
<thead>
<tr>
<th>Before treatment</th>
<th>After treatment</th>
<th>Age matched normal control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head-up tilting test</td>
<td>Supine</td>
<td>104</td>
</tr>
<tr>
<td></td>
<td>5 min after tilting</td>
<td>71</td>
</tr>
<tr>
<td></td>
<td>Supine</td>
<td>58</td>
</tr>
<tr>
<td></td>
<td>5 min after tilting</td>
<td>70</td>
</tr>
<tr>
<td></td>
<td>Supine</td>
<td>73</td>
</tr>
<tr>
<td></td>
<td>5 min after tilting</td>
<td>129</td>
</tr>
<tr>
<td></td>
<td>L palm</td>
<td>0.33</td>
</tr>
<tr>
<td></td>
<td>R palm</td>
<td>0.39</td>
</tr>
<tr>
<td></td>
<td>L Sole</td>
<td>0.67</td>
</tr>
<tr>
<td></td>
<td>R Sole</td>
<td>0.61</td>
</tr>
<tr>
<td></td>
<td>Local sweat response to acetylcholine</td>
<td>24</td>
</tr>
<tr>
<td></td>
<td>Total area of sweat droplets (mm²/cm²)</td>
<td>0.27</td>
</tr>
</tbody>
</table>

CORRESPONDENCE

Sandifer’s syndrome and gastro-oesophageal reflux disease

Perkin and Murray-Lyon’s Neurology and the gastrointestinal system reviews gastrointestinal disorders with neurological features.1 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-oesophageal reflux disease with spastic torticollics and dystonic body movements. Nodding and rotation of the head, neck extension, gurgling sounds, with-
Is inherited thrombophilia a risk factor for arterial stroke?

The paper of Ganesan et al adds the factor V Leiden 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.1,2 In their commentary on this paper, Brown and Bevan3 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 is 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.4,5 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 246 leucocytes/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 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 detected. Investigation of other disorders were normal. We concluded that our patient had neuroborreliosis and he was treated with ceftriaxone intravenously for 14 days. There was an almost complete recovery. The diagnosis of neuroborreliosis in our patient is supported by the clinical presentation with right hemiparesis, positive serology for Borrelia burgdorferi, the presence of IgG antibodies against Borrelia burgdorferi in the CSF, and the absence of antibiotic treatment. Based on the article by Oksi et al, it is very appealing to explain what happened in our patient by using their concept. Our patient had an aneurysm of the basilar artery. If cultitis is one of the patho-anatomical mechanisms in neuroborreliosis, it can also lead to formation of aneurysms or vascular infarction.

However, we postulate that the presence of the aneurysm in our patient was a coincidence. There are two other explanations for the xanthochromia through bilirubin in his CSF. The first is the raised protein content of the CSF (in a patient with protein C deficiency due to neuroborreliosis). Or, our patient had a vasculitis (supported by the pleiocytosis of the CSF and by the narrowing of the left anterior cerebral artery on angiogram) which can lead to subarachnoid haemorrhage with out the presence of an aneurysm, as was shown by Chehrenama et al.6

A causal relation between neuroborreliosis and the aneurysm is only based on circumstantial evidence. We do not agree that the reported cases of Oksi et al support this relation. Firstly, we think that only one of the three patients had neuroborreliosis. In the other two patients there was a pleocytosis or raised protein content in the CSF, a finding that is considered to be a necessity for the diagnosis of neuroborreliosis.7 Also, antibodies against Borrelia burgdorferi were not detected. Besides this, no evidence exists that in the one patient with neuroborrellosis and subarachnoid haemorrhage there is a causal relation with the aneurysm. He could indeed be one of those patients who happen to have an aneurysm.

For now, the answer to the question: “Intracranial aneurysms in three patients with disseminated Lyme borreliosis: cause or chance association?” should be chance association.

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samples using adequate techniques—for example, the polymerase chain reaction.

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BOOK REVIEWS


First impressions count, so it is important for a publisher to choose the right time to send a book to be reviewed. Unfortunately Butterworth-Heinemann’s timing for this book was awry. This volume arrived on my desk at the same time as I was struggling to improve a piece of software at home (which never refuses o...

A video is an excellent addition to any book on movement disorders. Unfortunately, the video refused to run on our modern video recorder at home (which never refuses offerings from the Disney corporation) and ran poorly on the state of the art equipment at Addenbrooke’s Hospital. Some of the clips were of poor quality—perfectly acceptable for very rare diseases but not for common conditions. The video covered the basics well and had some particularly florid examples of tics. More cross referencing between the book and video would have helped.

Despite its limitations I would recommend this book/video combination for the groups at whom it is aimed—mainly, primary care physicians and doctors in training. However, I thought that it might have been better written by a larger team, smaller, leaving the multimedia/multiauthor approach for more advanced textbooks, which aim to become

JERRY BROWN


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 functioning 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. The text and book 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 interested in neuroscience, although the way the book is packaged and presented would put most people off purchasing them, which is a shame.

Each chapter is clearly presented with high quality figures, which are often superior to those found in most medical neurobiology books. Furthermore, they often summarise key experimental results in a clear and concise fashion, and coupled to the well written prose, makes the book easy to understand and follow, if somewhat verbose. Questions are thrown in throughout the text and each chapter concludes with a summary, an objectives list, and a series of questions with a few key references cited. In addition, these books were impressive in their breadth of discussion within individual topics. For example, the pain section not only covers the conventional anatomical and pharmacological aspects of nociception but also discusses a range of antinociceptive treatments and strategies. As a result the reader is left with a much more balanced view than that which is traditionally presented in preclinical medical training and which is often at variance with that which is seen in clinical practice. It is this aspect of the books that is perhaps of most value and which could be applied to the new style of medical training currently being developed in this country. However constrained I must admit it 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.

ROGER BARKER


This is a book of 172 pages dedicated to the memory of Frank Morrell. It is a multiauthor text, originating largely from North America (with a notable United Kingdom contribution from the Maudsley Hospital). After a historical review including a description of recording techniques, novel approaches to using electrocoagulography 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. Electrocoagulography findings in extratemporal epilepsy are then dealt with, confirming that restricted frontal lobe abnorma...

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. However, the work in this book keeps its feet on the ground where necessary.

A comprehensive multicentre contribution follows, describing the findings in cortical dysplasia, and the way these probably incomprehensive results are then thrown into the pool of surgical experience when compared with patients with other structural lesions. There are then three chapters on studies in the mesial temporal region, involving patient selection, prognosis, and methodology. The multiauthor combination of acute and chronic electrocoagulography techniques. Some of this is then applied to a chapter on hemispherectomy.

The book finishes with a chapter on the applications of image guided surgery to...
Intraoperative electrophysiology, which is probably one of the most spicy contributions, confirming that a multimodal approach to the application of these investigations will probably be the most fruitful approach in the medium term. Those units contemplating similar work will find this book very useful in terms of selecting some of the techniques that they intend to include or exclude, with natural effects on their resources and clientele. Specialised units which already perform similar work will also find this a useful review. Inevitably this book will be of interest to a relatively selective readership, to whom it is thoroughly recommended.

SIMON BONIFACE


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 Tulio 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, on 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 neurologists, 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 second section reviews the different cognitive aspects and explores the role of neurodegenerative conditions in the understanding of organisation of memory. Executive functions in both subcortical and cortical dementia syndromes, episodic and semantic memory, and non-declarative memory are systematically covered. The discussion of disintegration of distinct memory systems in different degenerative conditions will be of interest to psychologists and doctors alike, although this section will be of special interest to neuropsychologists.

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 neurobiology of the dementias and other neurodegenerative conditions.

CLARE GALTON