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 photoclonulsive reaction with generalised spike wave activity may be provoked by IPS and is almost confined to patients with generalised epilepsy. There are, however, reports on patients with partial epilepsy and a photoconvulsive reaction. We report on two patients with known photoclonulsive 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 orolialimentary automatism, 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 T6, patient 1 showed provocation of anterotemporal (F8, patient 1) and posterotemporal (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 symptomology in the first patient included orolialimentary automatism, indicating a seizure origin in the amygdalohippocampal 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 F7, and the occurrence of a photoclonulsive reaction in patient 2 raises the question whether both phenomena were

**Spike activity during EEG registration in patient 1:** (A) without provocation; (B) while IPS.

**LETTERS TO THE EDITOR**

Spike activity during EEG registration in patient 1: (A) without provocation; (B) while IPS.
interrelated. Similar constellations were previously reported in individual patients with photoconvulsive 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 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 GABA-ergic 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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EEG on 4, 7, 10, and 19 June. It shows PSD on 7 June. The amplitude of the PSD is 150–200 µV and the frequency is 1.5–2 Hz.

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 it 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 hypotonus and hyporeflexia without Babinski’s sign. Serum glutamic oxaloacetic transami-

nase, 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. EEG 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, levomepromazine, and phenobarbitone.
Chlorpromazine, levomepromazine, and phe
obarbitone. 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 concentration after taking lithium for more than 1
week, and it is thought that a serum lithium con-
tcentration is toxic above 1.5 mmol/l, so her serum
concentration is likely to be high
eough to be toxic. Her symptoms such as for
getfulness, 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 EEG emerged 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
hypoactivity and hyporeflexia were uncom-
mon compared with previous reports. ”
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
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 ten-
don reflexes, and pes cavus are usually found in
males by the second decade, whereas in fem-

We describe a new Cx32 point mutation (Ala
Val) in genetically established identical twins with similar CMT phenotypes 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 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 diminished, 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 somatosensory potentials were all
absent. Twin 2 had identical clinical manifes-
tations, except that the left plantar reflex
was floxor 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
m/s, 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 re-
flexes were extensor. Her median and pero-
neal 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 grandmother was not examined,
but had a long history of a slowly progressive
europathy.

The presence of the 17p11.2–p12 duplica-
tion was excluded with the DNA sequencing
quantitative polymerase chain reaction with five
microsatellite markers contained within the
involved segment.

Sequencing Cx32 with the ABI TM
Primer Cycle Sequencing Ready Reaction
CMTX with the ABI TM Primer Cycle
Sense electropherogram of a segment of Cx32
exon 2 showing the C to T mutation at base pair 39 (upper trace) and the corresponding control
segment (lower trace).

Val amino acid substitution are clearly on the mild 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
showing of the central conduction time in
their brain stem auditory evoked potentials,’
but clinical manifestations secondary to cen-
tral dysfunction does not seem to be a frequent
finding. Paulson et al reported a patient who developed dysarthria and incor-
ded in high altitude skiing. His MRI showed
confluent, symmetric, white matter
changes. 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 fam-
ily with a mutation on code 93 whose clinical manifestations included tremor, brisk re-
flexes, 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-
parietal tract. Unfortunately no imaging or
evoked potential studies were possible.

Cx32 is a gap junction protein expressed in
the paranodal region and Schmidt–
Lanterman 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 rela-
tion between Cx32 and the structural organ-
isation or metabolic requirements of the
PNS. Another possibility is that other con-
nexin 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 muta-
tions 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 17p11.2 duplication there was remarkable clinical variability. 1,2 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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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. 1,2 The few cases related and studied with MRI strictly involve the cervical cord. 3,4 We add another patient with spontaneous bilateral vertebral artery dissection in whom the particularity was an isolated extensive ischaemia of the spinal cord from C4 to T5 vertebral levels.

A previously healthy 45 year old woman had paresthesiae of the right ankle lasting a few days. Ten days later, she had a right sided scapulohumeral pain and suddenly developed 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-Sèquard’s syndrome.

Biological investigations were normal. CSF protein was 0.34 g/l, glucose 2.54 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 extending 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 vasculatisation could be provided from the arterial cervical cord region. 5 The bilateral ischaemic lesions extending through several cervical and dorsal segments are in favour of watershed infarcts caused by hyperfusion due to bilateral vertebral artery dissection.

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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 catecholamine 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 concentra-
tion was reduced at rest, and its increase after tilting up was minimal. Suddomotor function 
was evaluated by sympathetic skin response (SSR) and local sweat response to acetylcholine 
(Ach).1-4

Before treatment, the SSR amplitude was decreased, and the number and area of sweat 
droplets were decreased in responses to tilt up was minimal. Sudomotor function 
over 30 mm Hg with normal heart rate 
Letters, Correspondence, Book reviews

was evaluated by sympathetic skin response 

SSR was evaluated by sympathetic skin response

(about 20%) and axonal degeneration (about 
10%). Electron microscopy showed a normal 
unmyelinated fibre density (30 945/mm²). Collagen pockets (15 000 /mm²), and dener-
vated Schwann cell subunits (12 000 /mm²) were present, but their densities were within 
the normal range for his age.1

A highly sensitive acetylcholinesterase 
(AChE) histochemical test (modified Tago’s method)5 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 
return of heart rate response in head up tilt test.

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 pre-
served 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, al-
though we cannot exclude the possibility that 
sympathetic neurons in the brainstem or spi-
nal cord induce the dysfunction of postgang-
ligonic fibres by a trans-synaptic effect. 1-4

Vitamin B12 is related to the methylation 
reaction regulated by S-adenosylhomocystine 
and S-adenosylmethionine.1 This reaction 
has a crucial role in the myelin formation 
process in the nervous system.1-5 Deficient 
catecholamine release as the basis of 
orthostatic hypotension in pernicious anemia. J 

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

<table>
<thead>
<tr>
<th>Head-up tilting test</th>
<th>Before treatment</th>
<th>After treatment</th>
<th>Age matched normal control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systolic blood pressure (mm Hg)</td>
<td>Supine</td>
<td>104</td>
<td>106</td>
</tr>
<tr>
<td></td>
<td>5 min after tilting</td>
<td>71</td>
<td>93</td>
</tr>
<tr>
<td></td>
<td>15 min after tilting</td>
<td>76</td>
<td>106</td>
</tr>
<tr>
<td>Heart rate (/min)</td>
<td>Supine</td>
<td>58</td>
<td>66</td>
</tr>
<tr>
<td></td>
<td>5 min after tilting</td>
<td>70</td>
<td>73</td>
</tr>
<tr>
<td></td>
<td>15 min after tilting</td>
<td>73</td>
<td>76</td>
</tr>
<tr>
<td>Noradrenaline (pg/ml)</td>
<td>Supine</td>
<td>94</td>
<td>129</td>
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<tr>
<td></td>
<td>5 min after tilting</td>
<td>286</td>
<td>258 - 752</td>
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<tr>
<td></td>
<td>Sympathetic skin response Amplitude (mV)</td>
<td>L palm</td>
<td>0.33</td>
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<tr>
<td></td>
<td>R palm</td>
<td>0.39</td>
<td>1.0</td>
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<tr>
<td></td>
<td>L Sole</td>
<td>0.67</td>
<td>1.9</td>
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<tr>
<td></td>
<td>R Sole</td>
<td>0.61</td>
<td>1.2</td>
</tr>
<tr>
<td>Local sweat response to acetylcholine</td>
<td>Number of sweat droplets (/cm²)</td>
<td>24</td>
<td>69</td>
</tr>
<tr>
<td></td>
<td>Total area of sweat droplets (nm²/cm²)</td>
<td>0.27</td>
<td>2.86</td>
</tr>
</tbody>
</table>

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sion associated with pernicious anemia: report 
of a case with complete recovery following vita-
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Sympathetic skin response in patients with 
multiple sclerosis compared with spinal cord 
transsection and normal control. Brain 1991; 
114:1381–94.

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 torticollis and dystonic body movements. Nodding and rotation of the 
head, neck extension, gurgling sounds, with- 
ring movements of the limbs, and severe hypo-
tonia have been reported.1,2 It is hypothesised 
that such positioning provides relief from discomfort caused by acid reflux.2 A causal 
relation between gastro-oesophageal reflux disease and the neurological manifestations of 
Sandifer's syndrome is supported by the reso-
lution of the manifestations on successful 
treatment of gastro-oesophageal reflux dis-
case.2,5 The clinical manifestation for almost 
 invariably arouse the suspicion of neurological 
disease and lead to unnecessary investigative 
procedures.2 The intermittent occurrence of 
torticollis with alternating directions, normal 
stenocleidomastoid muscles, and normal cer-
vical radiographic findings make Sandifer's 
syndrome a probable diagnosis and necessitate 
upper gastrointestinal studies.2,5

Most patients have no other abnormalities. 
Sandifer's syndrome is supported by the rela-
tively large number of patients with brain 
damage or metabolic disorders and is often 
interpreted as a feature of their basic dis-
order.1 A high prevalence of Sandifer's 
syndrome was reported in the Beuchman-de 
Lange syndrome.3 These findings may simply 
reflect the high prevalence of gastro-
oesophageal reflux disease among brain 
damaged children rather than a primary feature of these disorders.3

Early recognition and treatment of gastro-
oesophageal reflux disease in patients with 
Sandifer's syndrome enhances the success of 
medical management, curative for patients 
with no other disorders, and contributes to 
 improved quality of life for patients with brain 
damage.1,2

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CORRESPONDENCE
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 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 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 19 leucocytes/mm³; the protein content was 3.49 g/l. The IgG index was raised to 1.35. The CSF was xanthochromic, became of bilirubin. IgG antibodies against Borrelia burgdorferi in 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 detected. Investigations for other disorders were normal. We concluded that our patient had neuroborreliosis and he was treated with ceftriaxon intravenously for 14 days. There was an almost complete recovery.

The diagnosis of neuroborreliosis in this 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 another antibiotic treatment. Based on the article by Oksi et al,6 it is very appealing to explain what happened in our patient by using their treatment. Our patient had an aneurysm of the basilar artery. If culturing is one of the usual pathological 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 our CSF. The first is the raised protein content of the CSF (in a patient with meningitis 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.7

A causal relation between neuroborreliosis and the aneurysms 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 pleiocytosis or raised protein content in the CSF, a finding that is considered to be a necessity for the diagnosis of neuroborreliosis.8 Also, antibodies against Borrelia burgdorferi were not detected. Besides this, no evidence exists that in the one patient with neuroborreliosis 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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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—namely, primary care physicians and doctors in training. However, I thought that it might have been better written by a far smaller team, leaving the multidisciplinary/multiauthor approach for more advanced textbooks, which aim to become the definitive works on a subject.

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BOOK REVIEWS

Differential Diagnosis and Treatment of Movement Disorders (Book and Video Set). Edited by EDUARDO TOLOSA, WILLIAM C KOLNER, and OSCAR S GERSHANIK (£90.00). Published by Heinemann, Oxford, 1998.

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 patient’s primary orthostatic tremor in time for her daughter’s wedding in Australia. I was therefore disconcerted when I could find no reference to this disease in the index under primary, orthostatic, or tremor. I turned to the book on tremor and eventually found a single inadequate sentence describing it as a clinical variant of essential tremor and a reference to clonazepam as the first line treatment. Happily, the book passed my second test—treatment options for a patient with tardive dyskinesia.

With longer acquaintance the book is more impressive. There are some good reviews and key references listed, and a series of questions with a few


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 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 who are 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


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 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, 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