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 which may prove the epileptic nature and specify epileptic syndromes.

A photoconvulsive reaction with generalised spike wave activity may be provoked by IPS and photoconvulsive reaction. 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 generalised tonic-clonic seizure. 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 intermittent spikes showing 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-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 (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 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 F8 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 photovoltaic 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 photovoltaic reaction, which is supported by the studies of Ricci et al using neuromagnetic methods in humans with photovoltaic reaction to identify the location of the photovoltaic 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 photovoltaic reaction is a generalised phenomenon and not due to multifocal 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 photovoltaic reaction as the second appeared in only one of the patients.

S SEDDIGH
F THÖMKE
TH VOGT

Department of Neurology, University of Mainz, Germany

Correspondence to: Correspondence to: Dr Sussend, Neurologische Klinik und Poliklinik, Langenbeckstrasse 1, D 55101 Mainz, Germany.

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

Creutzfeld-Jakob-like syndrome was first reported by Smith and Kocen in 1988. Its symptoms resemble Creutzfeld-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 Creutzfeld-Jakob disease, but this returns to normal when the patient recovers.

There have been some case reports of Creutzfeld-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 Creutzfeld-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 hyporeflexia without Babinski’s sign. Serum glutamic oxaloacetic transaminase, glutamic pyruvic transaminase alkaline phosphatase, and creatine kinase was 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.
chlorpromazine, levomepromazine, and phe- 
nobarbital. Her CSF lithium concentration was 0.82 mmol/l on 4 June. According to Taguchi et al.,7 lithium concentration in CSF is about one fourth of the serum concentra- 
tion after taking lithium for more than 1 week, and it is pointed out 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, 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 after taking lithium for more than 1 week 
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 phenobarbital. We could not identify the role of phenobarbital injected from 4 to 6 June, but it might be possible it have some part in the induction of PSD, and her hypotonia and hyporeflexia are uncom-
mon compared with previous reports.8,9 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.1

HIDEYUKI KIKYO
Shinmaro 201, 3–12–8 Negishi, Taito-ku, 
Tokyo 116, Japan

TETSUO FURUKAWA
1–5–45 Yushima Bunkyo-ku, Tokyo 113, Japan

Correspondence to: Dr Hideyuki Kikyo, Depart-
ment of Neurology, Tokyo Medical and Dental 
University, Tokyo, Japan. email: kikyo@tmu.
tokyo.ac.jp

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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.1,2 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 
axonal 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 
fected males. Nerve conduction studies in 
fected males are usually, but not always, 
suggestive of a demyelinating process, 
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 neuropathy.

We describe a new Cx32 point mutation 
(Ala34 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 absent or depressed, 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 manifesta-
tions, 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 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 genetic analysis was not examined, 
but had a long history of a slowly progressive 
neuropathy.

The presence of the 17p11.2-p12 duplication 
was excluded by fluorescence in situ 
hybridization and the corresponding quantita-
tive polymerase chain reaction with five micro-
satellite 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 BsrGI and oligonucleotide 200 con-
trol chromosomes and the father’s DNA, no 
mutation was found. The mother was shown 
to harbour the mutation.

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

CMTX is now recognized as a frequent 
case of HMSN.3 Mutations have been detected 
in all domains of the protein and are 
postulated to be either non-functional or 
excite a dominant-negative effect. The clinical 
manifestations detected in this family with a 
novel point mutation leading to an Ala34 to 
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 planter responses in 
in all three people shown to carry the mutation. 
Involvement of the CNS in patients with 
CMTX mutations have been demonstrated 
by slowing of the central conduction time in 
their brain stem auditory evoked potentials,4,5 
but clinical manifestations secondary to cen-
tral dysfunction does not seem to be a frequent 
finding. Paulson et al.6 reported a patient who 
developed dystarthis and incoordination 
after high altitude skiing. His MRI showed 
confluent, symmetric, white matter 
changes. Another member of the family 
carrying the mutation had normal MRI7 
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.8 presented a 
family with a mutation on code 93 whose 
clinical manifestations included tremor, bril-
less, 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-
 spinal tract. Unfortunately no imaging or 
evoked potential studies were possible.

Cx32 is a gap junction protein expressed 
in the paranoid region and Schmidt-
Launieran incisures in the PNS, and in cell 
boils 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-
ization or metabolic requirements of the 
PNS. Another possibility is that other con-
exin 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. 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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W MARQUES JR M G SWEENEY N WOOD
University Department of Clinical Neurology, Institute of Neurology, Queen Square, London, UK
S J WROE
Department of Clinical Neurology, Ipswich Hospital NHS Trust, Ipswich, UK
W MARQUES
Department of Neurology, School of Medicine of Ribeirão Preto, Brazil
Correspondence to: Dr Nicholas Wood, Institute of Neurology, Queen Square, London WC1N 3BG, UK.
Telephone 020 7387 3611 ext 4559; fax 0044 171 278 5616.


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. 1-3 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 paresthesia of the right ankle lasting a few days. Ten days later, she had a right sided scapulothoracic 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 spinthalamic 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 0.53 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 signal high 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 cervical cord. 4,5 The bilateral ischaemic lesions extending through several cervical and dorsal segments are in favour of watershed infarcts caused by hypoperfusion due to bilateral vertebral artery dissection.

P GARNIER D MICHEL R PETRON O BEAUCHET
Department of Neurology
F LE BRAS F G BARRAL
Department of Neuroradiology, University Hospital Saint-Etienne, 42055 Saint-Etienne, France
Correspondence to: Dr D Michel, Service de Neurologie, Hôpital de Bellevue, 42055 Saint-Etienne, Cedex 2, France.


Autonomic dysfunction and orthostatic hypotension caused by vitamin B12 deficiency

Orthostatic hypotension sometimes is a reversible neurological complication of vitamin B12 deficiency. 6,7 Eisenhofer detected deficient sympathetic norepinephrine release in insulin tolerance testing, 8 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 lower limbs, 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 autonomic nervous system tests before and after vitamin B12 treatment

\[ \text{Table:} \]

<table>
<thead>
<tr>
<th>Test</th>
<th>Before Vitamin B12</th>
<th>After Vitamin B12</th>
<th>Improvement</th>
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<tbody>
<tr>
<td>Systolic blood pressure (mm Hg)</td>
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<td></td>
<td></td>
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<tr>
<td>Head-up tilting</td>
<td>104</td>
<td>106</td>
<td>112 - 135</td>
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<tr>
<td>Low back tilting</td>
<td>71</td>
<td>93</td>
<td>118 - 140</td>
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<tr>
<td>Heart rate (bpm)</td>
<td>5 min after tilting</td>
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<td>66</td>
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<tr>
<td>Noradrenaline (pg/ml)</td>
<td>5 min after tilting</td>
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<td>73</td>
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<tr>
<td>Sympathetic response</td>
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<td>Amplitude (mV)</td>
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<tr>
<td></td>
<td>R palm</td>
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<tr>
<td></td>
<td>R Sole</td>
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<td>Local sweat response to acetylcholine</td>
<td>Number of sweat droplets (cm²/cm²)</td>
<td>24</td>
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<td></td>
<td>Total area of sweat droplets (mm²/cm²)</td>
<td>0.27</td>
<td>2.86</td>
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</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 torticollis and dystonic body movements. Nodding and rotation of the head, neck extension, gurgling sounds, withstanding movements of the limbs, and severe constipation have been reported.4,5 It is hypothesised that such positioning provides relief from discomfort caused by acid reflux.4 A causal relation between gastro-oesophageal reflux disease and the neurological manifestations of Sandifer's syndrome is supported by the resolution of the manifestations on successful treatment of gastro-oesophageal reflux disease.4,6 The clinical manifestation of Sandifer syndrome enhances the success of medical management, is curative for patients with no other disorders, and contributes to improved quality of life for patients with brain damage or metabolic disorders and is often interpreted as a feature of their basic disorder.1 A high prevalence of Sandifer's syndrome was reported in the Beacham-de Lange syndrome.1 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.4 Early recognition and treatment of gastro-oesophageal reflux disease in patients with Sandifer's syndrome enhances the success of medical management, is curative for patients with no other disorders, and contributes to improved quality of life for patients with brain damage.1,4

DEMETRIOS S THEODOROPOULOS

RICHARD F LOCKEY

Division of Allergy and Immunology

H WORTH BOYCE, JR

Center for Stuttering Disorders, Department of Internal Medicine, University of South Florida College of Medicine, and James A Haley Veterans Hospital, 13000 Bruce B Downs Boulevard, Tampa, Florida 33612, USA

Correspondence to: Dr DS Theodoropoulos, Division of Allergy and Immunology, Department of Internal Medicine, University of South Florida College of Medicine and James A Haley Veterans Hospital (111D), 13000 Bruce B Downs Boulevard, Tampa, Florida 33612, USA.
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 Bevan4 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.5-7 It would therefore seem prudent to follow concentrations of protein C and protein S for at least this time period before concluding that any patient can be attributed to an inherited thrombophilia, particularly if the presence of such a disorder is to be managed by “lifelong anticoagulation”.

C R KENNEDY
Department of Paediatric Neurology, Southampton General Hospital, Southampton, UK


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. They 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 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, the presence of IgG antibodies against Borrelia burgdorferi in the CSF, and the finding of antitoxic 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. We do not agree that the 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 finding of antitoxic antibiotic treatment. In their commentary on this paper, 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. No evidence in support of this recommendation is cited.

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C R KENNEDY
Department of Paediatric Neurology, Southampton General Hospital, Southampton, UK

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

JARMO OKSI
Department of Internal Medicine, Turku University Central Hospital and Department of Medical Microbiology, Turku University, Finland

HANNU KALIMO REIJIO JMARTTILA MERJA MARJAMÄKI PIRKKO SONNINEN JUKKA NIKOSKELAINEN MATTI K VIJLANEN
Turku University, Turku University Central Hospital, and National Public Health Institute, Department in Turku, Finland


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/multi-author approach for more advanced textbooks, which aim to become the definitive works on a subject.

JERRY BROWN


This is a book of 172 pages dedicated to the memory of Frank Mellor. It is a multi-author text, originating largely from North America (with a notable United Kingdom contribution from the Maudsley Hospital). After a historical review including descriptions of diagnostic and recording techniques, novel approaches to using electrocardiography 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. Electrocardiography findings in extratemporal epilepsy are then dealt with, confirming that restricted frontal lobe abnorma lities predict a favourable outcome, particularly when combined with a well defined structural lesion. The technique of chronic electrocardiography is also reviewed, including demonstration of how it may be used 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. The book keeps its feet on the ground where necessary.

A comprehensive multicentre contribution follows, describing the findings in cortical dysplasia, and the way theory probably diverges from 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, volumetric MRI, and the combination of acute and chronic electrocorticography 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, 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