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 epileptogenic potentials, which may prove the epileptic nature and specify epileptic syndromes. A photokinetic reaction with generalised spike wave activity may be provoked by IPS and is almost confined to patients with generalised epilepsy. There are, however, some reports on patients with partial epilepsy and specifc stimuli like rubbing, cold wind, tactile stimuli may evoke spike activity 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 longitudinais inferius.

Spike activity during EEG registration in patient 1: (A) without provocation; (B) while IPS.
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 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 photovoltaic 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. It's symptoms resemble Creutzfeldt-Jakob disease but it is induced by drugs, particularly lithium, and most patients recover without sequel after discontinuation of drugs. It also displays a characteristic EEG similar to Creutzfeldt-Jakob disease, but this returns to normal when the patient recovers. There have been some case reports of Creutzfeldt-Jakob-like syndrome after that of Smith et al (table), but no paper seems to have described the detailed course of EEG changes. This paper presents a case of Creutzfeldt-Jakob-like syndrome possibly induced by lithium, levomepromazine, and phenobarbitone, in which we succeeded in recording the course of EEG changes.

A 65 year old woman was admitted to a hospital with coma and myoclonus. She had a history of manic and depressive disease for 8 years and had been treated with 200 mg lithium carbonate, 25 mg chlorpromazine, and 10 mg levomepromazine daily. Her first symptom was forgetfulness from 20 May, then she complained of appetite loss from 27 May, diarrhea 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, levodopa/benserazide.

<table>
<thead>
<tr>
<th>Drugs (other than lithium)</th>
<th>Authors</th>
<th>Age/sex</th>
<th>SL</th>
<th>PD</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>Smith et al</td>
<td>69/F</td>
<td>T (+)</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Primavera et al</td>
<td>50/F</td>
<td>T (+)</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Primavera et al</td>
<td>56/M</td>
<td>NT (+)</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>Takahashi et al</td>
<td>67/F</td>
<td>T (+)</td>
<td></td>
</tr>
<tr>
<td>Levodopa</td>
<td>Broussolle et al</td>
<td>70/F</td>
<td>NA (+)</td>
<td></td>
</tr>
<tr>
<td>Lorazepam</td>
<td>Cassanova et al</td>
<td>67/M</td>
<td>NT (+)</td>
<td></td>
</tr>
<tr>
<td>Norropetidine</td>
<td>Finelli</td>
<td>60/M</td>
<td>NT (+)</td>
<td></td>
</tr>
<tr>
<td>Phenobarbitone, levomepromazine</td>
<td>This case</td>
<td>65/S</td>
<td>T (+)</td>
<td></td>
</tr>
<tr>
<td>Madpar* others</td>
<td>Smith et al</td>
<td>72/F</td>
<td>NT (+)</td>
<td></td>
</tr>
<tr>
<td>Bromazepam, trihexyphenidyle, others</td>
<td>Masmoudi et al</td>
<td>66/F</td>
<td>NT</td>
<td>Pseudoperiodic</td>
</tr>
</tbody>
</table>

Li=lethium, SL=serum lithium concentration; PD=periodic discharge, T=toxic, NT=not toxic, NA=not assessed, Madpar=levodopa/benserazide.
Central nervous system involvement in a novel connexin 32 mutation affecting identical twins

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

At least 130 different mutations have been reported in the Cx32 gene causing peripheral neuropathy. Classically, distal weakness and atrophy initially involving the lower limbs, as well as sensory abnormalities, depressed tendon reflexes, and pes cavus are usually found in males by the second decade, whereas 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, although they are not quite as slow as in patients with CMT1A. In females, conduction velocities (CVs) may be in the normal range or only mildly reduced, as seen in axonal neuropathies.

We describe a new Cx32 point mutation (Ala175Val) 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 intrinsic tendinous contracture 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 decreased, but both plantar responses were extensor. His median, ulnar, and peroneal motor CVs were 33.0 m/s, 33.0 m/s, and 31.0 m/s, respectively, and the distal amplitudes were 0.7 mV, 5.0 mV, and 3.3 mV. The sensory potentials were all absent. Twin 2 had identical clinical manifestations, except that the left plantar reflex was flexor whereas the right was clearly extensor. His motor CVs and amplitudes of the same nerves described above were 30.0 m/s and 1.7 mV, 34.0 m/s and 6.0 mV, and 33.0 m/s and 4.0 mV, respectively. No sensory response was obtained. Their mother had minimal neuropathic features and both plantar reflexes were absent. Her median and peroneal motor CVs were 43.0 m/s and 37.0 m/s, and the median sensory CV was 40.0 m/s. Their sister and the mother’s brother were clinically and electrophysiologically normal. The maternal genetic study was not examined, but had a long history of a slowly progressive neuropathy.

The presence of the 17p11.2-p12 duplication was excluded. Gower et al2 performed a subsequent quantitative polymerase chain reaction with five microsatellite markers contained within the involved segment. Sequencing Cx32 with the ABI® Dye Primer Cycle Sequencing Ready Reaction detected a C→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 BbvI and oligotyping 200 control 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 determination 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 cause of HMSN.3 Mutations have been detected in all domains of the protein and are postulated to be either non-functional or exert a dominant-negative effect. The clinical manifestations detected in this family with a novel point mutation leading to an Ala175Val 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 chromosomes 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 slowing of the central conduction time in their brain stem auditory evoked potentials,4 but clinical manifestations secondary to central dysfunction does not seem to be a frequent finding. Paulson et al5 reported a patient who developed dysarthria and incoordination after high altitude skiing. His MRI showed confluence, symmetric, white matter changes. Another member of the family carrying the mutation had normal MRI6 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 al7 presented a family with a mutation on code 93 whose clinical manifestations included tremor, brisk reflex and spasticity. On MRI there was atrophy 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 corticospinal tract. Unfortunately no imaging or evoked potential studies were possible.

Cx32 is a gap junction protein expressed in the paranodal region and Schmidt-Lantermann incisures in the PNS, and in cell bodies and oligodendrocytes processes in the CNS. Why mutations in Cx32 usually lead only to PNS dysfunction is still an open question. Presumably, there is a unique relation between Cx32 and the structural organization of the PNS. Another possibility is that other connexin proteins might compensate for Cx32 dysfunction in the CNS and other tissues, but this remains to be investigated.

Although the clinical manifestations are extremely similar in most of the Cx32 neuropathies suggesting that different mutations do not cause different phenotypes, different degrees of severity and the presence of unusual signs, like the one we present here,
have already been described to occur with some mutations. There are only two previous reports relating to three pairs of identical twins with CMT and known genetic defects. In the two pairs with the 1p11.2 duplication there was remarkable clinical variability. We have also seen a pair of identical twins with a P0 mutation in whom there was marked variability in early ages (unpublished data). Apart from the asymmetry of toe responses in one of the probands, the genetically identical twins described here are phenotypically very similar, suggesting that the expression of this mutation was not influenced by other non-genetic factors.

Coden 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.

WMIR was supported by grants from CAPES, FAPESP, and FAEPB (Brazil).

#### Clinical features in vertebral artery dissection

We add another patient with spontaneous bilateral vertebral artery dissection. Some authors state that this condition does not seem to depend on non-genetic factors.

There are only two previous reports concerning bilateral vertebral artery dissection in patients with a 17p11.2 duplication there was manifestation does not seem to depend on non-genetic factors, as might be expected in a hemizygous condition.

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

Orthostatic hypotension sometimes is a reversible neurological complication of vitamin B12 deficiency. 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 distal parts of all the limbs. Deep tendon reflex was absent in the lower limbs, and Babinski’s sign was positive bilaterally. Mild limb ataxia was seen in the four limbs, and Romberg’s sign was positive.

Haematological studies disclosed mild macrocytic hyperchromic anaemia (haemoglobin 14.0 g/dl, mean corpuscular volume 104 fl, mean corpuscular haemoglobin concentration 35.2 pg), with a few (3%) hypersegmented polymorphonuclear cells. His serum vitamin B12 concentration was markedly decreased (38 pg/ml; normal 249–938 pg/ml). Intrinsinc factor and parietal cell antibodies were positive in the serum. Echo cardiography showed no evidence of heart failure. In a study of peripheral nerve conduction, amplitudes of sensory nerve action potentials were slightly decreased in the lower limbs. The somatosensory evoked potential on median nerve stimulation showed a moderately prolonged central conduction time. Urodynamic studies disclosed uninhibited neurogenic bladder with detrusor sphincter dyssynergia.

Results of the autonomic nervous system tests before and 6 months after treatment are shown in the table. When the patient was tilted up to 60 degrees, he experienced dizziness and a significant fall in systolic blood pressure.
over 30 mm Hg with normal heart rate response. His serum noradrenalin concentration was reduced at rest, and its increase after tilting up was minimal. Sudomotor function was evaluated by sympathetic skin response (SSR) and local sweat response to acetylcholine (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 (AChE) histochemical test (modified Tago’s) on 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 noradrenalin 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 noradrenalin 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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**CORRESPONDENCE**

**Sandifer’s syndrome and gastro-esophageal reflux disease**

Perkin and Murray-Lyon’s Neurology and the gastrointestinal system reviews gastrointestinal disorders with neurological features. 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 a condition which may lead to unnecessary investigative procedures. The intermittent occurrence of torticollis with alternating directions, normal sternocleidomastoid muscles, and normal cervical radiographic findings make Sandifer’s syndrome a probable diagnosis and necessitate further evaluation.

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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 arteri-also in this commentary on our paper, Brown and Bevan avoid this terminology. There is no evidence in sup-port 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 seem prudent to follow concentrations of antithrombin 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. It would therefore seem prudent not 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”.

C R KENNEDY
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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 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 enhance-ment with contrast. Examination of CSF disclosed 30 leucocytes/mm³; the protein content was 3.49 g/l. The IgG index was raised to 1.35. The CSF was xanthochromatic but could not do the same with the aneurysm of the anterior cerebral artery. They do not have any evidence that excludes the possibility that the aneurysm 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.

JOSÉ D POLET
HENDRI C WEINSTEIN
Department of Neurology, St. Lucas Andreas Hospital, Amsterdam, The Netherlands

Oksi et al reply:

As a reply to the comments by Polet and Weinstein on our article “Intracranial aneurysms in three patients with disseminated Lyme borreliosis: cause or chance association?” we present the following comments.

Polet and Weinstein consider that only one of our three patients had neuroborreliosis. In our article we do not claim that all the patients had neuroborreliosis. However, all three patients had disseminated Lyme borreliosis based on the detection of Borrelia burgdorferi DNA in plasma samples of two patients, and an intrathecal production of antibodies against Borrelia burgdorferi in the third patient. It is also relevant to raise the question: what is neuroborreliosis? Does it require invasion of the spirochetes to none, or several of the following: cerebral tissuaries, meninges, CSF, or cranial (or peripheral) nervos system. The authors state that an increased cell count or increased protein content is a necessity for neuroborreliosis, referring to the article by Garcia-Monco and Benach. However, this is not always the case as shown, for example in the studies by Lu and Coyle in which they describe patients with neuroborreliosis hav-ving DNA or antigens of Borrelia burgdorferi in CSF despite normal routine CSF analyses and absence of antibodies against Borrelia burgdorferi in CSF.

Polet and Weinstein present a case of a patient with an intracranial aneurysm and suspected neuroborreliosis. They write that antibodies against Borrelia burgdorferi were detected in the CSF. However, the total IgG was increased, and they do not state whether the antibodies were intrathecal or not. If we understood correctly, the authors think that, for some reason, could associate the focal narrowing of the left anteri-or cerebral artery with Borrelia burgdorfer but could not do the same with the narrowing of the basilar artery. They do not have any evidence that excludes the possibility that the aneurysm could also have been caused by Borrelia burgdorferi. As we refer to in our arti-cle, high numbers of inflammatory cells have been found in aneurysms. We consider that it is possible that the aetiology of inflammatory changes in both instances may be the same—although probably seen at a different time point. We and others have found that disseminated Lyme borreliosis is be associ-ated with cerebral vasculitis.

Polet and Weinstein give a straightforward answer to the question presented in the title of their article. However, that it is not yet possible to give an answer to this question. The previously known association of another spirochetal infection, syphilis, to aneurysm formation, indicates that there might be a causal relation between Borrelia burgdorferi infection and aneurysm formation. This possible causal relation is a hypothesis that could be tested in the future in a larger number of patients with coexisting Lyme disease and aneurysm. Definite answers would require analysis of a sufficient number of aneurysm


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 offers from the Disney corporation) and ran poorly on the state of the art equipment at Addenbrooke’s Hospital, Cambridge. 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 medical students. However, I thought that it might have been better written by a far smaller team, leaving the multimedia/multiauthour approach for more advanced textbooks, which aim to become definitive works on a subject.

JERRY BROWN


This is a book of 172 pages dedicated to the memory of Frank Morrell. It is a multiauthour text, originating largely from North America (with a notable United Kingdom contribution from the Maudsley Hospital). After a historical review including stimulation and recording techniques, novel approaches to using electroencephalography 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. Electroencephalography findings in extratemporal epilepsy are then dealt with, confirming that restricted frontal lobe abnormalities predict a favourable outcome, particularly when combined with a well defined structural lesion. The technique of chronic electroencephalography is also reviewed, including the demonstration of how parallel approaches can be applied to a chapter on hemispherectomy.

A comprehensive multicentre contribution follows, describing the findings in cortical dysplasia, and the way these probably limit the applications of image guided surgery to the surgical outcome when compared with patients with other structural lesions. There are then three chapters on studies in the mesial temporal region, involving patient selection, prognosis, volume, and 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...
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
Autonomic dysfunction and orthostatic hypotension caused by vitamin B12 deficiency

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