Reversible proximal myopathy in epilepsy related Cushing’s syndrome

Epilepsy can induce hypercortisolism secondary to altered temporolimbic modulation of the hypothalamic-pituitary secretory of ACTH.1–3 Hypercortisolism can produce a reversible myopathy. Epilepsy, however, has not been recognised to cause a symptomatic Cushing’s syndrome, in particular, has not been reported. We report on three such patients as well as their successful treatment with normalisation of serum cortisol using ketoconazole.3

Three women (table) with epilepsy who complained of easy fatigability, had mild to moderate weakness of trunk, neck, and upper and lower limb girdle muscles. All had irregular menses and hirsutism. They had increased serum ACTH concentrations and serum and 24 hour urinary cortisol concentrations. Weakness was persistent in two and intermittent, correlating exclusively with periods of seizure exacerbation and raised cortisol secretion in patient 3. In this patient, seizures would flare up every few months. At such times, she would experience daily to weekly seizures for one to three months with development of fatiguability and clinically documented weakness after two or three weeks. EMG showed myopathic changes characterised by normal resting activity, small units, and increased recruitment patterns in all three patients. EMG findings were normal between exacerbations in patient 3 on two out of two occasions. Deltoid muscle biopsy was carried out in patient 2 and showed mild non-specific myopathic changes with type 1 predominance. Dexamethasone suppressed ACTH and cortisol secretion in all patients. MRI studies of the pituitary and adrenal glands showed no evidence of tumour. Thyroid function tests, and gonadal steroid glands showed no evidence of tumour. Those findings raise the possibility that epilepsy may be the basis for a hypercortisolism induced proximal myopathy that is readily reversible by normalisation of cortisol concentrations using ketoconazole. Normalisation of cortisol may benefit epilepsy as well.

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Summary of patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td>32</td>
<td>43</td>
<td>31</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>CPS/SGS</td>
<td>CPS/SGS</td>
<td>CPS</td>
</tr>
<tr>
<td>EEG</td>
<td>L and R temp foci</td>
<td>R temp focus</td>
<td></td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>Phenyltoin phenobarbionate</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Irregular menses</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hirsutism</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Obesity</td>
<td>Mild</td>
<td>–</td>
<td>Moderate</td>
</tr>
<tr>
<td>Striae</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Weakness</td>
<td>Mild persistent</td>
<td>Moderate persistent</td>
<td>Moderate intermittent</td>
</tr>
<tr>
<td>Cortisol: Serum (pm) (1.4–10.7 µg/dl)</td>
<td>19.8–31.0</td>
<td>11.0–15.0</td>
<td>25.1–29.9</td>
</tr>
<tr>
<td>Urinary (20–90 µg/24h)</td>
<td>70.3–106.0</td>
<td>57–102</td>
<td>123–183</td>
</tr>
<tr>
<td>AlTH (20–80 pmol)</td>
<td>57–124</td>
<td>76.5–142.5</td>
<td></td>
</tr>
<tr>
<td>Dex supp (&lt;1 µg/dl)</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>EMG</td>
<td>Myopathic</td>
<td>Myopathic</td>
<td>Myopathic</td>
</tr>
<tr>
<td>Biopsy</td>
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<td>Non-specific myopathic</td>
<td>Not done</td>
</tr>
<tr>
<td>Treatment</td>
<td>Ketoconazole 200 mg four times daily</td>
<td>Ketoconazole 200 mg three times daily</td>
<td>Ketoconazole 200 mg three times daily</td>
</tr>
<tr>
<td>Follow up</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Myopathy</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>St freqn</td>
<td>92%</td>
<td>Seizure free</td>
<td></td>
</tr>
<tr>
<td>Menses</td>
<td>Regular</td>
<td>Hysterectomy for fibroids</td>
<td>Regular</td>
</tr>
</tbody>
</table>

CPS=complex partial seizures; SGS=secondary generalised seizures; L=left; R=right; temp=temporal; irregular menses=irregular or <25 or >32 day interval menstrual cycles; weakness: mild->4.5/5 strength, moderate->4/5 strength; Dex supp=dexamethasone suppression test; st freqn=seizure frequency during follow up period compared with baseline.


Serial magnetic resonance imaging shows separate medial and lateral medullary infarctions resulting in the hemimedullary syndrome

There have been several reports on MRI of the classic medullary vascular syndromes such as dorsolateral infarction and medial infarction.4 The syndrome with both dorsolateral and
medial infarctions is known as hemimedullary (Babinski-Nageotte) syndrome. However, because of the separate arterial topography supplying the medulla, the simultaneous occurrence of ischaemic lesions of its medial and lateral parts would be extremely rare. We report MRI of a patient with an ipsilateral lateral medullary infarction (Wallenberg’s syndrome) followed by a medial medullary infarction with a 6 month interval. The initial stroke event on the central facial weakness of this patient was previously reported. A 40 year old man noted weakness of the right limbs and dysarthria. Since the age of 38, the patient had been on an antihypertensive drug regimen. He was conscious, and his blood pressure was 134/80 mm Hg. Neurologically, there were noted right supranuclear facial weakness, paralysis of the left hypoglossal nerve, paralytic dysarthria, and right hemiparesis with prominent weakness of the arm. Deep tendon reflexes were exaggerated in the right limbs and Babinski’s sign was positive on the right side. Light touch and vibratory sensations were slightly decreased in the right leg, but temperature and pain sensations were well preserved. There was no diabetes mellitus or hyperlipidaemia. No abnormalities were noted on ECG, transthoracic echocardiogram, and Holter monitoring. Brain MRI at 10 days after the onset showed a high signal intensity area (T2 weighted image) in the left ventromedial part of the upper medulla, which was diagnosed as an infarct (figure A). Cerebral angiography performed at 12 days after onset showed mild atherosclerotic changes of the verteobasilar artery. The right facial weakness and left tongue paralysis were gradually improved and the patient was discharged with persisting mild right hemiparesis after one month.

Six months later, however, he suddenly experienced a floating sensation and gait difficulty. At that time he was alert and had a blood pressure of 110/60 mm Hg. Neurological examination showed left Horner’s syndrome, skew deviation with slight downward position of the left eyeball, clockwise rotatory nystagmus, impairment of temperature and pain sensations of the left face, paralysis of the left soft palate, hoarseness, slight dysarthria, and ataxia of the left limbs, decreased temperature and pain sensations of the neck and all parts of the right side, and hypophorhosis of the left half of the body. Slight right hemiparesis was present. Brain MRI performed on the next day showed a high signal intensity area (T2 weighted image) in the left dorsolateral portion of the upper to middle medulla. A repeat MRI study at one month after this second attack clearly showed an infarct in a left dorsolateral portion of the medulla with a reduction of oedema, and showed a reduction of an infarct in the left ventromedial part from the first stroke (figure B).

The patient presented here developed a left dorsolateral medullary infarction 6 months after the onset of an infarction involving the ventromedial part of the upper medulla. Both were clearly seen on MRI. The dorsolateral and medial part of the medulla are supplied by separate branches from the posterior inferior cerebellar artery and the anterior spinal artery, both of which are indirectly supplied by the vertebral artery. Hence the occlusion of the vertebral artery could produce simultaneous hemimedullary syndrome. However, serial infarctions of the separate topographic areas of the medulla resulting in a hemimedullary syndrome have been suggested, and serial events documented by MRI—such as in the present patient—have not been reported. The present case provided unequivocal evidence that the hemimedullary syndrome can occur as serial events of the medial and lateral medullary infarctions.

With the advent of MRI, it has recently become possible to see microvascular lesions in the brain stem that would in the past have been detected only by postmortem examination. In patients where such multiple ischaemic lesions occur at separate sites and times so as to form a complete Wallenberg syndrome, serial observations with MRI will certainly prove the pathogenic background of this syndrome and be of great clinical interest.

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Oculo-auroculo-vertebral spectrum disorder (Goldenhar’s syndrome) coexisting with schizophreniform disorder

Oculo-auroculo-vertebral spectrum disorder is a complex and heterogeneous inborn condition affecting primarily the face, skull, and other craniofacial structures. It may additionally affect the skeletal system, kidneys, heart, gastrointestinal tract, and eye. Familial occurrence has been reported, but most cases seem to be sporadic. Structural CNS and neurological abnormalities occur often with great variability. Mental retardation seems to be a common feature, but other relevant psychiatric disorders have not been reported.

We present what seems to be the first published case of a patient with an organic schizophreniform disorder and oculo-auroculo-vertebral spectrum disorder.

A 27 year old man was referred for psychiatric treatment by his general practitioner. The patient reported intermittent speech problems since the age of 9 which he had shamefully convinced, were the effect of “electrosomn”. He would often smell strange and sickening odours originating in his mother’s kitchen that nobody else would notice and were causing his problems. Officially he was studying business administration, but had not passed any of the necessary exams in the past 3 years. Mentally he thought he was absolutely sane.

From birth he showed facial asymmetry, right thumb dysplasia, and dysplasia of the right auricle. Neither the patient nor his mother knew of a relevant family history of psychiatric disorders or dysplasias of any kind. There were no further psychiatric treatments except for one short admission for somatisation disorder 2 years previously. No prenatal or perinatal abnormalities were known. The patient’s school performance had been average to good. He had first successfully trained and worked as a chemical laboratory assistant, but had been discharged from the polytechnic engineering course for no obvious reason. On psychiatric examination the patient was suspicious and reserved. Thought contents were restricted to his complaints; formal thought showed a loosening of associations and often tended to be illogical. Attention, comprehension, and concentration were mildly impaired. There were olfactory hallucinations (kitchen smells), but no other
Disabling stroke arising five months after internal carotid artery dissection

Dissection of the internal carotid artery is an increasingly recognised cause of acute ischaemic stroke in young adults and children. It may arise spontaneously or secondary to blunt or penetrating trauma. It has been reported after seemingly trivial incidents, such as reversing a car, washing hair, or holding a mobile telephone by flexing the neck against the shoulder. The incidence of carotid dissection is about 2.5–3/100 000/year—similar to aneurysmal subarachnoid haemorrhage.1 The most common presenting features are those of embolic or retroorbital, or hemispheric pain, Horner’s syndrome, and local cranial nerve palsies, plus potentially devastating cerebral ischaemic events. Although no trial data exist to support the use of anticoagulation, some vascular surgeons believe that anticoagulation does not increase the risk of intracranial haemorrhage.2–4 Given the potentially fatal or disabling consequences of carotid dissection, the diagnosis and management of carotid dissection is urgent.5


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medications included, there was no recurrence of seizures. Osmoprotectants and thiamine were not used. The patient was discharged from the hospital after a total of 13 days with the diagnosis of focal cortical dysplasia with disordered cortical development and intractable partial seizures. The patient was followed up in the epilepsy clinic and continued on the same treatment regimen. The seizures remained controlled, and the patient was able to continue his work and other activities without significant disruption.

It is likely that this patient's intractable partial seizures were related to the focal cortical dysplasia with disordered cortical development. The use of osmoprotectants and thiamine may have played a role in reducing the severity of the seizures, but the main treatment was the antiepileptic drug therapy. The patient's adherence to the treatment regimen and regular follow-up care were critical in achieving seizure control.

Conclusion:
Focal cortical dysplasia with disordered cortical development is a rare condition that can cause intractable partial seizures. Early diagnosis and appropriate treatment can lead to seizure control and improvement in quality of life. Further research is needed to understand the underlying mechanisms and to develop more effective treatments.

References:

Severe weight loss after withdrawal of chronic pizotifen treatment

A 36 year old man was admitted to hospitafor investigation of weight loss of 10 kg over 2 months. He had had classic migraine for over 20 years, and had been taking 1 mg pizotifen in the treatment of underweight patients. The patient had no clinical evidence of the heritable connective tissue disorders sometimes associated with multiple or recurrent dissections (Marfan's syndrome, Ehlers Danlos type IV, etc.). He clearly had three ischaemic episodes over five months attributable to the right internal carotid artery and we thought that predicting a recurrent stroke was going to be a major concern. His headache attacks were associated with a craving for carbohydrates, and weight gain, particularly induced by its powerful antiserotonin activity, an effect which has been used clinically in the treatment of obesity and convalescent patients. Although weight loss could theoretically follow discontinuation of chronic pizotifen treatment, this does not seem to be a noted side effect, and there are no published reports of severe weight changes. However, the manufacturers (Sandoz Pharmaceuticals) received a similar case report to this in 1986 in which a patient who had taken 1.5 mg daily for 2 years had lost 2 stone over 2 months after withdrawal of the treatment. Hence, it seems that marked body weight reduction may follow withdrawal of long term pizotifen treatment, and knowledge of this adverse effect may prevent excessive and unnecessary investigation of patients currently presenting with anorexia and severe weight loss.

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Successful treatment of intractable epilepsy partialis continua with multiple subpial transections

Cortical dysplasia is increasingly being recognised as an important cause of partial seizures including epilepsy partialis continua. With the advent of high resolution MRI it is now often possible to identify areas of cortical dysplasia, increasing the possibility of neurological surgery when seizures are refractory to medical treatment. We present a patient with intractable epilepsy partialis continua due to cortical dysplasia that was refractory to all medical treatments, was not evident on MRI, and was dramatically improved by multiple subpial transections. A 19 year old boy presented with the product of an uneventful pregnancy with no perinatal problems and normal early milestones developed simple partial seizures at the age of 14 years. These comprised clonic movements of the left side of his face, left arm, and left leg. Brain MRI was reported as normal and an EEG showed a right frontoparietal focus. Carbamazepine was started at a dose of 800 mg daily but had to be discontinued because of severe nausea. After a second recurrence of the left side simple partial seizures responded to the addition of phenytoin (300 mg daily). Over the next four years there was reduced academic performance and seizures were infrequent. At the age of 19 he developed clonic movements of the right side of his face and right arm. These partial seizures increased in frequency and after a week he was admitted to the hospital. Epilepsia partialis continua comprising continuous motor seizures of the right side of his face and right arm. There was no alteration of consciousness but there were seizures sufficiently distressing to require sedation and ventilatory support. Several antiepileptic drugs were tried over a two month period without success, including phenytoin, phenobarbitone, sodium valproate, vigabatrin, and infusions of clonazepam, lorazepam, and diazepam. The patient was transferred to this hospital, ventilated, and sedated on a third occasion. On examination there was marked hepatosplenomegaly but no focal neurological deficit was evident. Investigations were aimed at identifying a progressive degenerative disorder in view of the cognitive decline, involvement of both hemispheres at different times, and the hepatosplenomegaly. A full metabolic screen was normal. Liver, bone marrow, skin, and muscle biopsies were non-contributory and a screen for mitochondrial DNA mutations was negative. The MRI was performed on a 0.5 T Vector (GE) instrument with T1 and T2 weighted axial and coronal sequences including volume acquisitions, and again no abnormality was detected. His EEG showed spike and sharp waves over the left central cortex, and during seizures runs of repetitive polyphasic discharges were recorded. Electroencephalography and a biopsy of the left premotor cortex were performed. The cortex appeared macroscopically normal but electrocorticography showed polyphasic discharges over the area that was biopsied. Histological examination showed areas of focal cortical dysplasia with disrupted lamination, large dysplastic neurons occurring in clusters and aggregates of astrocytes (figure). During this period of investigation his seizures could only be controlled initially on a thiopentone infusion that was sufficient to cause burst suppression on EEG. This resulted in impairment of liver function and coagulopathy and so was therefore replaced with a propofol infusion. High doses of up to 1000 mg/hour were required to control his seizures, but this was associated with prolonged bleeding time and gastrointestinal haemorrhage and had to be discontinued. Other antiepileptic drugs were tried without success. Vigabatrin at a dose of 4g daily, carbamazepine <3g daily, gabapentin <2.4 g daily, piracetam <24 g daily, and acetazolamide <1 g daily did not control seizures. Phenytoin induced myelosuppression and was withdrawn. Some improvement was obtained with a combination of ketamine, phenobarbitone, and midazolam but at doses that rendered the patient unconscious. In view of the intractability of the seizures and biopsy findings, multiple subpial transections were undertaken. Before the transections were performed high amplitude spikes were recorded over the left prefrontal and motor cortex on electrocorticography. Guided by the electrocorticography, multiple subpial transections were performed on the left precentral and postcentral gyri and after this the electrocorticography showed no epileptiform discharges. Postoperatively there was marked improvement in seizures, it was possible to discontinue intravenous antiepileptic drugs and the patient was extubated. The hepatosplenomegaly resolved. He subsequently developed a brief self limiting clonic movements of the right arm. There was no neurological deficit attributable to the surgical procedure but rehabilitation was hampered by a critical illness polyneuropathy. Nine months after surgery the patient was able to walk with a wheelchair bound but is beginning to mobilise, with no recurrence of his EPC. Cortical dysplasia is increasingly being recognised as an important and treatable cause of intractable seizures.
There have recently been major advances in structural brain imaging that allow detection of cortical abnormalities with high resolution MRI, although a considerable proportion of cases are not identifiable.

The histological diagnosis, complications, and failure of the patient to respond to pharmacological measures led us to perform multiple subpial transections with dramatic effect. The results of surgical treatment for cortical dysplasia depend on the site and grade of the lesion and type of procedure performed. The most established techniques are lesionectomy, resection of epileptogenic tissue as assessed by electrocorticography, or a combination of the two procedures. The use of multiple subpial transections has also been described and used in an attempt to avoid any deficit resulting from excision of primary sensorimotor cortex. This technique has been successfully used to end EPC due to Rasmussen's syndrome, but this is the first reported case we are aware of in which it has been used as a treatment for intractable epilepsy partialis continua due to cortical dysplasia.

The outcome in terms of seizure control was good and our patient was spared the hemiparesis that would have resulted from resection of the motor cortex. We conclude that multiple subpial transections should therefore be considered in patients with medically refractory EPC secondary to cortical dysplasia, even when structural imaging is unremarkable.

CORRESPONDENCE

Sjögren's syndrome in patients with chronic idiopathic axonal polyneuropathy

We appreciate the results of the study by van Dijk et al and add some results of our own regarding peripheral neuropathy and primary Sjögren's syndrome.

The proportion of patients who fulfil the diagnostic criteria for Sjögren's syndrome certainly depends on disease classification. Grant et al propose a distinct syndrome which includes patients with peripheral neuropathy and sicca complex. In their study minor salivary gland biopsy was positive in 73% of patients, which is in accordance with our studies. In our studies and those of Grant et al labial gland biopsy was not performed until sicca symptoms were established. This supports the desirability of searching for a
non-invasive test which should replace salivary gland biopsy.

In our study of 44 patients with chronic axonal neuropathy of small nerve fibres primary Sjögren’s syndrome was confirmed in 23%. The smaller proportion (4.6%) of patients with peripheral Sjögren’s syndrome as described in the study of van Dijk et al.1 It seems that van Dijk et al. as well as Grant et al.2,3 ignored a group of patients with small nerve fibre involvement lacking the psychophysiological assessment of thermal specific and thermal pain sensitivity as well as electrophysiological evaluation of the autonomic nervous system.

We agree with the authors that in patients with peripheral neuropathy sicca symptoms are often unrecognised. All of our patients were referred to the outpatients’ clinic because of positive sensory symptoms. Mainly they had burning pain. In most patients routine electrophysiological findings were within normal limits whereas thermal specific and thermal pain sensitivity assessment as well as electrophysiological determination of autonomic nervous system function disclosed distinct abnormalities.

We are aware that there is no specific test to evaluate Sjögren’s syndrome or peripheral neuropathy with sicca complex as described by Kennett and Harding.1 However, there is no reason to perform labial gland biopsy in each patient with chronic idiopathic axonal neuropathy as described in the study of van Dijk et al. Finally, we want to stress the need for a non-invasive method for diagnosing Sjögren’s syndrome.1

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Authors’ reply: We thank Denišič and Meh for their comments on our study regarding Sjögren’s syndrome in patients with chronic idiopathic axonal polyneuropathy (CIAP). As another example of the wide range of peripheral nerve disorders in Sjögren’s syndrome, they stress the importance of recognising patients with small nerve fibre disease. We agree that these patients were excluded from our study, as axonal nerve fibre dysfunction on electrophysiological examination was required to confirm the clinical diagnosis of polyneuropathy in our study, and the routine neurophysiological investigations often do not show abnormalities in patients with small nerve fibre disease. When small nerve fibre dysfunction of unknown cause is present, we also perform an investigation for Sjögren’s syndrome as we described. We do not recommend performing a labial salivary gland biopsy in every patient with CIAP. As in the figure, we suggest performing further tests for Sjögren’s syndrome in men with CIAP only when ocular or oral sicca symptoms are present and only a labial salivary gland biopsy when objective tests for tear gland and salivary gland dysfunction or serological abnormalities are also present. However, in women with CIAP we suggest a full investigation for Sjögren’s syndrome that includes a questionnaire for ocular and oral symptoms, a Schirmer’s test or Rose bengal staining score, and determination of serological abnormalities; when an abnormal result is found in any of these tests, a sublabial salivary gland biopsy should be performed to see if a diagnosis of Sjögren’s syndrome can be made.

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BOOK REVIEWS


With an ever increasing number of neurology texts concentrating on diagnosis and medical science, a book with an emphasis on therapeutics is a welcome addition. The key problem is that whereas older treatments may change little, up to date ones change in the time it takes such a book to come to fruition. Nevertheless this text does cover a broad range of therapeutic issues with access to key publications in a way that would be difficult to find in another single volume.

It is divided into sections, for example headache, degenerative diseases, and seizure disorders. Each starts with a brief overview and goes on to chapters dedicated to specific topics. Common diseases are given substantial coverage but the choice of more minor topics is curious. Whole chapters are devoted to botulism, diphtheritic neuropathy, and organophosphate poisoning and a few lines only to tuberculosis or AIDS and its complications. The emphasis varies throughout the book. A very valuable chapter on Parkinson’s disease devotes three pages to diagnosis and 40 pages to topical issues in therapeutics, whereas the chapter on myasthenia gravis devotes 14 pages to diagnosis and eight to treatment. In some chapters the discussion of treatment is little more than a list of drugs and their side effects. The more successful chapters give an analysis of relevant publications and try to give up to date treatment options for specific clinical situations. Others are more didactic, drawing on the authors’ personal experiences. The authors are Amer-

I cannot imagine ever getting any sensual enjoyment from handling a text on CD ROM but thumbing through some books will always be a pleasure. Among them are those that are beautifully produced on glossy paper with high quality illustrations. This is such a book and is one of a series of medical atlases from this publisher. It starts with 40 pages of well spaced text providing a succinct summary of some important aspects of epilepsy. Where possible there are tabulated data in support of the text—for example assessing risk of seizures after different kinds of head injury. But this book is not about text, it is about pictures. They include clinical photographs, EEG tracings, extensive neuroimaging, and microscopic and macroscopic pathological specimens. There are also a few graphs illustrating epidemiological points. Each figure has a clear and comprehensive figure legend. The pathology illustrations, both gross and microscopic are the high point of the atlas. The neuroimaging illustrates relevant pathology but surprisingly most of the scans are CT with only a handful of MRI. The EEGs cover a range of topics from common syndromes to curiosities such as the EEG background of Angelman’s syndrome. Most are clear but some tracings have come out rather faint. The electrode montages are somewhat disappointingly listed under the tracings rather than incorporated into the illustrations. Most descriptions do not have arrows to point to the area of interest but as the book is likely to be bought by those who have an established interest in epilepsy, rather than by novices, this is perhaps less critical.

For the discerning reader who wishes to know more than just the simple facts regarding the normal function of the pituitary gland and the pathophysiology which can affect it, this book offers a very readable starting point. Despite its relatively small size, the pituitary is an extremely important player in the endocrine team and a wide array of clinical features can arise from its dysfunction. Not surprisingly, traditional textbooks often approach this fascinating topic in purely clinical terms, either as a brief overview of the relevant facts, or as a comprehensive reference. Comprehensive references are usually bulky and seen as places to look when faced with a particular clinical problem.


Neurocysticercosis is the commonest chronic parasitic infestation of the central nervous system. Although malaria is perhaps the commonest acute parasitic illness, as far as chronic infestation is concerned neurocysticercosis affects many countries. Interest in neurocysticercosis and its clinical manifestation has been intense in the past 15 years, since the advent of CT in many countries where it is endemic. Epilepsy is the commonest neurological presentation and reports from all over the world show that in many countries it is the prime cause for epilepsy at various ages, especially in adults. Neurocysticercosis is now seen worldwide in people who emigrate, travel, or visit countries where it is endemic. Because of the ease of travel neurologists are perhaps more aware of the existence of the condition but in those who live in countries where it is endemic. The condition is endemic in Central and South America, India, and many parts of Africa. The scientific literature on the topic is divided between the Spanish and English languages. This book is divided into 14 chapters, only three of which are written in English. For a non-Spanish speaker the book offers little value but on the other hand, it is most valuable to people in the Spanish speaking world. The information is up to date with the advent of CT imaging, up to date immunology, and extensive photographs of pathological specimens.

This may sound a bit “politically incorrect” but homogeneity is important in any book and the fact that this book is written in mixed languages does not really serve this purpose. It is much easier for this book to be appreciated by a Spanish rather than an English reader, who will have difficulty in comprehending many of the chapters.

I do not think that in these very financially restrained times I can recommend a book which is mainly written in Spanish, to university departments or neurological units in the English speaking world.

Letters, Correspondence, Book reviews

R A SHAKIR

Progressing from the video cassettes to the CD-ROM is a logical step. The ILAE classification of epileptic seizures is now standard teaching throughout the world. The video illustrated classification of epileptic seizures was compiled by the Commission of the ILAE from collected EEG video telemetry tapes, mainly from Europe. The editors point out that some of the material is unclear from the source and hence the CD-ROM picture is not optimal. This is certainly true in many cases. It is understandable how difficult it is to produce uniform video recording conditions of epileptic seizures from many countries of the world and at different periods throughout the past 15 years or so. The CD-ROM atlas is accompanied by a manual which attempts to explain the classification of epileptic seizures as well as the epilepsies and then goes on to give details of every case shown on the CD-ROM. Loading the CD-ROM into various computers is rather cumbersome as one has to wait for the loading process to happen and there are no direct short cuts. After the preliminaries on the CD-ROM six sections of the CD are displayed and the user can click and access any one of them. The first is a guided tour of what is available and the second section is on seizure classification. The third section is the video library and case histories, for which it is necessary to refer to the manual. There is an information section and a very interesting self assessment section which is set at various levels of knowledge of epilepsy syndrome. The scoring is on the ability to recognize correctly various seizure types and the scoring system is graded according to the level of error. A special applications section is very interesting; the user can compile a sequence of seizures—for instance, for teaching purposes, and this can be run in whatever way the user requires. Producing this CD-ROM atlas is an excellent idea which will serve not only medical students and doctors but also all of those involved in the care of the patient with epilepsy. Nurses and EEG technologists, both in training and in practice will find the CD most valuable. There are, however, some criticisms and these centre on the quality of some of the videos, which are very difficult to see. The absence of an EEG montage on screen is a great disadvantage. There is a possibility of adding a running commentary on the screen so that the user does not have to refer to the manual which tends to be too descriptive and at times imprecise. The size of the picture on the CD-ROM is another issue which needs to be considered and whether a larger picture is technically feasible. To the amateur user of computers, which perhaps describes many in epilepsy (myself included), I find this CD very interesting and I am sure the Commission will fine tune the CD-ROM in years to come to make it rather more user friendly. Apart from the criticisms above, this is a most valuable addition to our knowledge and understanding of epileptic seizures. Many neurology departments throughout the world will find this a most interesting purchase.

R.A. Shakir


It is unfair and unreasonable to review volume 11 of the Handbook of Neuropsychology without considering it in the context of the preceding 10 volumes. This magnificent enterprise started in 1988 and has been steered through to its completion by Boller and Grafman: it is truly monumental, consisting of some 3–4000 pages of text and covering virtually every aspect of neuropsychology from adult to childhood disorders, classic syndromes to modern cognitive neuropsychology, imaging to computational networks, movement to consciousness, memory to language, and assessment to treatment. The editors have assembled a star studded cast which is transatlantic, but sadly deficient in contributors from Japan.

The bulk of Volume 11 concerns the complex issue of action and cognition edited by Jeannerod. It combines chapters on anatomy, computational modelling, and neuropsychology. It is up to date and is well referenced. The final two smaller chapters are devoted to emerging techniques (functional MRI and transcranial magnetic stimulation) and current views on consciousness. This volume lacks perhaps the coherence of some early volumes and seems very expensive, in that a fifth of the book is dedicated to a cumulative subject and author index.

It is a cliché to say that “everyone should buy it”, but I really think that in this instance no self respecting neuropsychological unit or university library should be without the complete handbook. Unfortunately, it is beyond the pocket of most individual people. It is unlikely to be superseded his century, at least, I imagine, by these editors, who deserve a medal for their contribution to neuropsychology.

John Hodges
Reversible proximal myopathy in epilepsy related Cushing's syndrome

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