LETTERS TO THE EDITOR

Reversible proximal myopathy in epilepsy related Cushing's syndrome

Epilepsy can induce hypercortisolism secondary to altered temporolimbic modulation of the hypothalamopituitary secretion of ACTH.[1 2] Hypercortisolism can produce a reversible myopathy. Epilepsy, however, has not been recognised to cause a symptomatic Cushing's syndrome. The development of proximal myopathy as part of an epilepsy based 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 fatiguability, 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 I 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 and creatine kinase concentrations were normal. All serum concentrations of antiepileptic drugs were in the therapeutic range.

Treatment with ketoconazole (200 mg three to four times a day) normalised the 4 00 pm serum cortisol and 24 hour urinary cortisol concentrations in all three patients. Normalisation of cortisol concentrations was associated with alleviation of the complaint of increased fatiguability, normalisation of strength and EMG, and a reduction in seizure frequency in all three patients (table). Discontinuation of ketoconazole for three months in all three patients was associated with a recurrence of the myopathy within two to three weeks with subsequent recovery again within three weeks after reinstitution of ketoconazole.

Proximal muscle weakness in all three patients could be attributed to hypercortisolism. This conclusion is supported by the documentation of hypercortisolism, the failure to find other possible causes, and the reversibility of the myopathy and accompanying EMG features by normalisation of serum cortisol concentrations using ketoconazole treatment.[3] Hypercortisolism, in turn, may have been related to abnormal temporolimbic modulation of hypothalamopituitary secretion of ACTH.[1 2] This possibility was suggested by the presence of active, focal temporal epileptiform discharges in all three patients, the absence of any pituitary or adrenal tumour or hyperplasia, the absence of abdominal striae characteristic of classic Cushing's syndrome, and the absence of emotional depression, clinically and by the Hamilton scale, which can also raise serum cortisol. The intermittent occurrence of this Cushing's myopathy and its relation with antecedent seizure exacerbation in patient 3 offers further support for a possible epileptic causality. The description of this previously unreported epilepsy based Cushing's myopathy is important because it represents a readily treatable cause of fatiguability and muscle weakness which may otherwise be underrecognised and attributed to postictal effects or medication toxicity.

Some adrenocorticosteroids, including cortisol, are glutamatergic, as well as bicuculline or picrotoxin-like antagonists of γ-aminobutyric acid A (GABA-A) receptor in the brain and exert potent anxiogenic, proconvulsant, and convulsant effects.[4 5] Hypercortisolism, therefore, may potentially exacerbate seizure disorders. The favourable seizure response to normalisation of serum cortisol concentrations in all three patients is consistent with this notion and the possibility that temporolimbic epilepsy may not only induce hypercortisolism but that the hypercortisolism, in turn, may exacerbate temporolimbic discharges leading to a positive feedback loop and a vicious circle.

These 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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1 Gallagher BB, Flanigin HF, King DW, *et al*. The effect of electrical stimulation of medial temporal lobe structures in epileptic patients upon ACTH, prolactin, and growth hormone. *Neurol* 1987;**37**:299–303.
2 Mandell AJ, Chapman LF, Rand RW, *et al*. Plasma corticosteroids: changes in concentration after stimulation of hippocampus and amygdala. *Science* 1963;**139**:1212.
3 Sonino N. The use of ketoconazole as an inhibitor of steroid production. *N Engl J Med* 1987;**317**:812–8.
4 Paul SM, Purdy RH. Neuroactive steroids. *FASEB J* 1992;**6**:2311–22.
5 Deutsch SI, Mastropaolo J, Hitri A. GABA-active steroids: endogenous modulators of GABA-gated chloride ion conductance. *Clin Neuropharmacol* 1992;**15**:352–64.

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.[1–4] The syndrome with both dorsolateral and

Summary of patients

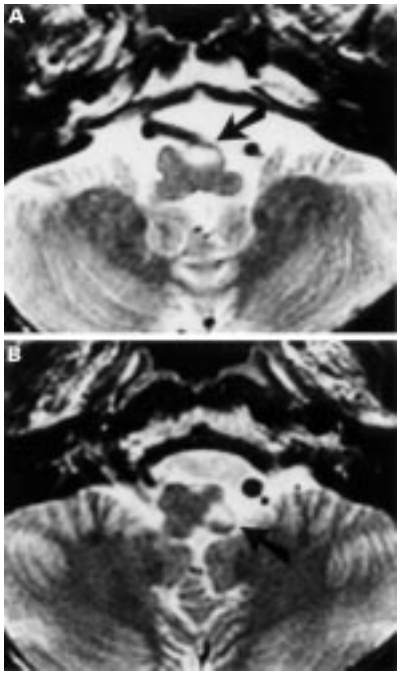
	Patient 1	*Patient 2*	*Patient 3*
Age (y)	32	43	31
Epilepsy	CPS/SGS	CPS/SGS	CPS
EEG	L and R temp foci	R temp focus	L temp focus
Medications	Carbamazepine	Phenytoin phenobarbitone	Carbamazepine phenytoin
Irregular menses	+	+	+
Hirsutism	+	+	+
Obesity	Mild	–	Moderate
Striae	–	–	–
Weakness	Mild persistent	Moderate persistent	Moderate intermittent
Cortisol:			
Serum (pm) (1.4–10.7 μg/dl)	19.8–31.0	11.0–15.0	25.1–29.9
Urinary (20–90 μg/24h)	70.3–106.0	57–102	123–183
ACTH (20–90 pg/ml)	57–124	72–120	76.5–142.5
Dex supp (<1 μg/dl)	+	+	+
EMG	Myopathic	Myopathic	Myopathic
Biopsy	Not done	Non-specific myopathic	Not done
Treatment	Ketoconazole 200 mg four times daily	Ketoconazole 200 mg three times daily	Ketoconazole 200 mg three times daily
Outcome:			
Follow up (y)	3	2	5
Myopathy	Absent	Absent	Absent
Sz freqn	92%	Seizure free	32%
Menses	Regular	Hysterectomy for fibroids	Regular

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; sz freqn=seizure frequency during follow up period compared with baseline.

medial infarctions is known as hemimedullary (Babinski-Nageotte) syndrome.[5] 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.[6 7] 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.[8]

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 vertebrobasilar 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 160/106 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 dysphagia, cerebellar ataxia of the left limbs, decreased temperature and pain sensations of the neck and all parts of the right side, and hypohidrosis 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).



Axial T2 weighted MRI (5000/96, TR/TE). High signal intensity areas (arrows) are noted in the left ventromedial part of the upper medulla (A) and the left dorsolateral part of the upper to middle medulla (B) 6 months after the first stroke.

The patient presented here developed a left dorsolateral medullary infarction 6 months after the onset of an infarction involving the left ventromedial part of the upper medulla. Both were clearly seen on MRI. The dorsolateral part 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.[2 6] However, serial infarctions of the separate topographic areas of the medulla resulting in a hemimedullary syndrome have been suggested,[3–5] 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 certain distinct syndrome, serial observations with MRI will certainly prove the pathogenic background of this syndrome and be of great clinical interest.

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1 Gan R, Noronha A. The medullary vascular syndromes revisited. *J Neurol* 1995;**242**:195–202.
2 Vuilleumier P, Bogousslavsky J, Regli F. Infarction of the lower brainstem. Clinical, aetiological, and MRI-topographic correlations. *Brain* 1995;**118**:1013–25.
3 Kim JS, Kim HG, Chung CS. Medial medullary syndrome. Report of 18 new patients and a review of the literature. *Stroke* 1995;**26**:1548–52.
4 Bassetti C, Bogousslavsky J, Mattle H, *et al*. Medial medullary stroke: report of seven patients and review of the literature. *Neurology* 1997;**48**:882–90.
5 Currier RD. Syndromes of the medulla oblongata. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*. Vol 2. Amsterdam: North-Holland, 1969:217–37.
6 Mossuto-Agatiello L, Kniahynicki C. The hemimedullary syndrome: case report and review of the literature. *J Neurol* 1990;**237**:208–12.
7 Nakane H, Okada Y, Sadoshima S, *et al*. Babinski-Nageotte syndrome on magnetic resonance imaging. *Stroke* 1991;**22**:272–5.
8 Terao S, Takatsu S, Izumi M, *et al*. Central facial weakness due to medial medullary infarction: the course of facial corticobulbar fibres. *J Neurol Neurosurg Psychiatry* 1997;**63**:391–3.

Oculo-auriculo-vertebral spectrum disorder (Goldenhar "syndrome") coexisting with schizophreniform disorder

Oculo-auriculo-vertebral spectrum disorder is a complex and heterogeneous inborn condition affecting primarily aural, oral, and manibular development with an estimated incidence of between 1/3500 and 1/26 000 births. It is thought to be connected to the development of the first and second branchial arch. 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.[1] Structural CNS and neurological abnormalities occur often, but with great variability.[2] Mental retardation seems to be a common feature,[1] 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-auriculo-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 was unshakably convinced, were the effect of "electrosmog". 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 he had discontinued a subsequent 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

sensory impairments. Delusional ideas—but no autochthonous delusions—were present (control by electrosmog), they were not systematised and had a low affective impact. There were no delusions of reference. Mood was blunt and flat. There were no psychomotor abnormalities, but marked apathy was noted. Neuropsychological testing showed normal intelligence, normal visual memory, reduced speed of visual processing, and, despite the discrete attention deficit, normal accuracy, concentration, and speed. Physical examination confirmed dysplasias of the right auricle and the right thumb as well as scoliosis of the cervical and thoracic spine. Cranial nerves were unaffected, but right sided conductive hearing loss (audiometrically confirmed) and marked facial asymmetry with a hypoplastic right half were prominent. Ear, nose, and throat examination showed submucous cleft palate, malocclusion and hypoplasia of the maxilla, mandibular asymmetry, and cluttering with mild stuttering.

Routine blood and urine tests were normal. Electrocardiography, echocardiography, and chest radiography showed no cardiac or pulmonary abnormalities. Radiography showed right sided rib deformation, aplasia of the left 12th rib, a hypoplastic atlas, a plump odontoid process, basilar impression, complete (C5/7) and incomplete (C2/3) fusion of vertebrae, rotational scoliosis, and segmental synostoses of the cervical and thoracic spine. Asymmetry of the mandible and hypoplasia of the maxilla was cephalometrically confirmed on skull radiography. Cranial MRI showed a hypoplastic vermis cerebelli inferior but no other abnormalities. An EEG showed no ictal activity. Cortical magnetic stimulation was normal, whereas the blink reflex showed markedly lowered responses on the right side (trigeminal nerve; R1) and the masseter reflex produced no right sided electrophysiological response, but normal responses on the left. Acoustic evoked potentials showed a peripheral conductive delay on the right side. The karyotype was 46, XY. Fluorescence in situ hybridisation techniques showed no microdeletions on chromosomes 22q13.3 or 10p13/14.

We treated the patient with the atypical neuroleptic drug olanzapine (15 mg/day) and supportive psychotherapy. This resulted in a mild but significant amelioration of the disturbances of thought, concentration, attention, and comprehension, the rather low intensity olfactory hallucinations and delusional ideas diminished markedly.

This patient with normal intelligence exhibited multiple abnormalities: oral (submucous cleft palate, malocclusion), phoniatric (cluttering with stuttering), auricular (right sided conductive hearing loss and dysplasia of the auricle), facial (asymmetry), skeletal (right sided aplasia of the 12th rib, fused cervical vertebrae, and other dysplasias of the cervical and thoracic spine, abnormalities of the cranial base, right thumb dysplasia). Oculo-auriculo-vertebral spectrum disorder lacks clear minimal diagnostic criteria, but according to Gorlin *et al*[1] our patient would qualify for this diagnosis.

There is no final answer to the question whether this schizophreniform disorder is idiopathic or symptomatic, as an unequivocal distinction between idiopathic and symptomatic schizophrenia cannot be made by phenomenology.[3] But despite the age of the patient, there are arguments for a symptomatic aetiology: The lack of autochthonous (or primary) delusions, of delusions of reference, and of Schneiderian first rank symptoms, the exclusively olfactory hallucinations, the absence of a positive family history of psychiatric disorders, the neither schizoid nor schizotypal premorbid personality, and the deficient pattern of thought disorders and cognitive abilities and at the same time the organic abnormalities affecting other systems of the head (speech, cleft palate, cranial base, hearing loss, facial asymmetry, cerebellar structures) support the idea of an organic background to the disorder.[3] We think that inborn syndromes of the head and neck—particularly oculo-auriculo-vertebral spectrum disorder—have received too little attention from neuropsychiatrists. Given their embryological position, these conditions might help develop some hypotheses about the aetiology of psychiatric disorders.

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1 Gorlin RJ, Cohen M, Levin LS. *Syndromes of the head and neck.* 3rd ed. New York: Oxford University Press, 1990.
2 Schrander-Stumpel CTRM, Die-Smulders CEM, Hennekam RCM, *et al.* Oculoauriculovertebral spectrum and cerebral anomalies. *J Med Genet* 1992;**29**:326–31.
3 Lewis SW. The secondary schizophrenias. In: Hirsch SR, Weinberger DL, eds. *Schizophrenia.* Oxford: Blackwell Science, 1995:324–40.

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 ipsilateral temporal, retroorbital, or hemicranial 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 anticoagulants, most cerebrovascular specialists advocate initial intravenous heparin then three to six months of warfarin treatment if there have been ischaemic episodes. There is often an interval between onset of symptoms and cerebral infarction enabling the diagnosis to be made and treatment to be instituted.[2] Given the potentially fatal or disabling consequences of carotid dissection this window of opportunity is not to be missed. About 80% of ischaemic strokes arise within the first seven days although they can occur four to five weeks after the onset of symptoms.[2 3] We report a patient who developed a disabling stroke five months after ipsilateral carotid dissection. This has implications for instituting treatment in a patient seen weeks or months after the incident, and for the duration of subsequent anticoagulation.

A 24 year old man was involved in a road traffic accident. Emergency fire services were required to free him from the vehicle and he remains amnesic for the event. He sustained soft tissue injuries to the face, chest, and arms, but no fractures. No surgery was required. No neurological sequelae were noted although he mentioned mild visual blurring in retrospect. A non-contrast CT was performed the day after the accident which showed a small right peripheral parietooccipital infarct (but this had been reported as a contusion). The patient was discharged and made a full recovery from his injuries.

About four months later he suddenly developed a left facial weakness. There was associated right retro-orbital and temporal headache. Over three to four days the headache and facial weakness cleared. Medical attention was not sought.

Just over five months (158 days) after the traffic accident he was admitted to our unit. That morning he had suddenly developed a dense left hemiparesis affecting the arm and face more than the leg. The right sided headache had returned. A repeat CT showed maturation of the previous occipitoparietal infarct and a new infarct in the corona radiata on the right, extending into the internal capsule. A colour flow Doppler/duplex scan of the cervical carotid arteries showed normal appearances on the left side, a normal right external carotid artery, but reduced diastolic flow in the right common carotid artery. The origin of the right internal carotid artery was patent but the Doppler waveform was severely damped with the typical "bidirectional" signal associated with distal occlusive disease and commonly seen in dissections. The diagnosis of dissection of the right internal carotid artery was confirmed by T1 weighted axial MRI of the neck. These showed characteristic high signal in the vessel wall due to haemorrhage with associated reduction in diameter of the true lumen. The patient was treated immediately with intravenous heparin. Seven days later warfarin was started for six months. His hemiparesis gradually improved.

The right internal carotid artery probably dissected at the time of the road traffic accident and the infarct seen on the initial CT had the characteristics of a peripheral embolic lesion in the territory of posterior superficial middle cerebral artery branches. A second (presumably) embolic insult causing transient facial weakness arose four months later. Another five weeks passed before the disabling left hemiparesis. During the five months since the traffic accident neither the patient nor his family could recall any occasions when he had complained of a new neck or head pain relating to further neck trauma. It seems likely that he had had only one insult to the right internal carotid artery but that over five months the vessel had failed to heal spontaneously and the damaged intima continued to act as a focus for thrombus formation.

Recurrent dissection of the same extracranial vessel is extremely unusual, and occurred in only one of 81 patients followed up for a

median of almost three years by Bassetti *et al*.[4] Recurrent dissection involving other extracranial vessels is slightly more common, with an incidence of about 1% a year.[5] Our 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 think that persisting damage, rather than true recurrence, was the likeliest cause of his stroke.

It is not uncommon to be asked to see young patients with ischaemic stroke several weeks after the event. Even when arterial dissection is suspected or proved, if the patient has had no further episodes for a month or more it is tempting to prescribe antiplatelet agents rather than submitting the patient for formal anticoagulation. This case shows that disabling stroke can occur as long as five months after the initial dissection; thus anticoagulation should probably be considered even when there is considerable delay in referral. Although empirical, a minimum period of six months on warfarin would seem appropriate. If repeat duplex ultrasound remains abnormal at that time, extension to 12 months may be necessary.

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1 Giroud M, Fayolle H, André N, *et al*. Incidence of internal carotid dissection in the community of Dijon. *J Neurol Neurosurg Psychiatry* 1995;**58**:1443.
2 Biousse V, D'Anglejan-Chatillon J, Touboul P-J, *et al*. Time course of symptoms in extracranial carotid artery dissections. *Stroke* 1995;**26**:235–9.
3 Hilton-Jones D, Warlow CP. Non-penetrating arterial trauma and cerebral infarction in the young. *Lancet* 1985;i:1435–8.
4 Bassetti C, Carruzzo A, Sturzenegger M, *et al*. Recurrence of cervical artery dissection: a prospective study of 81 patients. *Stroke* 1996;**27**:1804–7.
5 Schievink WI, Mokri B, O'Fallon WM. Recurrent spontaneous cervical artery dissection. *N Engl J Med* 1994;**330**:393–7.

Severe weight loss after withdrawal of chronic pizotifen treatment

A 36 year old woman was admitted to hospital for investigation of weight loss of 10 kg over 2 months. She had had classic migraine for over 20 years, and had been taking 1 mg pizotifen (Sanomigran) daily continuously as migraine prophylaxis with good effect. Eight weeks before admission, this drug had been discontinued. A week later, she developed frontal headaches with nausea. She became anorexic and began to lose weight quickly. General and full neurological examination showed no abnormality. Investigations including full blood count, erythrocyte sedimentation rate, urea and electrolytes, liver function tests, random cortisol, blood glucose, and thyroid function tests were all normal. A pregnancy test was negative, and gastroscopy, including duodenal biopsy showed no abnormality. Brain CT was normal. Pizotifen was restarted and she immediately felt better. Her appetite improved, headaches stopped, at 1 month she had regained 5 kg, and by 2 months was back to her normal weight, remaining asymptomatic.

Pizotifen is widely used for migraine prophylaxis, where it modifies humoral mechanisms inducing headache by effects on serotonin and histamine.[1] Treatment is often associated with increased appetite, a craving for carbohydrates, and weight gain, probably induced by its powerful antiserotonin activity, an effect which has been used clinically in the treatment of anorexic and convalescent patients.[2] 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 extensive and unnecessary investigation in those subsequently presenting with anorexia and severe weight loss.

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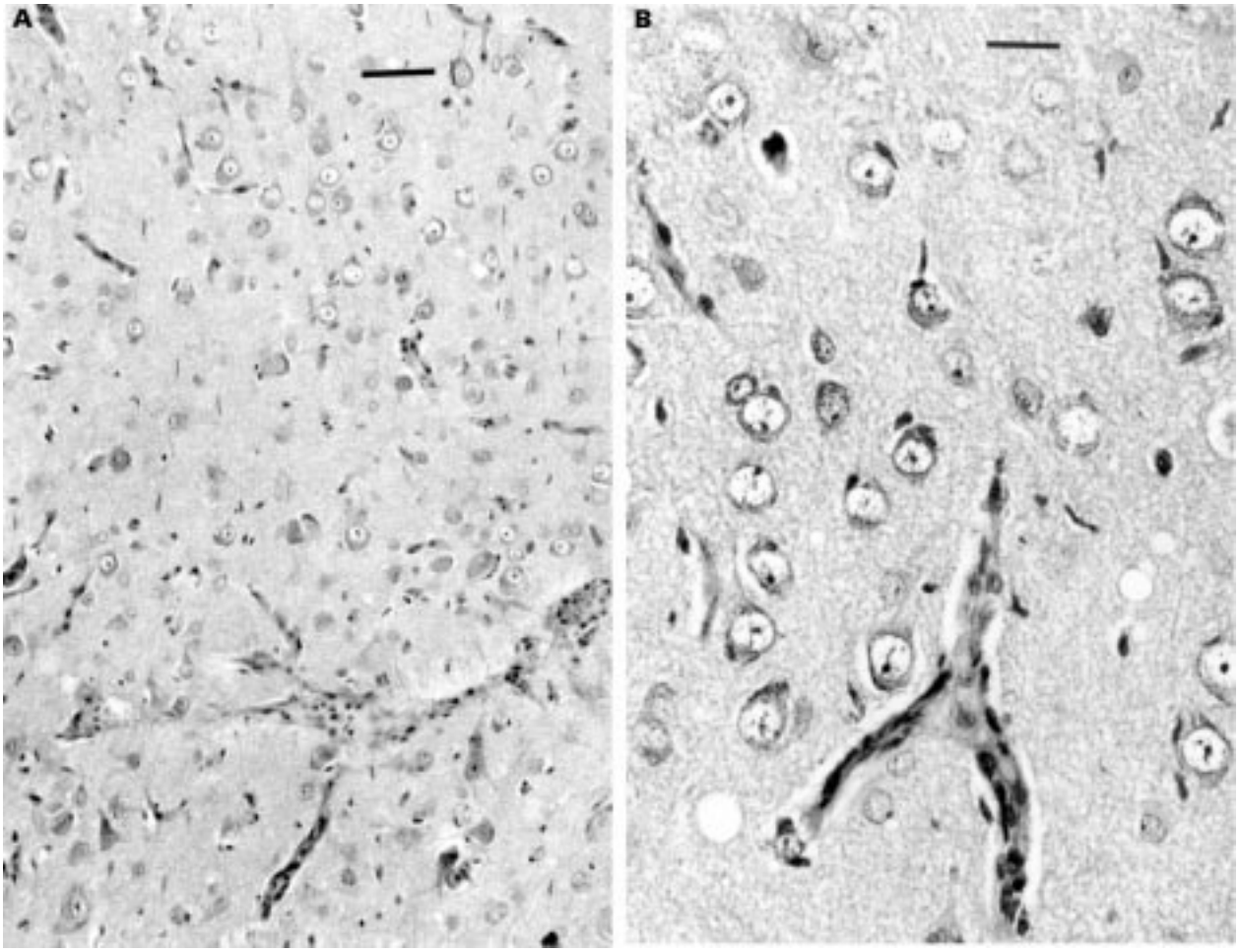
1 In: Reynolds JEF, ed. *Martindale. The extra pharmacopoeia.* 31st ed. London:Royal Society of Pharmacology, 1996:484.
2 Dolecek R. Experimental and clinical results with pizotifen in the treatment of underweight patients. *Pharmatherapeutica* 1980;**2**:363–71.

Successful treatment of intractable epilepsia partialis continua with multiple subpial transections

Cortical dysplasia is increasingly being recognised as an important cause of partial seizures including epilepsia partialis continua. With the advent of high resolution MRI it is now often possible to identify areas of cortical dysplasia, increasing the possibility of neurosurgical intervention when seizures are refractory to medical treatment. We present a patient with intractable epilepsia 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 man who was 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 and he remained seizure free for a year. A subsequent recurrence of the left sided simple partial seizures responded to the addition of phenytoin (300 mg daily). Over the next four years there was reduced academic attainment but 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 his local hospital with epilepsia partialis continua comprising continuous motor seizures of the right side of his face and right arm. There was no alteration of consciousness ictally but his seizures were sufficiently distressing to require sedation and ventilation. 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 thiopentone infusion. 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 noncontributory and a screen for mitochondrial DNA mutations was negative. The MRI was performed on a 0.5 T Vectra (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. Electrocorticography and a biopsy of the left premotor cortex was 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 disordered cortical 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 at doses 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 prolongation of 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 degree of control was obtained with a combination of ketamine, phenobarbitone, and midazolam but at doses that rendered the patient unconscious. In view of the intractable nature of the seizures and biopsy findings, multiple subpial transections were undertaken. Before the transections were performed high amplitude spikes were recorded over the left premotor 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 had only a few 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 discharge he remains 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



Photomicromicrographs of the excised specimens. (A) Showing dysplastic cortex with disordered cortical lamination, malorientated nerve cells, and focal clustering (haematoxylin and eosin, originally ×160, bar represents 25 μm). (B) Showing large dysplastic nerve cells (haematoxylin and eosin, originally ×400, bar represents 62.5 μm).

of partial seizures.[1 2] There have recently been major advances in structural brain imaging that allow detection of cortical abnormalities with high resolution MRI,[2–4] although a considerable proportion of cases are not identifiable.[3 4]

The histological diagnosis, complications, and failure of the patient to respond to pharmacological measures led us to perform multiple subplial 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.[4] 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 subplial transections has also been described[5] and used in an attempt to avoid any deficit resulting from excision of primary sensorimotor cortex.[5 6] This technique has been successfully used to end EPC due to Rasmussen's syndrome,[6] but this is the first reported case we are aware of in which it has been used as a treatment for intractable epilepsia 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.

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1 Desbiens R, Berkovic SF, Dubeau F, *et al*. Life-threatening focal status epilepticus due to occult cortical dysplasia. *Arch Neurol* 1993;**50**:695–700.
2 Raymond RR, Fish DR, Sisodiya SM, *et al*. Abnormalities of gyration, heteropias, tuberous sclerosis, focal cortical dysplasia, microdysgenesis, dysembryoplastic neuroepithelial tumour and dysgenesis of the archicortex in epilepsy: clinical, EEG and neuroimaging features in 100 adult patients. *Brain* 1995;**118**: 629–60.
3 Kuzniecky RI. MRI in developmental disorders of the cerebral cortex. *Epilepsia* 1994;**35**(suppl 6):S44–56.
4 Kuzniecky R, Murro A, King D, *et al*. MRI in childhood intractable partial epilepsies: pathological correlations. *Neurology* 1993;**43**: 681–7.
5 Morrell F, Whisler WW, Bleck TP. Multiple subpial transection: a new approach to the surgical treatment of focal epilepsy. *J Neurosurg* 1989;**70**:231–9.
6 Sawhney IM, Robertson IJA, Polkey CE, *et al*. Multiple subpial transection: a review of 21 cases. *J Neurol Neurosurg Psychiatry* 1995;**58**: 344–9.

CORRESPONDENCE

Sjögren's syndrome in patients with chronic idiopathic axonal polyneuropathy

We appreciate the results of the study by van Dijk *et al*[1] 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.[2] In their study minor salivary gland biopsy was positive in 73% of patients, which is in accordance with our studies.[3 4] In our studies and those of Grant *et al*[2] 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%) found by van Dijk *et al*[1] can be explained by the strict criteria used by inclusion of patients in our study. In other papers we stress the use of psychophysical methods besides routine electrophysiological tests in primary Sjögren's syndrome.[3 4] It seems that Dijk *et al*[1] as well as Grant *et al*[2] ignored a group of patients with small nerve fibre involvement lacking the psychophysical assessment of thermal specific and thermal pain sensitivity as well as electrophysical 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 outpatient's 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 defined by Kennett and Harding.[5] 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*.[1] Finally, we want to stress the need for a non-invasive method for diagnosing Sjögren's syndrome.[3]

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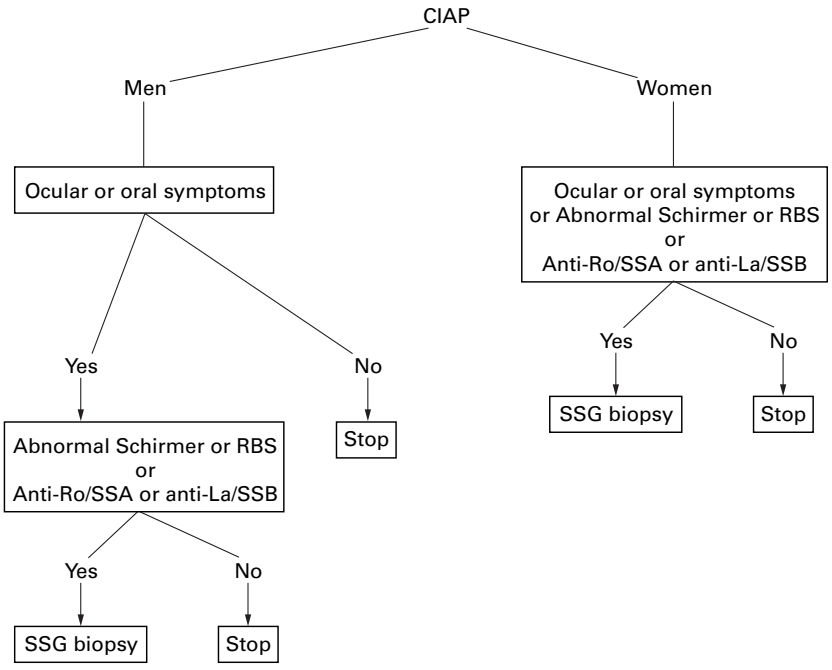
1 van Dijk GW, Notermans NC, Kater L, *et al.* Sjögren's syndrome in patients with chronic idiopathic axonal polyneuropathy. *J Neurol Neurosurg Psychiatry* 1997;**63**:376–8.
2 Grant IA, Hunder GG, Homburger HA, *et al.* Peripheral neuropathy associated with sicca complex. *Neurology* 1997;**48**:855–62.
3 Denišlič M, Meh D. Neurophysiological assessment of peripheral neuropathy in primary Sjögren's syndrome. *Clin Invest* 1994;**72**:822–9.
4 Denišlič M, Meh D. Early asymmetric neuropathy in primary Sjögren's syndrome. *J Neurol* 1997;**244**:383–7.
5 Kennett RP, Harding AE. Peripheral neuropathy associated with the sicca syndrome. *J Neurol Neurosurg Psychiatry* 1986;**49**:90–2.

Authors' reply:

We thank Denišlič and Meh for their comments on our study regarding Sjögren's syndrome in patients with chronic idiopathic axonal polyneuropathy (CIAP).[1] 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.



Diagnostic approach to Sjögren's syndrome in patients with chronic idiopathic axonal polyneuropathy (CIAP). SSG = sublabial salivary gland biopsy.

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1 Van Dijk GW, Notermans NC, Kater L, *et al.* Sjogren's syndrome in patients with chronic idiopathic axonal polyneuropathy. *J Neurol Neurosurg Psychiatry* 1997;**63**:376–8.

BOOK REVIEWS

Neurology Practice Guidelines. Edited by RICHARD LECHTENBERG, HENRY S SCHUTTA. (Pp 544; US$150). New York: Marcel Dekker, 1998. ISBN 0-8247-0104-6.

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 Ameri-

can and some familiar treatments are omitted—for example pizotifen for migraine prophylaxis. Others may sound strange to British ears, for example intravenous dihydroergotamine features prominently in treatment of chronic daily headache. Key references are cited only at the end of each chapter, which gives a free flowing text but makes it difficult to identify sources of specific information. There are many tables but no illustrations and no colour to break 500 pages of print, and although the text is readable, this is a missed opportunity.

The book is expensive at $150, but it does stand apart from most neurological texts, going some way to being a manual of current therapeutics. I should have liked to see a more consistent emphasis on treatment in all the chapters.

MARK MANFORD

An Atlas of Epilepsy. Edited by D F SMITH, R E APPLETON, J M MACKENZIE, D W CHADWICK. (Pp 158; £45.00). Carnforth: Parthenon Publishing, 1998. ISBN 1-85070-488-0.

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 injuries. But this book is not about text, it is about pictures. They include clinical photographs, EEG tracings, extensive neuroimaging, and microscopic and macroscopic pathology 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 points satisfactorily 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 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 illustrations 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.

The main use of such a book is as a source of illustrations for teaching material and as such it will prove valuable to epileptologists and neurological libraries. It seems well priced for this sort of text.

MARK MANFORD

Headache. Edited by PETER J GOADSBY, STEPHEN D SILBERSTEIN. (Pp 411; £60.00). Oxford: Butterworth Heinemann, 1997. ISBN 0-7506-9871-3.

This book is divided into three sections: migraine; other primary headaches; and secondary headaches. The pathophysiology of migraine is covered with clarity, although I doubt if I will ever quite come to grips with 14 different serotonin receptors. Symptomatic and prophylactic treatments of migraine are considered in detail with up to date coverage of available treatments and mention of drugs in development. Many women comment on the effect of menstruation on their migraine and one chapter tries to put this into pathophysiological perspective and suggests modes of treatment. There is also a useful description of some of the rarer idiopathic headaches.

The authors state that "tension type headache has in the past been an ill defined syndrome". Unfortunately, little in the ensuing pages suggests a major change in its status and the classification of chronic daily headache remains problematic. Some recognisable syndromes are described such as analgesic associated headache. In the chapter on tumours, the authors discuss the evidence surrounding the vexing question of when to scan and when not to scan. Other secondary headaches considered involve the temporomandibular joint, the neck, and painful ophthalmoplegia, topics rarely covered elsewhere. This section is generally well illustrated with scans and the basic science section has graphs and diagrams covering salient points.

The text is well referenced and the authors bring a strong critical appraisal to each topic. The result is a book that seems to be authoritative in breadth and depth, is securely grounded in clinical and scientific research, and has a coherent structure. It should prove to be invaluable to the jobbing neurologist, for whom headaches are bread and low fat spread.

MARK MANFORD

Diseases of the Pituitary. Edited by MARGARET E WIERMAN. (Pp 405; $125.00). New Jersey: Humana Press, 1997. ISBN 0-896-03364-3.

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 field in one of two ways, either as a brief overview of the salient facts, or as a comprehensive reference. Comprehensive references are usually bulky and seen as places to look when faced with a particular clinical problem.

Diseases of the Pituitary is different however, in that it attempts to combine comprehensiveness within a readable size and style. The book begins with an up to date explanation of the ontogeny of pituitary cell types and explains how recent advances in molecular biology have considerably enhanced our understanding in this area, as well as elucidating the mechanisms underlying various human syndromes arising from aberrant development. After covering the broader topic of hypopituitarism, subsequent chapters review the normal physiology and pathology of each of the anterior pituitary hormones. Although the chapters have been written by one or more experts in each field, the editor has managed to retain a unified style throughout. Of particular note is the structured approach to the diagnosis and treatment of pituitary deficiency or excess in which the authors discuss current practices and recent developments in such a way as to guide the clinician, but without trying to impose set rules or regimes. Furthermore, the text is clearly and extensively referenced and encourages the more inquisitive among us to seek out the original literature, especially in those areas where a consensus opinion is lacking. The book concludes with useful chapters covering radiological, surgical, and immunohistochemical aspects of pituitary disease.

The one feature of the book that would benefit from change is its illustration. In several places I think that an additional figure would aid description, or that a colour plate would have been more appropriate. This is especially true in the section on immunohistochemistry. However, this does not detract greatly from a book that should be recommended to anybody with more than a passing interest in the pituitary gland.

MARK GURNELL

Neurocysticercosis. Edited by CAMILO ARRIAGADA R, JORGE NOGALES-GAETE, WERNER APT B. (Pp 333; US$120.) Chile: Arrynog Ediciones, 1997. ISBN 956-272-689-4.

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 people worldwide. 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 clinical 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 not endemic, the experience is limited. Having a book on the subject is of immense interest to many doctors, not only in the neurological world but also in the disciplines of infectious diseases and tropical medicine.

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 modern 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 two 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 most 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.

R A SHAKIR

Atlas of Epileptic Seizures - Classical Examples for CD-ROM: International League Against Epilepsy: the Medicine Group (£20).

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 recognise 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

Handbook of Neuropsychology. Volume 11. Edited by MARC JEANNEROD (series editors P BOLLER, J GRAFMAN). (Pp 496; NLG 390). Amsterdam: Elsevier Science, 1997. ISBN (Series) 0-444-90491-1. ISBN (Vol H) 0-444-82467-7.

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 covers 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 dedicated 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