LETTERS TO THE EDITOR

Accessory middle cerebral artery and moyamoya disease

A rare association of moyamoya disease with the accessory middle cerebral artery was seen in two patients. The terminal portions of bilateral internal carotid arteries and their vicinities were markedly stenotic and so-called moyamoya vessels developed at the base of the brain. The left accessory middle cerebral artery originating near the anterior communicating artery supplied the left anterior frontal lobe in both patients. Although the accessory middle cerebral artery coursed in the vicinity of the markedly stenotic terminal portion of the left internal carotid artery, the artery was not stenotic. This finding implies that the steno-occlusive changes in the cerebral vasculature in moyamoya disease have topological predilection to the distal internal carotid arteries.

Moyamoya disease is characterised by angiographic features of steno-occlusive changes of the terminal portions of bilateral intracranial internal carotid arteries as well as dilated perforating arteries at the base of the brain known as “moyamoya” vessels. The clinical manifestation of moyamoya disease is typically brain ischaemia in the paediatric population and brain haemorrhage in adults. The accessory middle cerebral artery is a variation of middle cerebral artery branching and its incidence has been reported to be 0.3–4.0%. The accessory middle cerebral artery originates from either the proximal or distal horizontal portion of the anterior cerebral artery couring parallel to the horizontal portion of the middle cerebral artery and reaches the anterior frontal lobe. Patient 1, a 32 year old woman, had presented with a transient ischaemic attack of right hemiparesis 2 years before the current episode. Results of initial MRI imaging performed at another hospital were interpreted as normal, but MR angiography showed steno-occlusive changes of the terminal portions of bilateral intracranial internal carotid arteries. Stenotic changes were more severe on the right than on the left side. Although the proximal portion of the left middle cerebral artery was markedly stenotic on MR angiography, the left accessory middle cerebral artery was clearly shown to be without stenosis (fig 1 A). The patient was treated conservatively.

No recurrent ischaemic attack had occurred over a period of 2 years until the patient began to experience transient ischaemic attacks of right hemiparesis several times a month. She was referred to us for further evaluation. She was neurologically normal and history was not contributory except for mild hypertension for 2 years. Digital subtraction angiography showed progressive steno-occlusive changes of the terminal portions of the internal carotid arteries as well as development of moyamoya vessels, which were consistent with the diagnosis of moyamoya disease (fig 1 B). Most moyamoya vessels on the left side originated from the accessory middle cerebral artery. This patient subsequently underwent bypass surgery bilaterally. Frequency of transient ischaemic attacks reduced markedly during the follow up period of 3 months postoperatively.

Patient 2 was a 30 year old man admitted for re-evaluation of moyamoya disease. This patient experienced occasional headache and vomiting at the age of 5 years. At the age of 11 years, transient ischaemic attacks of right hemiparesis developed to a rate of one a week. The diagnosis of moyamoya disease was established by cerebral angiography. He underwent bilateral bypass surgery subsequently at the age of 13 years in another hospital. He had experienced no ischaemic episodes for 17 years thereafter and he thought that moyamoya disease was cured. He came to our hospital when he had minor head trauma at the age of 30 years and was advised to re-evaluate the disease.

At admission, the patient was neurologically normal. Brain MRI showed no parenchymal abnormality. Single photon emission computed tomography was normal. Right carotid angiography showed severe stenotic
change of the proximal right middle cerebral artery, but the right anterior cerebral artery was normal. The left accessory middle cerebral artery originated near the anterior communicating artery (fig 2 A). Left carotid angiography showed severe stenosis at the terminal portion of the internal carotid artery with moderate development of moyamoya vessels (fig 2 B). The left accessory middle cerebral artery was not stenotic despite the vicinity of the markedly stenotic distal internal carotid artery and middle cerebral artery. Moyamoya vessels were not supplied by the left accessory middle cerebral artery. The patient was conservatively followed up for 6 months without any ischaemic episodes.

An association of the accessory middle cerebral artery and cerebral aneurysms has been well documented.1,4 Moyamoya disease is highly associated with primitive carotid-basilar anastomosis, such as with the primitive trigeminal arteries and their variants.1 To our knowledge, however, an association of the accessory middle cerebral artery with moyamoya disease has not been reported. The accessory middle cerebral artery was first regarded as a hypertrophied recurrent artery.1 However, it is now thought to be a cortical branch of the middle cerebral artery supplying the anterior frontal lobe, which is annexed to the embryological early artery, the anterior cerebral artery.1 The accessory middle cerebral artery can serve as a collateral blood supply when the internal carotid artery or middle cerebral artery, or both are stenotic or occluded, as was the case in our patient.

Our patients are interesting in that (a) the accessory middle cerebral artery was associated with moyamoya disease and (b) the accessory middle cerebral artery was not stenotic even though the distal internal carotid artery and the proximal middle cerebral artery showed steno-occlusive changes. Stenotic changes were not seen in the accessory middle cerebral artery although it coursed in the vicinity of the stenotic horizontal portion of the middle cerebral artery. This implies that susceptibility to arterial stenotic change is limited to the distal portion of the internal carotid artery and the proximal middle cerebral artery but not to the accessory middle cerebral artery even though all of these vessels are in close proximity. The cause of moyamoya disease is still unknown, but we think that there is a topographic difference in the predilection to stenotic changes in the cerebral vasculature in the disease.

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Maternal age is not a risk factor for Parkinson’s disease

The aging process is associated with an accumulation of oxidative damage to mitochondrial DNA (mtDNA).1,4 Mutations and deletions of mtDNA accumulate with aging in various tissues including germ lines.1 The resulting mitochondrial defects, if transmitted to offspring through the maternal line, could potentially play a part in the pathogenesis of various neurodegenerative illnesses including Parkinson’s disease.2 In addition, advanced maternal age at the time of conception results in an increased risk of genetic birth defects. This is in part due to chromosomal mutations and malfunctions within the ova which are in turn due to the increased age of the ovary and the ova. We examined the age of mothers of Parkinson’s disease patients with Parkinson’s disease and control patients to evaluate whether maternal age may be a risk factor in the development of Parkinson’s disease.

Subjects were recruited from the Movement Disorders Clinic at the University of Virginia. We interviewed 612 consecutive patients with Parkinson’s disease and 376 consecutive spousal controls regarding their mother’s age at the time of the subject’s birth (maternal age). The diagnosis of Parkinson’s disease was based on internationally accepted criteria. At least two of the following three criteria had to be present: rest tremor, cogwheel rigidity, and bradykinesia. Asymmetry of these features at the time of diagnosis and at onset had to be present. Exclusion criteria included presence of atypical features, presence of a pre-existing possible cause, definite absence of response to levodopa, and a non-progressive course over at least 3 years. Moreover, 69% of the patients with Parkinson’s disease had been examined on multiple occasions thus increasing both our confidence in and accuracy of the diagnosis of Parkinson’s disease.

The original data of 1005 subjects was reduced to obtain groups of equal size as well as groups similar in sex proportion and age. Firstly, we excluded 79 cases in which maternal age was missing. This elimination of cases even though all of these cases are in close proximity. The cause of moyamoya disease is still unknown, but we think that there is a topographic difference in the predilection to stenotic changes in the cerebral vasculature in the disease.

The results of this study suggest that genetic mutations acquired by aging oocytes are not pivotal in the development of Parkinson’s disease. If acquired mtDNA mutations of female gametes were a significant factor in the pathogenesis of Parkinson’s disease, maternal age at birth would be higher for patients with Parkinson’s disease than for age

Figure 1 Proportion of subjects by maternal age.
matched controls. These data show no difference in maternal age at birth between patients and controls. Thus, transmission to offspring of somatic mtDNA mutations that accumulate as the mothers age is unlikely to play a part in the cause of Parkinson's disease. However, the results of this study do not exclude a role for inherited abnormalities of mtDNA mutations in this disease. Homoplasmic polymorphisms or heteroplasmic sequence abnormalities could still account for a proportion of those with Parkinson's disease. Indeed, either of these possibilities, especially heteroplasm, is consistent with the high degree of variability in the clinical expression of mtDNA mutations and the apparent sporadic occurrence of this disease.

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Hippocampal involvement in identical twins with neurofibromatosis type 1

Neurofibromatosis type 1 (NF1) is an autosomal dominant disorder due to a defect on the long arm of chromosome 17. Since 1986, when Cohen et al first drew attention to changes on MRI in NF1, it has increasingly been recognised that up to 75% of children with NF1 have lesions, seen most often in the brain stem, cerebellum, optic tracts, and basal ganglia. They are asymptomatic, usually less prominent with age, and the limited available postmortem data suggest that they involve areas of spongiform or vacuolar change. It is unclear how often temporal lobe structures are involved and figures range from 0% to 16%. We report on a pair of identical twins with NF1, with prominent bilateral changes in hippocampal MRI, one of whom presented with a major amnesic syndrome.

The twins, aged 15, were the only children of non-consanguineous unaffected parents with no known family history. Twin 1 had been considered bright throughout his schooling, until in summer 1999 he was noted to have greater than 15 cafè au lait spots and axillary freckling. Visual acuities were normal and there were no major focal neurological findings. Because of the acute memory problems herpes simplex encephalitis was considered despite a normal EEG and CSF examination and he was given acyclovir, without benefit. His brain MRI showed multiple areas of increased signal in the medulla,pons,midbrain, and internal capsule regions (fig 1 A), prominent bilateral lesions in the hippocampus, amygdala, and medial temporal cortical region (fig 1 C), and enlargement of the optic chiasm with increased signal strongly suggestive of an optic nerve glioma. Extensive blood tests including treponemal serology and chest radiographs were normal. Psychometric testing confirmed twin 1 to have an IQ in the high average range but to have a dense generalised anterograde amnesia. He was able to recall a story immediately it was told but at 5 minutes could only recollect one out of 20 details. Similarly, his immediate copy of the Rey-Osterrieth complex figure was normal (32/36) but he had almost no recall of the figure at 30 minutes (1/2 out of 36). Repeat MRI and neuropsychological assessment 1 year later showed no significant changes.

Because of the lack of a clear diagnosis on twin 1 we also studied his identical twin brother. Southern blot analysis confirmed their monozygosity. Aged 2, twin 2 developed a right facial plexiform neurofibroma and aged 8 a plexiform neurofibroma involving his tongue and larynx was surgically excised. He also had typical cutaneous lesions. His brain MRI showed remarkably similar changes to those seen in twin 1 in the medulla,pons,midbrain, internal capsule (fig 1 B), and hippocampal regions (fig 1 D) although the changes were less marked in the limbic regions in twin 2 than twin 1. The sole major difference was the normal optic chiasm in twin 2. Neuropsychometry on twin 2 disclosed a full scale IQ in the average range with no evidence of significant amnesic problem.

There are three main areas of interest that arise from these twins. The first is how remarkably similar are the changes on brain MRI. The second is that unlike previous studies they are discordant for optic nerve gliomas. The third, and perhaps most intriguing, is twin 1’s amnesic syndrome, the cause of which remains unestablished. He does not seem to have any of the conditions known to cause profound amnesia with changes on MRI, which include herpes simplex encephalitis, paraneoplastic limbic encephalitis, and possibly neurosyphilis. As he has NF1 associated with bilateral hippocampal changes on MRI it is tempting to speculate that the combination of the minor head injury and the lesions in the limbic cortex are responsible. Less likely explanations are malignant gliomatous transformation of hippocampal hamartoma or spread of malignant tissue from his optic nerve glioma.

Changes in MRI specifically within the hippocampus have apparently not been the subject of detailed analysis in NF1 and this paper suggests the need for further study of hippocampal structure and function in NF1.

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Paroxysmal hypertension during a complex partial seizure

The autonomic mechanisms involved in neurogenic paroxysmal hypertension are not understood. We present the first demonstration of the precise haemodynamic and autonomic changes during a complex partial seizure.

A 50 year old headmaster was investigated for an 8 year history of recurrent absence attacks, stereotyped in nature and of sudden onset, each lasting about half a minute. He became pale, sweaty, and mentally withdrawn initially, but after 10 minutes, he suddenly became pale, sweaty, and withdrawn for about 30 seconds. No loss of muscle tone was seen and he later confirmed that this was a typical absence attack. Coinciding with the onset of his symptoms, MNSA increased briefly for 3 seconds associated with a sudden increase in blood pressure from 138/95 to 222/150 mm Hg over 10 seconds. Heart rate simultaneously increased from 65 to 98 bpm (fig 1). Over the next 20 seconds, blood pressure and heart rate decreased and there was a major burst of MNSA followed by reciprocal oscillation of blood pressure with MNSA (0.1 Hz) as blood pressure reached normal levels. During recovery, he complained of his usual transitory headache. Venous noradrenaline (norepinephrine) concentrations were 1650 pmol/l and 5250 pmol/l before tilt and during recovery respectively. Normal values in our laboratory before and after 10 minutes of tilt are 456 (SD 50) and 705 (SD 74) pmol/l. His absence symptoms could not be reproduced by rapidly increasing blood pressure to similar values (250/120 mm Hg for 30 seconds) with an intravenous bolus of epinephrine (100 µg). One week later, an EEG during a similar absence attack showed sharp waves arising from the left frontotemporal area (fig 2). Subsequent continuous EEG and blood pressure monitoring confirmed that focal seizure activity was simultaneous with paroxysmal hypertension. Studies with MRI showed hippocampal atrophy consistent with the diagnosis of complex partial seizure disorder. His absences were abolished with 400 mg carbamazepine daily and he has remained free of symptoms for 6 months.

![Figure 1](image1)

**Figure 1** A 2 minute recording of blood pressure (BP), muscle nerve sympathetic activity (MNSA), and heart rate (ECG) during an absence attack after 10 minutes of head up tilt. At 30 seconds there was sudden mental withdrawal and a rapid increase in MNSA followed by a severe and paroxysmal increase in blood pressure and heart rate. As blood decreased, MNSA increased and when blood pressure normalised there was a marked baseline shift in MNSA. During recovery, blood pressure and MNSA oscillated reciprocally (0.1 Hz).†

This is the first demonstration of paroxysmal neurogenic hypertension triggered by a seizure in a patient with complex partial seizure. The diagnosis of complex partial seizure was supported by the following: focal EEG changes during a subsequent absence seizure; no reproduction of absence symptoms during drug induced paroxysmal hypertension; characteristic hippocampal atrophy on MRI; and complete response to anticonvulsant drugs. Other possible diagnoses including brain stem tumour, phaeochromocytoma, and renal artery stenosis were excluded by appropriate imaging and neuroendocrine analysis. Pseudoseizures were excluded on the basis of the EEG findings and the rapid response to treatment. Although rapid increases in MNSA and heart rate have been found during panic attacks, paroxysmal hypertension and loss of consciousness are not consistent features.1

The paroxysm consisted of simultaneous hypertension and tachycardia associated with sweating and facial pallor during the absence attack. We suggest that this is secondary to a generalised increase in sympathetic activity causing vasoconstriction and increased cardiac output. This is supported by (a) increased MNSA and heart rate despite progressive rise in blood pressure; (b) sympatoexcitation with blood pressure overshoot; (c) noradrenaline increased to over seven times the normal tilt levels; (d) prominent low frequency (0.1 Hz) oscillations in blood pressure and heart rate during recovery. These low frequency oscillations (0.1 Hz) are thought to be secondary to changes in brain stem sympathetic activity separate from the effects of respiration, which are generally of a higher frequency (0.5 Hz). We emphasise that the initial increase in MNSA occurred when blood pressure was increasing and so was not baroreflex mediated as would be expected for respiratory or normal brain stem low frequency oscillations.1

We hypothesise that this generalised increase in sympathetic activity is permitted by a transient interruption of baroreflex feedback inhibition during the seizure. We think that this is a unique recording of transient baroreflex failure characterised by a rapid and generalised increase in sympathetic activity, overriding the baroreflex afferents in the brain stem. It has long been suspected that paroxysmal hypertension occurs in complex partial seizures but to date, ambulatory monitoring has only demonstrated changes in heart rate.1 Ambulatory beat to beat blood pressure monitoring would allow closer study of this phenomenon and its possible relation to sudden cardiac death in epileptic patients. Finally, this a good example of an episodic medical condition which may be very difficult to diagnose. Occasionally, when an episode is seen fortuitously in the laboratory, we may identify pathophysiology previously suspected but not actually seen.

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Compression by looping and perforation of the facial nerve by the anterior inferior cerebellar artery: a possible cause of facial tic

Hemifacial spasm is a distressing, common, and well defined condition, which is difficult to treat. It is characterised by clonic and tonic contractions of the muscles supplied by the facial nerve, is intermittent, and is usually worsened by emotional upsets. A clinical picture similar to that of hemifacial spasm is also seen in trigeminal neuralgia, another condition for which widely varying pathophysiological bases, including vascular compression, have been proposed.1

In our case postmortem exploration of the posterior cranial fossa disclosed a strikingly abnormal relation between the anterior inferior cerebellar artery and VIIth cranial nerve. Such abnormal vascular anomalies around the facial nerve are repeatedly reported in the literature, and they seem to be closely correlated with hemifacial spasm.2

We took the cadaver of a man who had died aged 51, from the dissection material held by the First Department of Anatomy at the University of Vienna.

A square 2 cm×2 cm was drilled in the centre of the skull cap, formalin:water (1:5; 30 ml) was injected subdurally. The cadaver was left in the cold room for 24 hours. Then a mixture of phenol:formalin:water (3:1:10) was introduced into the femoral artery about 5 cm below the inguinal ligament through a 3 mm diameter cannula. Before preparation the cadaver remained in this phenol:formalin:water mixture for about 6 to 8 months.

The vascular anomaly reported was noted when the brain was moved carefully backwards. The brain, which was otherwise normal, was then dissected out of the cranial cavity. The external radius of the anterior inferior cerebellar artery was determined by a digital gauge.

During the exploration of the posterior cranial fossa of the cadaver, we encountered an unusual course of the right anterior inferior cerebellar artery (1.20 mm diameter), which arose from the lower third of the basilar artery and passed through the facial nerve 1.5 cm from the cerebellopontine angle, forming a ring around the nerve about 0.5 cm proximal to the point of penetration and compressing it at several points (fig 1). The anterior inferior cerebellar artery first ran vertically downwards closely following the basilar and the right vertebral arteries. Then it took a horizontal course until it made a V shaped angle before penetrating the seventh cranial nerve. After it had passed through the facial nerve, with about one third of the fibres above it and two thirds below, its course made a ring around the seventh cranial nerve and disappeared between the cerebellum and the pons into the deep tissue. Along its course a branch went off to the choroid plexus of the fourth ventricle.

The cause and treatment of hemifacial spasm remain controversial. Whereas some theories postulate a cerebral or brain stem mechanism for its origin, others suggest that the causative lesion is within the facial nerve, either within the posterior fossa or at a more distal location. Our unusual finding lends support to the neurovascular compression theory to explain the aetiology of this disorder as we would like to point out the unusual nature of this

Figure 1 Inferior surface of the brain. A=Anterior inferior cerebellar artery; F=facial nerve (seventh cranial nerve); C=cerebellum; P=pons; V=vestibulocochlear nerve (eighth cranial nerve).

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anatomical variety of the anterior inferior cerebellar artery.

During an investigation of the posterior cranial fossa, we became aware of an abnormal relation between a vessel and a nerve, which is not described in the current textbooks and encyclopedias. Although there is much literature reporting a close relation between the symptom “facial tic” and vessel variety; our variety—that is, with both transfixion of the facial nerve and an arterial loop around the same nerve—has not been described in the specialist literature, nor has it been mentioned in the most recent review of variants.

Several authors have provided illustrations of a loop formed by the anterior inferior cerebellar artery, but without elaborating further on the topographic relation between the artery and the internal acoustic meatus or the seventh and eighth cranial nerves. It has been asserted that this vessel seldom appears on radiographs.

Typical hemifacial spasm is caused by vessels on the anterocaudal side of the nerve, whereas atypical hemifacial spasm is caused by vessels on the posterocaudal side of the nerve.

The relevant aspect of this article lies in its emphasis on the connection between the neurological symptoms and this anatomical variety of a nerve.

The deceased had begun to experience intermittent symptoms of varying intensity in his face at the age of 49. These symptoms took the form of uncontrollable twitching at the right corner of his mouth, ipsilateral hearing impairment, retroauricular cramps, and retroauricular sensory impairment. According to the case history, the deceased had undergone a full otorhinolaryngological examination and pure tone speech audiometry during his lifetime. Thus, it was possible to diagnose the perceptive unilateral acoustic hypoauscusia on the right.

Early auditory evoked potential studies had also been performed showing an increase in latency and a decrease in amplitude without any deterioration of morphology of the waves. It had not been possible to use MR for the diagnosis as he had a pacemaker in place. None of these symptoms responded to therapy with botulinum toxin.

In our case the compression of the facial nerve at several points could have led to irritation of the region supplied by the posterior auricular nerve and in this way to the symptoms described above.

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Effects of topiramate on cognition

This letter concerns the recently published study by Thompson et al, regarding the authors’ findings on cognitive effects with topiramate. Firstly, I want to correct the authors’ mischaracterisation of a review paper of mine. The authors state that “the literature on antiepileptic drugs ... emphasised positive psychotropic effects,” referencing only a 1998 review in which I discussed cognitive and psychotropic effects of antiepileptic drugs. Although I mention some positive effects, I also discussed negative effects, including studies from my own centre that have shown significant negative psychotropic and cognitive drug effects.

Secondly, I provide some perspective on the report of Thompson et al of clinically significant cognitive decline in patients treated with topiramate as adjunctive therapy. The authors correctly conclude that “caution is warranted in the interpretation of the findings due to methodological limitations of the study design.” Because their study was retrospective and observational, it is subject to considerable subject selection bias. For example, five of the 18 patients were specifically included in the topiramate sample because they reported cognitive effects.

The only way to minimise effects that may bias study conclusions is to conduct a prospective randomised controlled study. Two such studies have recently compared topiramate and valproate as add on therapy to carbamazepine, using comprehensive neurophysiological batteries to objectively measure drug effects. At the end of 3 months of maintenance therapy, only one of 17 (6%) variables in one study and only two of 30 variables (7%) in the other were significantly worse for topiramate compared with valproate. For the three variables with statistically significant differences, the mean differences in change scores were modest. Analysis of individual data showed that scores were unchanged or even improved in most patients receiving topiramate and valproate. Statistically significant differences could be accounted for by a minority of patients receiving topiramate in whom scores deteriorated >1 SD from baseline. I suggest that the patients reported by Thompson et al likely represent a similar subgroup of patients.

Physicians should be aware that a subgroup of patients treated with topiramate may experience clinically significant cognitive effects. When these effects occur, they are generally apparent to the patient or family members and can therefore be monitored with routine clinical evaluations. Alternatively, a brief cognitive test (for example, a verbal fluency test or symbol digit modalities test) should easily detect changes of the magnitude reported. In a subgroup of patients, topiramate may need to be discontinued if cognitive effects do not resolve over time with slowed titration or dosage reduction.

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Postoperative hearing loss due to venous congestion at the inferior colliculus, or cochlear dysfunction?

I read with interest the article by Strauss et al. on postoperative contralateral hearing loss which developed on the third day after microvascular decompression for trigeminal neuralgia. The patient noted symptoms to venous congestion at the ipsilateral inferior colliculus after dissection of the ponto-trigeminal vein, which was documented by MRI. Symptoms resolved partially after intravenous nimodipine for a total of 19 days. The authors’ explanation for the delayed postoperative hypacusis, however, merits further discussion. Strauss et al. provided preoperative and postoperative recordings of brain stem auditory evoked potentials: postoperatively, after stimulation on the operated side, ipsilateral waves I through V, and contralateral waves II through V are all clearly identifiable, contrasting with stimulation of the non-operated side, despite only a small wave V bilaterally, but no other components. This pattern suggests a left sided lesion involving the generator of wave I—presumably the auditory nerve near the cochlea—and is also consistent with the patient’s pans cochlear hearing loss.1,2 By contrast, brain stem lesions—even damaging the cochlear nucleus—are usually not associated with pure tone hearing loss, but rather with abnormal auditory localisation or interaural time discrimination,3 as auditory impulses are conveyed bilaterally in the brain stem.1,4,5

Furthermore, a brain stem lesion causing profound hearing loss is likely to produce also contralateral wave IV/V abnormalities after stimulation on the non-affected side, but even the severest brain stem lesions, such as in evolving brain death, do not affect wave I.4 The vascular supply of the mesencephalic brain stem differs from that of the inner ear, the first being fed by meningeal arteries via the posterior cerebral or superior cerebellar artery, and the second by the superior petrosal or transverse sinus, and the internal jugular vein.6 Thus, the causal pathway within the brain stem, the “isolated vasospasm theory” seems unlikely.

Strauss replies:

We appreciate Kofler’s comment on our paper and his interest in this unusual and still poorly understood clinical picture. We agree that the brain stem auditory potentials (BAEPs) after contralateral stimulation do not clearly point to a lesion of the ipsilateral colliculus; however, to our knowledge the neurophysiology of auditory pathways within the brain stem is not yet fully understood. For example, in our series of more than 300 cases of acoustic neurinoma monitored using BAEPs, we have never seen that the contralateral wave V is much more pronounced compared with the ipsilateral wave V. The advantage of this case report is the preoperative and postoperative radiological and clinical documentation. The delayed onset of symptoms several days after the surgical procedure, the lack of effect of calcium blocker therapy—actually the patient’s pure tone audiogram and speech discrimination deteriorated under nimodipine treatment—and the hearing improvement after heparinisation do not suggest vasospasm as the underlying pathophysiological mechanism. The literature on this rare and important phenomenon of postoperative hearing loss after cerebellopontine angle surgery is purely speculative. By contrast in this case report follows a straight course, which started at surgery with dissection of the pontotrigeminal vein, followed by a delayed contralateral hearing loss, and ended with a lesion of the ipsilateral colliculus. This lesion was not documented on preoperative MRI. Taking these findings into consideration, together with the neurophysiological findings of BAEPs in a still not fully understood auditory pathway within the brain stem, the “isolated vasospasm theory” seems unlikely.

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This book has arisen because of the comparative dearth of studies examining the emotional and behavioural consequences of focal brain lesions and, if nothing else provides a useful component of the repertoire in 18 chapters written by experts in the field. The first three chapters set the scene by providing a conceptual overview and discussions of methodological issues. The rest are devoted to specific topics and generally take the form of either examinations of the evidence for the localisation of specific neuropsychiatric syndromes (depression, mania, obsessive compulsive disorder, psychosis) or reviews of the effects of lesions of particular structures (thalamus, basal ganglia, frontal lobe, temporal lobe). In their overview, Cummings and Bogousslavsky group the diseases into mood, thought, affect, and motivation considered in this book under the rubric “fundamental dysfunctions” which they point out are associated with interruptions of limbic and frontosubcortical circuitry. This diffuse nature of these circuits is reflected in the finding that lesions of disparate areas can produce, for example, psychotic phenomena and that lesions in specific structures—for example, thalamus or basal ganglia, can result in a range of behavioural and emotional abnormalities. It is thus often difficult to make strong inferences from this literature about the neural basis of emotion and behaviour. Indeed some chapters are essentially categorised lists of studies without much in the way of theoretical underpinning. Several chapters, by Habib (disorders of motivation), Eisinger and Gelen (behavioural and emotional changes after focal frontal damage) and Tranel (neural correlates of violent behaviour), stand out. These stray from localisation and bring in evidence from basic neuroscience and human functional neuroimaging to inform and interpret fascinating case histories in terms of a functional neuroanatomy of emotion and behaviour.

EILEEN JOYCE

Update in neurology for general practitioners. By P O'ROTON, D BATEMAN, G FULLER, P NEWMAN, D PARK, R SHAKER, and O YOUNG. (Pp 218 + 2 CD—ROM, £80.00.) Published by University of Bath/British and Spine Foundation/Royal College of General Practitioner’s, Bath, 2000.

The seven authors of this text are to be congratulated on their innovative approach and the freshness they bring to the evaluation of the common neurological problems encountered in primary care. This text and the two accompanying CD ROMs represent a cooperative venture between the British Brain and Spine Foundation, the Royal College of General Practitioners and the University of Bath.

The text addresses the commonly encountered problems of headache, sensory disturbance, cerebrovascular disease, dementia, confusion, movement disorders, fits and fants, and back, leg, and arm pain. The approach is a simple one—namely, a definition of the condition or conditions, who gets it? What causes it? What do we do? What is the differential diagnosis? How is it investigated and managed? This pragmatic approach pays special attention to the prevalence of each disorder in the community and contains “red flags” which suggest diagnostic doubt and uncertainty. The two accompanying CD ROMs outline learning outcomes, case histories, self assessment questionnaires, and case pathways with respect to investigation, management, and prognosis. Each such case is linked with the text providing assignments in medical audit, comment, documentation of data, and “setting standards”. Incorporated in all of this are useful components such as data collection sheets for specific symptoms and summary sheets to aid and improve documentation.

I found this an interesting book and enjoyed finding my way around the CDs, which were easy to use. I would imagine that this would be a valuable tool for the busy general practitioner and should help set standards in primary care that would aid patients and ensure thorough evaluation before referral. The only thing missing was guidance demarcating between evaluation in the primary care setting from the need to refer on for a specialist opinion. All of us occasionally think, under moments of work overload, that our colleagues in primary care could be a little more circumspect in whom they refer (although the truth of the matter is that most referrals are thoughtful and necessary). A text such as this with no obvious competitor, is long overdue, written by neurologists who, along with their general practitioner colleague, have adopted a common sense approach to their task. I would strongly recommend this book with its accompanying CD ROMs to my general practitioner colleagues and others (with von Cramon) “that recovery of visual function is possible and meaningful” (p 34). Of course, complete V1 lesions are almost always accompanied by additional damage, when animal work demonstrates a reduced residual capacity. But a complete but restricted V1 remnant in the monkey still allows further recovery to take place.

Anyone wishing to find practical approaches for helping patients across a range of visual disorders will find this useful, humane, and dedicated book. He or she will have to work through a compact monograph and share Zihl’s experience with him, rather than find a collection of shorthand recipes. It will be rewarding work.

IAN BONE


This is the second series on neuropsychological rehabilitation fostered by Barbara Wilson and Ian Robertson (the first being Neural repair, transplantation, and rehabilitation by R Barker and S Dunnett). The renewed interest is welcome. Josef Zihl is one of the first clinical scientists, together with collaborators such as von Cramon, to have put a concentrated effort into rehabilitation of visual disorders, spanning a period of 2 decades. This book is a testament of this pioneering work in Germany, stemming from an older tradition going back to Poppelreuter, and the current book should make the developments better known outside that country. It remains something of a curiosity that rehabilitation is scarcely pursued in Britain and the United States, especially given the seminal animal work (by Cowey in Britain and Mohler and Wurtz in the United States) clearly demonstrated the potential for recovery after V1 lesions in the monkey, with practice regimes. Indeed, Zihl’s own work acknowledges the influence of these animal studies on his own research.

The Series preface describes this work as a “modular handbook.” Handbook it is not. Rather it is a compact and relatively short account that is more like a progress report, full of interesting approaches described in midstream of the research, which is still to be completed. It is broad ranging, the largest and most advanced section (about half of the book) being on visual field disorders, but progressing to visual acuity, and visual space perception (including Balint’s syndrome), visual agnosia, and a final brief section on the special problems of central scotomas. Each section starts usefully with evidence of spontaneous recovery from the lesion, before the baseline from which rehaesthetic efforts must be assessed. Such efforts are almost entirely based on intensive practice (what I have called “grindsight”). Zihl is very cautious, raising the evidence well, stating such as “our observations produce preliminary evidence that spatial contrast sensitivity can, in principle, be improved by specific and systematic practice. However, the limited number of cases does not allow definite conclusions to be drawn ......” (p 96). He is also careful to distinguish between measurable effects in the laboratory and their “ecological validity” to the subject in terms of “its import to everyday life. In some regards he is over-cautious—for example, in his speculation (with von Cramon) “that recovery of visual field defects can only be expected with complete striate cortex injury” (p 34). Of course, complete V1 lesions are almost always accompanied by additional damage, when animal work demonstrates a reduced residual capacity. But a complete but restricted V1 remnant in the monkey still allows further recovery to take place.


L. WEISKRANTZ

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Dr Lawrence Robbins, from Rush Medical College, Chicago has revised his deservedly popular 1994 text on the management of idiopathic headache syndromes. He shares a wealth of clinical experience in the drug therapy of difficult headache patients, with necessarily brief descriptions of clinical features and pathogenesis. Although there is an extensive list of references at the end of the book, they are not cited directly in the text, and the book reads as one man's recipe book rather than a systematic review of published trials, or of the pharmacological or clinical evidence supporting management decisions. It is inevitable that the range of drugs recommended is American, with more emphasis on narcotic and barbiturate combinations and no mention of domperidone or pizotifen, but there is a useful glossary of United States trade names as an appendix.

The chapters dealing with the acute and preventative management of migraine, tension headache, and cluster headache are followed by illustrative case histories, from which it is soon apparent that the tendency of United States patients to see many neurologists and complain more about side effects can lead to the use of an extremely wide range of drugs, sometimes making one's own multiple efforts on behalf of a challenging tertiary referral seem straightforward in comparison. There is a particularly useful chapter on headache in patients aged over 50, post-traumatic headache, lumbar puncture headache, chronic paroxysmal hemicrania, and SUNCT.

Although filled with much clinical wisdom from a vastly experienced author, the book has the feel of a catalogue; for European readers it is more likely to have a role as a desk reference text.

RICHARD PEATFIELD


This is a short text of 306 pages in 17 chapters covering vascular dementia. Although filled with much clinical wisdom from a vastly experienced author, there are only 11 authors so many of the chapters have authors in common. The principal editor, Dr Meyer, coauthors no less than 10 of the 17 chapters. As a result of this many authorities are not included among the list of contributors and some of the chapters are not fully authoritative.

The text gives a fairly good overview of the subject, although there are some unusually prominent inclusions including full chapters on plasmapheresis and estrogen replacement therapy in the treatment of vascular dementia, neither of which are recognised treatments. Some of the more important recently recognised conceptual issues, including the very high preponderance of mixed dementia (vascular dementia and Alzheimer's disease) and the issue of diagnosis before reaching the state of being demented are scarcely covered at all. The limitations of the current formal diagnostic criteria for vascular dementia are touched on, but not considered in detail and the chapter on diagnosis really does not help in this respect.

Overall, the book provides an adequate background to the subject but a person interested in the topic would need to look elsewhere for some of the more recent critical issues.

JOHN BOWLER


A wide range of textbooks are now available on the topic of headache, ranging from short notes of 40 or 50 pages up to encyclopaedic tomes. This book falls into the middle of the range, being an easy read of 165 pages.

The work is divided into three broad categories: (1) pathophysiology and epidemiology of headache, (2) primary headache disorders, and (3) secondary headache disorders.

In the first section, the starting point is the International Headache Society (IHS) classification but the bridge to the clinical approach to patients is rapid and the use of disability assessment tools is discussed. The epidemiology of the various forms of headache is then examined, together with impact on the sufferer. An entire chapter is devoted to diagnostic testing, particularly to exclude ominous causes of headache, and the pathophysiology of primary headache is discussed, the focus being on migraine. Genetics, anatomy, and physiology are all investigated. This whole section is concise and informative, giving a very good overview of the subject.

The rest of the book is divided into chapters dealing with specific types of headache, the first section looking at migraine, tension-type headache, chronic daily headache, cluster headache, and related symptoms, the final section covering secondary headache, including post-traumatic headache, headache associated with disease of the intracranial cavity, sinus headache, headache associated with CNS infection, headache associated with pregnancy and breast feeding, and geriatric headache. Within these chapters, all types of presentation and management are discussed, the emphasis being on use and application of the IHS classification in a clinical setting.

This book undoubtedly contains the full range of information necessary to deal with the treatment of headache. However, the clinical chapters are written from a predominantly American perspective and, therefore, the therapeutic approaches in particular do not necessarily correlate with those used in the United Kingdom. Because of the comprehensive nature of the work, there is a danger that the reader may not appreciate the relative importance of some of the various conditions discussed. For instance, my own experience has shown that the greatest clinic problem, even in a primary care setting, comprises patients with chronic daily headache and analgesic dependence. Important basic clinical issues such as this may be lost within the wealth of information set out here.

My overall impression is that this is a very comprehensive book which would serve as a good reference. However, although it would be extremely useful for the primary care doctor in the United States where more specialisation exists, its value in the United Kingdom may be limited to the secondary care doctor or to the few primary care doctors who have a specific interest in headache.