Retro-ocular headache with autonomic features resembling “continuous” cluster headache in lateral medullary infarction

Headache is a common, although underemphasized, complaint of lateral medullary infarction. More than half of patients with posteriorinferior cerebellar artery (PICA) infarcts develop headache. Fisher found headache in 22 out of 41 (54%) patients with lateral medullary infarction and, more recently, Kuwabara and Hirayama reported headache in 26 out of 34 (76%) patients with Wallenberg’s syndrome. Less than 5% of patients with PICA infarcts, however, develop pericranial, hemicranial headache and although sympathetic dysfunction in the form of Horner’s syndrome, is a well known manifestation of lateral medullary infarction,1 signs of parasympathetic overactivity, such as lacrimation, eye injection, and nasal congestion, have never been described in lateral medullary infarction. Here we report on a patient with a lateral medullary infarction who developed anterior hemicranial pain accompanied by severe and persistent autonomic parasympathetic activation.

This heavy smoking 37 year old man came to our hospital due to acute vertigo, lack of coordination of his left limbs, numbness in his right limbs, dysarthria, and dysphagia. An angiographic study performed in another hospital 3 months earlier due to intermittent claudication and decrease in left radial pulse had disclosed atherosclerotic changes in the lower cervical and in the left subclavian arteries. General examination showed reduced pulses in the left radial and right dorsalis pedis arteries. Abnormal signs at neurological examination included hiccup, nystagmus, left Horner’s syndrome, and facial hypoesthesia, left velophalatine weakness, right hemicorporal hypoesthesia, and ocular signs of parasympathetic overactivity, such as lacrimation, eye injection, and nasal congestion, had been never described in lateral medullary infarction. Here we report on a patient with a lateral medullary infarction who developed anterior hemicranial pain accompanied by severe and persistent autonomic parasympathetic activation.

From the beginning of his clinical picture this patient complained of two clearly differentiated head pains. The first one was located in the right occiput and progressively disappeared during the first week after the acute stroke. The second was a very disturbing, continuous headache located anteriorly, mainly in the left retro-ocular and temporal region. This pain was described as moderate to severe, steady, or boring and constantly accompanied by ipsilateral conjunctival injection, lacrimation, and nostril blockage or rhinorrhea (fig 2). The patient also had one or two dramatic daily exacerbations of unbearable pain intensity together with an increase in autonomic symptoms and signs lasting about 2–4 hours. The pain did not significantly respond to either oral or intravenous analgesics (paracetamol, aspirin, metimazol, and NSAID). Verapamil, 240 mg daily, plus sodium naproxen, 1100 mg daily, slightly reduced the pain for 2 or 3 weeks. Medications containing ergotamine, methysergide, and agonists of the 5-HT1/B/D receptors were not prescribed. The pain remained unchanged for several days, after which it began to progressively improve and disappeared 6 months after the stroke.

To the best of our knowledge, this is the first reported case of lateral medullary infarction with unilateral anterior headache, sympathetic dysfunction, and parasympathetic autonomic activation, all this resembling a “continuous” cluster-like headache syndrome. The pathophysiology of trigeminal autonomic cephalalgias, including cluster syndrome, is largely unknown. It has been proposed, on anatomical grounds, that an inflammatory process in the cavernous sinus, as a point of intersection of the first division of the trigeminal nerve and the cranial sympathetic and cranial parasympathetic nerves, would be the pathophysiological explanation for these headaches.

As commented on, headache is a frequent complaint in lateral medullary infarction. The posterior is the most common location for headache, 65% in the series of Kuwabara and Hirayama and 49% in Fisher’s series 1,2 which concurs with Wolff’s finding on stimulating the vertebral artery the pain is referred to an occipital-suboccipital-nuchal area. This posterior pain was also reported by our patient during the first week, probably being related to the thrombus formation in the vertebral artery. The most important pain experienced by our patient, however, was a moderate to severe, unilateral, retro-ocular headache. Anterior pain affecting the eye region is much less frequent in lateral medullary infarction, but was already considered by Fisher as typical of this syndrome and probably resulting from a lesion in the nucleus of the descending root of the trigeminal nerve, as anterior pain cannot be induced on stimulating the vertebral artery and as this anterior headache is usually succeeded by numbness. The central medullary lesion also accounts for this autonomic symptoms and signs seen in this patient. Sym pathetic dysfunction (Horner’s syndrome) is a well known sign of Wallenberg’s syndrome due to damage of the sympathetic tract within the PICA infarction and it seems logical to try to explain the parasympathetic activation as secondary to irritation of the adjacent superior salivatory nucleus by the infarct area or to an imbalance between the two autonomic systems.

In conclusion, this patient shows for the first time that all the semiology typical of cluster and other trigeminal-autonomic headaches can be secondary to a pure central lesion, located in the lateral medulla, this supporting the contention that flow changes occasionally found in cluster headache cases in the cavernous sinus do not generate the disorder, but are in fact a consequence to the pain. This agrees with the proposal that the activation of the cavernous sinus region does not relate specifically to cluster headache, but it is a trigemino-novascular autonomic reflex to first division pain. In fact, the only difference between our case and typical cluster headache—the periodicity of the pain attacks—is easily explained by the distinct central generators giving rise to trigeminovascular system activation—the hypothalamus for idiopathic cluster headache3 and the established lesion of the key structure of the trigeminal nucleus caudalis, in this case.

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![Figure 1](image1.png) T2 weighted MRI showing an infarct in the left PICA territory.

![Figure 2](image2.png) Close up picture of the patient’s eyes showing left palpebral piosis and conjunctival injection.

Chronic autonomic neuropathy in a patient with primary Sjögren’s syndrome

Several investigators have described the autonomic neuropathy in Sjögren’s syndrome. However, only a few have documented the details of dysautonomia, or the pathology of the nerves or of other organs such as eccrine sweat glands. We report on a patient with Sjögren’s syndrome in whom dysautonomia was the dominant feature, and describe histological findings for the sural nerves and the eccrine glands.

A 39 year old woman visited our hospital because of frequent fainting attacks and diminished sweating. Her history included attacks of dizziness on standing since the age of 27. At the age of 30, she experienced a fainting attack on standing. At the age of 36, she noticed dry eyes and focal loss of sweating on the left forehead. She also developed amenorrhea at the age of 27. The family history was unremarkable.

She was 164.5 cm tall and weighed 36 kg (body mass index 13.4). Neurological examination showed bilateral ptosis and isocoric pupils (3 mm in diameter; measured using infrared photography) with an irregular margin. The light reflex was absent and the accommodation reflex was tonic. Muscle tone and power were normal. The tendon reflexes were absent and plantar responses were flexor. Sensation and coordination were normal. The skin was generally dry, and spontaneous sweating was present only over the right forehead and T-10 and T-11 dermatomes in hot conditions. The urinary system was normal.

Results of routine laboratory examinations, chest radiography, thyroid function tests, and thyroid hormone tests were normal. Urinary porphobilinogen (0.5 mg/day), α-methylenadolinic acid (1.8 mg/day), and serum vitamin B12 (679 pg/ml) were normal. Urinary porphobilinogen (0.5 mg/day), α-methylenadolinic acid (1.8 mg/day), and serum vitamin B12 (679 pg/ml) were normal. Fainting attacks on standing disappeared after the treatment. The patient was treated with oral prednisolone (40 mg/day) and l-threo-3, 4-dihydroxyphenylserine (200 mg/day). The dose of prednisolone was tapered to 20 mg/day. Fainting attacks on standing disappeared after the treatment. The patient had bilateral tonic pupils, bilateral Horner’s syndrome, hyporeflexia, orthostatic hypotension, abnormal cardiovascular reflexes, reduced lacrimation, and segmental anhidrosis. Examination of the autonomic nervous system disclosed parasympathetic and postganglionic sympathetic injury including sudomotor involvement. Since Ross reported the presence of tonic pupils and sympathetic denervation in his patient (Ross’ syndrome), several reports have confirmed that autonomic dysfunction in Adie’s syndrome may be more widespread than previously recognized. However, severe orthostatic hypotension as seen in our patient is rare. The clinical condition of our patient seemed different from that of acute autonomic neuropathy, because the dysautonomia started insidiously and progressed slowly over many years. Our patient did not have any other systemic disease except Sjögren’s syndrome. Peripheral neuropathy with Sjögren’s syndrome may represent a distinct syndrome, in which sensory polyneuropathy predominates but there is some autonomic dysfunction or Adie’s pupils. Griffin et al described autonomic deficits in patients with ataxic sensory polyneuropathy with Sjögren’s syndrome. In their patients, histological examination of sural nerves showed a preferential loss of large myelinated fibres, which is different from the findings of our patient. We concluded that the diagnosis of our patient was a subtype of polyneuropathy with Sjögren’s syndrome.

The marked reduction in the density of unmyelinated fibres seen in our patient may reflect damage of the postganglionic sympathetic efferent projection. We showed the severe degeneration of postganglionic sympathetic cholinergic nerves innervating the eccrine glands by histopathological morphometric analysis. Examination of the eccrine glands would provide useful information for assessment of autonomic function in patients with anhidrosis.

We thank Dr Shinji Oono, Department of Ophthalmology, Saga Medical School, Japan, for his help and advice in the examination of pupils.

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C1/C2 rotary subluxation due to spasmodic torticollis

Rotary subluxation of the atlantoaxial complex is encountered more often in children than in adults. It is usually associated with a clear history of cervical trauma, upper respiratory infection, recent head or neck surgery, or rheumatoid arthritis. This paper reports on an adult presenting with C1/C2 rotary subluxation in whom all of these causes were absent. The subsequent clinical course

Electron microscopic morphometric evaluation of the transverse profiles of secretory coils in the skin and nerve terminals and unmyelinated axons around secretory coils

<table>
<thead>
<tr>
<th>Control†</th>
<th>Patient‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perimeter of transverse profile (µm/Profile)</td>
<td>173 (55)</td>
</tr>
<tr>
<td>Area of transverse profile (µm²/Profile)</td>
<td>2017 (1100)</td>
</tr>
<tr>
<td>Nerve terminals (n)</td>
<td>9.1 (4.8)</td>
</tr>
<tr>
<td>Unmyelinated axons (n)*</td>
<td>12.7 (6.2)</td>
</tr>
</tbody>
</table>

†Mean (SD) of six control subjects in whom 14–38 transverse profiles of secretory coils in each individual were morphometrically evaluated.

‡Mean of 38 transverse profiles of secretory coils. Minimum–maximum values are shown in parentheses.

*Number/100 µm of perimeter of transverse profiles.
showed that the patient in fact had spasmodic torticollis which we think was the cause of the rotary subluxation at C1/C2.

A 37 year old man presented in November 1997 with a history of an insidious onset of progressive deformity of the neck associated with severe utilitarian head tilt and left sternocleidomastoid and trapezius muscles and occipital pain from April of that year. He had had mild neck pain for several years. The occipital pain was left sided and became increasingly severe. Electromyography was not performed. He had been off work for 6 months and found it increasingly difficult to sleep in a comfortable position. The only relevant history was one of anxiety attacks.

On examination there were no signs of rheumatoid arthritis. The patient had a classic “cock robin” deformation with his head tilted to the left and turning to the right. This was associated with spasm and tenderness, but no obvious hypertrophy, of the left sternocleidomastoid and trapezius which was thought to be voluntary as it subsided when the anaesthetic, the element of spasm and tilting of his neck which was thought to be due to persistent subluxation and dislocation. He exhibited symptoms of the “cock robin” deformation. Spinal CT in the neutral position confirmed a C1/C2 rotary subluxation which reduced with the head turned towards the left and was exaggerated turning to the right. There was no history of recent neck injury, rheumatoid arthritis, or pharyngeal infection and thus a cause for the C1/C2 rotary subluxation was not apparent at that stage. T1 weighted MRI of the neck showed no evidence of a primary vascular lesion causing and presenting a surgical stabilisation of C1/C2 was suggested.

The patient was placed in halo-traction for a week, and further CT was performed. This showed significant improvement but not total correction of the rotary subluxation. As reduction was not total, it was decided not to perform transarticular screw fixation of C1/C2 but a posterior modified Gallie fusion was performed with a screw and hooks of maximum reduction. A halo-vest was applied. Check radiography was satisfactory and the patient reported a very pleasing relief of the pain and spasm which he had preoperatively. The halo-vest was maintained for 8 weeks during which his pain and spasm had completely resolved. However, shortly after removing the halo, he had a recurrence of the pain and spasm in the left sternocleidomastoid although the severe occipital pain was still completely relieved. His neck posturing was now variable and typical of the dystonia which he had preoperatively. The halo-vest was main-

The halo-vest was main-

...should be considered. Botulinum toxin will not resolve the subluxation, but was necessary in this case to control the underlying dystonia. External 

braces and collars rarely control the forceful movements of cervical dystonia, and the toxin 

can take some days or even weeks to work, so we recommend treatment as soon as dystonia is diagnosed. In theory botulinum toxin might enhance the effect of or interfere with recovery from acute muscle paralysis following general anaesthesia. However, no such reactions have been reported in 12 years of extensive experience worldwide with botulinum toxin, and therefore it is probably safe to give injections even before neck surgery. When a patient with spasmodic (variable posturing) torticollis develops a fixed and sufficiently extreme “cock robin” posture, the clinician should consider investigation by plain radiography and CT to exclude rotary subluxation, even if the muscle spasm is intermittent.

S DALVIE

A P MOORE

FINDLAY


Bradshaw RB. Perphenazine dystonia presenting as recurrent dislocation of the jaw. J Laryngol Otol 1969;83:79–82.


Ataxic form of Guillain-Barré syndrome associated with anti-GD1b IgG antibody

Richter proposed an ataxic variant of Guillain-Barré syndrome, in which patients have severe ataxia of the cerebellar type at the onset of Guillain-Barré syndrome but no ophthalmoplegia or severe loss of proprioceptive sense. Patients with ataxic Guillain-Barré syndrome have distal paresthesias, areflexia, and raised CSF protein concentrations. Kusunoki et al reported that of 149 patients who had anti-GQ1b IgG antibodies without profound weakness, five had acute self limited ataxia without ophthalmoplegia. The nosology of these patients, however, was not discussed. Of our 340 consecutive patients who had anti-GQ1b IgG, six had no external ophthalmoplegia and one had minimal external ophthalmoplegia. The two patients of these seven anti-GQ1b-positive patients were consistent with an “ataxic form of Guillain-Barré syndrome” (Yuki et al, unpublished observations). Tentative diagnoses made by the primary physicians were Guillain-Barré syndrome associated with anti-GD1b IgG antibody.


8 Bradshaw RB. Perphenazine dystonia presenting as recurrent dislocation of the jaw. J Laryngol Otol 1969;83:79–82.


The outcome of tuberculous meningitis is influenced by the stage of the disease at the start of treatment. Initiation of antituberculous therapy is often delayed because of the inad-

equacy of presently available laboratory tests. Management of the course of the illness is possible if the diagnosis is made early. The three isozymes of ADA are not similarly distributed in infected brain tissue. The only enzyme present in the meninges is ADA, which is involved in purine catabolism, exists in at least three forms. ADA is a monomeric protein with a molecular mass of approximately 35 kDa and two ADA molecules joined via a connecting protein form the dimeric connective tissue. The between batch coefficient of variation of the enzyme was >2 U/l. ADA2 isoenzyme analysis is allowing for the measurement of ADA activity in the cerebrospinal fluid of the diagnosis of tuberculous meningitis. Patient characteristics and inves-
tigator's study diagnosed in patients with acute onset of symptoms, pyrexia, a CSF neutrophilia with high protein, and low CSF glucose. Acute bacterial meningitis is diagnosed if there was a predominately lymphocytic pleocytosis, a normal or moderately reduced CSF glucose (≥0.8 g/l), and negative serology and bacterial, fungal, and mycobac-
terial cultures. Analysis of CSF was considered normal when there were <5 leucocytes/mm³, protein <0.45 g/l, and negative culture and serology. Matched CSF and serum samples were frozen within 6 hours of collection, stored at −20°C (the enzymes are stable for at least 4 weeks) and analysed within 1 week of collection. Tests were performed with laboratory staff unaware of the diagnosis. Total ADA activity was measured by an enzymatic spec-

trophotometric method on a Cobas Mira auto-

analysers (Roche Diagnostics, Switzerland). Erythro-9-(2-hydroxy-3-nonyl)-adenine, a selec-
tive ADA, and ADA, isomerised to the reaction mixture at a concentration of 200 µM, allowing for the measurement of ADA activity (in the same enzymatic system). The between batch coefficient of the test at a dilution of 6 U/l was 8%. The propor-
tion of total ADA which comprised ADA, could only be reliably estimated when total ADA was >2 U/l. ADA isoenzyme analysis is therefore only reported in the groups with a mean total CSF ADA of >2 U/l.

Analysis of adenosine deaminase

isoenzyme-2 (ADA) in cerebrospinal

fluid in the diagnosis of tuberculous meningitis

The outcome of tuberculous meningitis is influenced by the stage of the disease at the start of treatment. Initiation of antituberculous therapy is often delayed because of the inad-

equacy of presently available laboratory tests. Management of the course of the illness is possible if the diagnosis is made early. The three isozymes of ADA are not similarly distributed in infected brain tissue. The only enzyme present in the meninges is ADA, which is involved in purine catabolism, exists in at least three forms. ADA is a monomeric protein with a molecular mass of approximately 35 kDa and two ADA molecules joined via a connecting protein form the dimeric
All results are median and range, except for %ADA 2 (mean and range). TBM = tuberculous meningitis; *Three subjects with CM had %ADA 2 > 80%.

Comparison of total CSF and serum ADA and CSF ADA, in the diagnostic categories is shown in the table. Total CSF ADA was highest in patients with tuberculous meningitis. Using a cut off of 26 U/l, the test was 90.9% sensitive in detecting tuberculous meningitis (10 of 11). The specificity was 94% (47 of 50) in all patients and 77.3% (17 of 22) in those with cryptococcal meningitis or acute bacterial meningitis. There were no significant differences between those with tuberculous meningitis established by culture and probable disease. Similarly, there were no significant differences in the CSF ADA concentrations in HIV positive and negative patients in the acute bacterial meningitis (mean 4.88 U/l vs 3.71 U/l; p=0.49) and normal lumbar puncture groups (mean 0.74 U/l vs 0.12 U/l; p=0.14).

Data were analysed using EpiInfo 6.04 (CDC, Atlanta) and PRISM 2.01 (GraphPad Software, USA). Continuous variables were compared using analysis of variance (ANOVA) and a 5% level of significance was used.

We were able to differentiate patients with tuberculous meningitis from those with aseptic meningitis or a normal lumbar puncture on the basis of the total CSF ADA. However, there was overlap between patients with tuberculous meningitis and those with cryptococcal meningitis or acute bacterial meningitis. A proportion of ADA, isoenzyme of > 80% seems to be a reliable marker of tuberculous meningitis, yielding a sensitivity of 100% and specificity of 86.4%. The only other diagnostic category with patients who had > 80% ADA 2 was cryptococcal meningitis, which is easily diagnosed on Indian ink staining and serology. Serum ADA concentrations were not useful in differentiating the cause of meningitis.

The laboratory technique for measuring ADA is inexpensive (about £1 per test), relatively simple to perform, and can be adapted to an autoanalyzer. It may thus be used in laboratories with limited resources. Measurement of ADA, produces results rapidly, thus potentially decreasing delays before therapy for tuberculous meningitis is initiated. These results seem promising and may make a valuable contribution to the early and accurate diagnosis of tuberculous meningitis. The use of CSF ADA, in the diagnosis of tuberculous meningitis should be further evaluated in larger series, including patients with other lymphocytic meningitides and different settings.

We thank the study partners; also Sally Ann Martin, Mark Ferreira, Renee Wilson, and laboratory staff at Gold Fields West Hospital; Janice Paiker and staff of the department of Chemical Pathology, South African Institute for Medical Research (SAIMR). PS was funded by the Medical Research Council, Department of Health and Gold Fields of South Africa. ES is supported by the Special Trustees of St Thomas’ Hospital.

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Unilateral focal lesions in the rostralateral medulla influence chemosensitiveness and breathing measured during wakefulness, sleep, and exercise

We have read with interest the article entitled Unilateral focal lesions in the rostralateral medulla influence chemosensitiveness and breathing measured during wakefulness, sleep and exercise by Morrell et al, which shows that unilateral ischaemic lesions of the rostralateral medulla may lead to an abnormal ventilatory CO2 response and sleep apnoea. We have recently conducted a similar study on five patients with syringobulbia. Syringobulbia has a predilection for autonomic nuclei of the cardiorespiratory network localised in the caudal medulla and this may cause severe respiratory and cardiovascular abnormalities. On occasion, a syrinx may extend to the rostral and ventral medulla.

Out of five patients with syringobulbia studied with MRI, ventilatory CO2 response and polysomnography, one patient had bilateral syringomyelic cavities in the caudal dorsal medulla with unilateral extension to the rostralateral medulla (figure). This 40 year old patient showed the following respiratory abnormalities: end tidal CO2, 47.3 mm Hg; p 0.1, 0.21; ventilatory CO2 response, 1.78 l/mm Hg; apnoea index, 52 events/hour of sleep, with a total number of 212 obstructive sleep apnoeas, four central apnoeas, and 39 hypopnoeas. Maximal duration of obstructive sleep apnoeas was 125 seconds and oxygen saturation values during apnoeic episodes were less than 50%. There was also evidence of severe autonomic dysfunctions with orthostatic hypotension, arterial hypotension at rest, and complete loss of sinus arrhythmia. Despite the severity of the respiratory abnormalities recorded, the patient refused to receive any respiratory support, and to date has not developed any cardiorespiratory complication during a follow up of 9 years.

It seems that extension of the syrinx to ventral and rostral medullary areas may lead to more severe respiratory and cardiovascular abnormalities. Three stages in the progression of syringobulbia may be described in involvement of autonomic and respiratory structures: (1) initial involvement of the
caudal and dorsolateral medulla with damage to the nucleus tractus solitarius, vagal motor nucleus, and nucleus ambiguus which may impair cardiovascular reflexes; (2) ventral extension with involvement of the intermediolateral reticular formation; and (3) further ventral and rostral extension to the anterolateral surface of the rostral medulla, involving vasomotor neurons and central chemoreceptors. These last two stages may be accompanied by severe respiratory abnormalities and arterial hypotension.

We have encountered similar difficulties to those described by Morrell et al in outlining small lesions in the medulla by using MRI. Unless the cervicomедullary junction is studied with thin slices, small cavities may be easily overlooked. In such patients, chest and abdominal movements due to respiratory abnormalities may be more extensive than in our study. Nevertheless, both papers highlight the gross sleep apnoea in these patients and the importance of carrying out nocturnal polysomnography to identify any abnormalities in breathing during sleep; this point is emphasised in an excellent editorial by Malow.

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The authors reply:
We were interested to read the paper by Nogues et al, and we are grateful to the authors for bringing it to our attention. One of the aims of our study was to investigate the effect of unilateral lesions in humans at sites previously defined as being important for chemosensitivity in animals. For this reason we studied patients with relativity small focal lesions in the rostrolateral medulla. Nogues et al have investigated the effect of syringomyelia and syringobulbia on chemosensitivity and breathing during sleep; the MR images of their patients show the lesions to

BOOK REVIEWS


One is naturally a little wary when asked to review a book entitled Neurologic Catastrophes in the Emergency Department. Why me? springs to mind. This said, swallowing my pride, I thoroughly enjoyed this book. Although the content is medical, parts on: ischaemic stroke, haemorrhage, cord compression, infections etc, the presentation is special. It is an extremely accessible book, clearly laid out with key points of management and pharmacology appearing in shaded boxes (called capsules). The book is generously illustrated with informative diagrams (and I refer the reader, in particular, to that explaining assessment of the Glasgow coma scale) depicting procedures and anatomy. There are bountiful radiological images and a few select colour plates of clinical cases.

The text is a monograph but Dr Wijdick’s opinions and practices are clearly stated as such and on the whole the evidence, or lack of it, for a management decision is described and referenced. Although American, the principles are transatlantic and this should in no way deter the English reader.

The content covers that dealt with by the neurologist, neurosurgeon, and casualty officer. It should be compulsory reading for those on the front line receiving such emergencies and may then play a part in preventing an emergency becoming a catastrophe. On the other hand, while providing the experienced neurologist with an enjoyable read, hopefully most are already fully acquainted with the up to date and appropriate management of a neurological emergency!

GILLIAN HALL

Handbook of Clinical Neurology; Systemic Diseases, Part III. By MI AMINOFF and CG GOETZ (Handbook of Clinical Neurology series edited by PJ Vinken and GW Bruyn). (Pp748, US$264.50). Published by Elsevier Science, Amsterdam, 1998. ISBN 0444812903. This is volume 71 in the series of the Handbook of Clinical Neurology. It is part III, the third and last volume dedicated to the neurology of systemic disease and, therefore, updates the previous volumes on this subject.

CORRECTION

Davies NP, Eunson LH, Gregory RP, et al. Clinical, electrophysiological, and molecular genetic studies in a new family with paramyotonia congenita. J Neurol Neurosurg Psychiatry 2000; 68:504–507. During the editorial process, figure 1 was reproduced incorrectly. The correct figure is shown here:

There is no shortage of books on multiple sclerosis and interestingly it has to be said what justifies each new publication. With this offering, the Chief Medical Officer and the Vice President of the Clinical Programs Department of the United States National Multiple Sclerosis Society enter the fray. When on home territory (such as the National Multiple Sclerosis Society studies on the emotional effects of multiple sclerosis on patients and families) the material is original and helpful. Also to those working in all the disciplines mentioned above. By necessity, however, it gets a little heavy going from time to time and although illustrated with radiological images and histology, the text is hardly succinct enough to offer some relief. I was left wondering why these two particular plates (one a midbrain perivascular haemorrhage and the other of a vasculitis with fibrinoid necrosis) did to deserve such special attention.

GILLIAN HALL


This is the best handbook on sleep disorders in neurology that I know. It gives as well as comprehensive information about the many and common sleep problems that accompany neurological illness. The text is well organised with chapters on sleep symptoms such as and sleep disorders in conditions such as multiple sclerosis and migraine. The forward spells it out – “the British never seem to get bored of discussing about the weather, whilst Americans are equally prepared to discuss about their own sleep patterns”. This is fair comment on the cottage industry state of United Kingdom sleep medicine versus the GB (Giga-buck) values of the United States. Indeed, we owe much of what we know about sleep to the American pioneers, who are well represented in this book.

It may reflect the parsimonious state of British sleep medicine to complain that there is too much of the neuroonomologist who will do a “sophisticated neuroanatomical assessment of the type best coordinated by a clinical neuroscientist” in this book. Surely there is still the foremost need to make a definite diagnosis by clinical history rather than sleep laboratory. I still think that a good psychiatrist is of greater value in sleep medicine than an EEG machine. It is a pity to discuss psychological factors is sadly missed. Still, the science presented is state of the art, although surprisingly there is little emphasis on molecular biology, which is an expanding field of sleep research in 2000. HLA systems are mentioned, but not clock genes or the many gene determined neurological disorders such as the Prader-Willis syndrome, which have important implications for sleep medicine. The book unfortunately was written before the important discovery of the hypothalamic orexin system involvement in sleep and atonia.

There are a few minor criticisms. The index is useful (no modafinil, no mela-
din). The references are very comprehen-
sive but not selective (a near universal book fault); the illustrations, which are mainly EEGs and actimetry recordings, are adequate but boring. Meaningless sentences are no more common than in most medical writing, but we could do without “the diagnostic cri-
teria for multiple sclerosis served to identify patients with this condition”, as well as “sleep is an active and integral component of the central nervous system (CNS) and may be one of its most important functions”. Treatment information can be inadequate. There are four lines on modafinil in a book of 422 pages. Physicians who want to know how anti-epileptic drugs impinge on sleep are referred to “the many excellent texts avail-
able”. What about sleep problems with new dopamine agonists, or after stereotactic surgery? Does melatonin deficiency insomnia in elderly people really exist? How does the idea that “chronopharmacological considerations will prove to be of importance in TBI” turn into an exact timetable for giving drugs after head injury?

Despite these problems it is most useful to have authoritative writing about conditions as diverse as familial insomnia and narcolepsy collected into one volume. Special mention must be made of an excellent chapter on insomnia by Lugaresi and his col-
leagues, discussion about dissociated states of brain and mind by Mahowald and Schenck, as well as Chadanar’s critical review of the role of melatonin in sleep disorders. There is a diverse outstanding list of contributing editors: W. Thorpy, and the editor, Antonio Culebras, gives a good description of the biology and neuroanatomy of sleep as well as outstanding chapters on neuromotor disorders, head-
aches and multiple sclerosis.

Sleep disorders are important in all areas of neurology. Unless you already have one of the many comprehensive texts on sleep medicine, money spent on this book is well worth it. Your practice will improve.

DAVID PARKES


The applications of transcranial magnetic stimulation (TMS) have broadened significantly in the past few years and the number of publications involving this technique have increased dramatically. This book is very well timed, therefore, and the editors are all well known in this field. It is a book of about 360 pages and is published as a supplement to the journal Clinical Neuro-
physiology. It is divided into five sections, covering methodology, physiology, clinical neuro-
physiology, psychiatry and cognition. Most of the authors have written extensively on TMS, so the chapters represent a fairly good distil-
ation of relevant details into brief texts, each accompanied by a set of references, which will lead new readers to pursue their own special interests and allow those who have been involved in TMS for some years to catch up with recent developments. The latter applies in particular to the community of neurophysiologists who have been using TMS in clinical and non-clinical studies of the motor system, who are now interested to see this technique being applied outside this system.

The various sections include modelling the stimulating field, its haemodynamic effect and role in mapping, the acquisition of single cell responses in M1, and the recent use in animal and human studies. The paired pulse and rapid rate paradigms are discussed, with clinical, physiological, and pharmacological applications and in the clinical section there is a series of papers describing several.

Thomas Brandt's masterpiece Vertigo remains mandatory reading for clinicians interested in this symptom. The basic sciences underpinning vertigo, together with the clinical diagnosis and management of all disorders characterised by this symptom, are comprehensively reported, extensively referenced, and beautifully illustrated. The book is detailed and exhaustive in its subject matter. The only area which seems to have been omitted in any depth is that of the cardiovascular causes of dizziness and vertigo.

There is a broad introduction covering the pathophysiology underpinning vestibular disorders, vertigo, dizziness, and falls; the clinical assessment of such a patient; and the management strategies, together with their rationale. Common disorders—namely, acute peripheral vestibular episodes and Menière's disease, are well covered, in addition to the rarer disorders such as the Ramsay-Hunt syndrome, bilateral vestibular failure, and autoimmune disorders associated with vestibular dysfunction. In this section, the author's views on certain disorders are reflected particularly in the chapters on vestibular neuritis and peripheral vestibular paroxysmia, and it is regrettable that a little more of the controversy in the literature is not highlighted. The management section of the Menière's chapter shows little in depth discussion of trials of medication which, with one or two exceptions, have been extremely poorly designed. Moreover, the incidence of hearing loss in association with gentamicin installation is not mentioned, although surgical treatments in general are well discussed. The chapter on perilymph fistulas is somewhat didactic in the light of many surgeons' views that the validity of this entity must be questioned.

Central vestibular disorders are clearly explained with an excellent and easily understood introduction outlining vestibular disorders in the different planes of action of the vestibulo-ocular reflex. Illustrations and diagrams make some difficult concepts accessible. There are two particularly valuable chapters on the vestibular cortex and its disorders and vestibular epilepsy, both areas that to date have been poorly investigated and understood, but which represent areas ripe for research with new imaging techniques and perceptual tests. Brandt then moves on to postural and standing vertigo which is possibly the "topic of the decade" for those with a vestibular interest, as the introduction of particle repositioning manoeuvres has provided a "cure" for a very common vestibular syndrome although, intriguingly, the underlying science of exactly what we are doing with this technique is somewhat didactic in the light of many surgeons' views are revealed, but, that aside, I cannot recommend this book too highly to guide both the experienced and the uninitiated through the minefield of pitfalls that beset the clinician trying to sort out vertigo. The limiting factor will be the price but certainly it is a book, and possibly the book, which should be in every library used by neurologists and otologists.

LINDA LUXON


Instant answers—from pocket books to reduced, handy text—this is the theme of our times. Nothing captures the attention better than the success of the Penguin 60s or in medicine the proliferation of summary or concise texts. These books are exceedingly difficult to do well and require a distinct approach to that of writing a conventional textbook. All too often a summary or concise text is a poorly edited version of a larger tome.

A model example of what a concise specialist book should look like in style and content is provided by David Perkin, in Mosby's Color Atlas and Text of Neurology. This book uses an imaginative combination of illustrations, colour coded boxes which highlight "the take home points", and bullet pointed narrative to deliver a very readable book. Part of the success of the book is the focus on common conditions with a sufficient nod to minutiae within each field. An emphasis on common and treatable is evident throughout with appropriate weighting on risk factors and diagnostic criteria where relevant. Tables and colour illustrations are skilfully deployed around the text to assist the reader. Inevitably, in a single author book, idiosyncratic views are revealed, but, that aside, I cannot recommend this book too highly to guide both the experienced and the uninitiated through the minefield of pitfalls that beset the clinician trying to sort out vertigo. The limiting factor will be the price but certainly it is a book, and possibly the book, which should be in every library used by neurologists and otologists.

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Retro-ocular headache with autonomic features resembling "continuous" cluster headache in lateral medullary infarction
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