

# PostScript

## CORRESPONDENCE

### The naming of parts

Many deplore the journalistic trend to label well recognised conditions by acronyms, or by recently invented names—commonly to no useful purpose. Thus neurologists may not welcome yet another two names, recorded in past literature but not in general currency.

Umapathi *et al* give a most valuable and timely review of the dropped head and bent spine syndromes.<sup>1</sup> But why perpetuate the ungainly phrase *head ptosis*? *Ptosis*, Greek πτωσις = falling, has traditionally been applied only to the upper eyelid and to prolapse of any of the viscera or of the breasts. *Head drop* is short and its meaning is unequivocal.

*Camptocormia*, Greek κάμπτω = bent; κορμός = trunk of a tree: but, like a few others,<sup>2</sup> Umapathi *et al* apply it to signify a bent spine. *Camptocormia* was first an illness occurring among soldiers in World Wars I and II, and was regarded as a sign of hysteria. *Ankylosing spondylitis* is a more frequent cause. It is an ostentatious word that portends something more mysterious than a bent back or neck. To be similarly pretentious, can we not survive without such euphuisms?

J M S Pearce

Anlaby, East Yorks, UK; jmsp@freenet.co.uk

### References

- 1 Umapathi T, Choudhry V, Cornblath D, *et al*. Head drop and camptocormia. *J Neurol Neurosurg Psychiatry* 2002;73:1-8.
- 2 Sinel M, Eisenberg MS. Two unusual gait disturbances: astasia, abasia and camptocormia. *Arch Phys Med Rehabil* 1990;71:1078-80.

### Head drop and camptocormia

The article by Umapathi *et al*<sup>1</sup> in this journal referred to the original use of the term *camptocormia* by Souques in 1915,<sup>2</sup> though functional bent back was first described by Brodie in 1837. Mlle Rosanoff-Saloff supported Souques' case study with a photographic record of this soldier's bent back and his recovery. According to the English translation abstract in Southard's fine collection of shell shock cases<sup>3</sup> this soldier was wounded five months previously by a bullet that entered along the auxiliary border of the scapula and emerged near the spine. "He spat blood for several days ... and when he got up his trunk and thighs were found to be in a state of moderate flexion upon the pelvis, the trunk being bent almost at a right angle." He was able to bend his trunk still further forward than 'its habitual contracted position' and it was evident that there was contraction of the muscles of the abdominal wall and of the iliopsoas. "No motor, sensory, reflex, trophic, vasomotor, electrical, visceral or X-ray disorders could be found." The application of plaster corsets 'cured' this man's deformity within six weeks.

The poilus spoke of this condition as *cintrage* (arching), suggesting that it was not an uncommon affliction of the French soldier. Seemingly only recorded by French

neurologists, Roussy and Lhermitte reported two subsequent cases.<sup>3</sup> An infantryman was thrown into the air by the bursting of a shell, rendered unconscious and recovered experiencing violent pains in the back. He remained stooped to the right. His bent back was corrected by the application of plaster corsets. The other reported case was that of a chasseur who was buried in an explosion, knocked unconscious, and experienced acute respiratory distress, and subsequent mutism and *camptocormia*. One séance of electrical treatment corrected the improper attitude of the trunk, though he did continue to experience "a few persistent lumbar pains".

It would be difficult to doubt the probability that psychological factors influenced these men's recuperation. To describe these soldiers as hysterical,<sup>4</sup> though this was the terminology used during this period, or indeed that they suffered functional bent back, is probably unfair. They may well have suffered acute traumatic spinal injury and reactive muscle spasm (and contractures). Persistent stooping in shallow trenches, in appalling conditions of deprivation and danger, may have been contributing factors weakening the tone of paraspinal muscles. However, these case reports suggest that the traumatic injury alone may be sufficient explanation for the bent spines. The management of *camptocormia* in the first world war was to provide biomechanical supports, such as corsets, apparently with good results. The psychological therapies of "persuasive re-education" were additive rather than pivotal, and faradisation (and other tortures) used only "if necessary".<sup>3</sup>

The Sandler triad of low self esteem with confusion of identity, sadomasochistic behaviour toward military authorities, and impotence<sup>4</sup> were, in 1947, proposed as being an essential part of *camptocormia*. Umapathi's recognised causes of *camptocormia* and the contributing factors however implicate organicity, as indeed do the original case reports.

A D Macleod

Psychiatric Consultation Service, Christchurch Hospital, Private Bag 4710, Christchurch, New Zealand; PCS@cdhb.govt.nz

### References

- 1 Umapathi T, Choudhry V, Cornblath D, *et al*. Head drop and camptocormia. *J Neurol Neurosurg Psychiatry* 2002;73:1-7.
- 2 Souques A, Rosanoff-Saloff M. La camptocormie. *Rev Neurol (Paris)* 1914;22:937-9.
- 3 Southard EE. *Shell-shock and other neuropsychiatric problems presented in 589 case histories from the war literature, 1914-1918*. Boston: WM Leonard, 1919.
- 4 Sandler SA. *Camptocormia, or the functional bent back*. *Psychosom Med* 1947;9:197-204.

### Author's reply

We would like to thank Dr J M S Pearce for his comments.

We agree with him on the proliferation of medical terms referring to similar if not identical conditions. One of the chief aims of writing this paper is to thread a line of commonality through the various names in literature,

which in essence refer to an anterior curvature of the spine. Hence the title "Head drop and *camptocormia*, the spectrum of bent-spine disorders".

However, we would like to disagree with Dr Pearce labelling the spinal deformity seen in *ankylosing spondylitis* as *camptocormia*. In arthritic conditions and diseases that affect bone, the spinal deformity is fixed. In the bent-spine disorders referred to in the paper, the deformity may reduce considerably or even disappear with change in position, for example when supine. We would therefore prefer to reserve the phrases *head drop* (used interchangeably with *head ptosis*) and *camptocormia* to neurological conditions that affect the strength or tone of the muscles controlling spinal posture.

As aficionados of medical history, we very much enjoy Dr A D Macleod's letter. We agree that organic factors might have contributed to the *camptocormia* in soldiers believed to have been suffering from hysteria. It would have not been unexpected for patients, like the man described by Southard with a bullet wound near the spine,<sup>1</sup> to have developed spasm or even denervation of thoracic paraspinal muscles.

T Umapathi

Department of Neurology, National Neuroscience Institute, 11 Jalan Tan Tock Seng, 308433 Singapore; tumapathi@yahoo.com

### Reference

- 1 Southard EE. *Shell shock and other neuropsychiatric problems presented in 589 case histories from the war literature, 1914-1918*. Boston: WM Leonard, 1919.

### Infection and multiple sclerosis

The article by Hawkes<sup>1</sup> and the editorial commentary about the role of infectious agents in multiple sclerosis (MS) examined this question from a new viewpoint based on epidemiological observations.<sup>2</sup> Several infectious agents, most not sexually transmitted, were reported to be associated with MS according to epidemiological data, serology in CSF and blood, or demonstration of pathogens in tissue. A relation with measles virus (MV) has been an early and most consistent finding. More recently, higher prevalence and higher titres of antibodies against human herpesvirus 6 (HHV6), but not other herpesviruses, were shown in MS patients compared to control groups, suggesting different exposure to HHV6 in MS.<sup>3</sup> HHV6, like vaccine strain MV and certain wild type MV, uses the membrane cofactor protein (MCP; CD46) as a receptor for entry into cells. This suggests a possible involvement of CD46 in MS.

The possibility of a particular isoform of CD46 predisposing MS patients to infection is unlikely because all isoforms have similar affinity to MV. Increased levels of soluble CD46 have been reported in the serum and cerebrospinal fluid of MS patients, more in those who have HHV6 DNA.<sup>4</sup> One interpretation of these findings involved increased activity of the complement system in MS. However, experimental studies show no influence of inflammatory cytokines on CD46 expression and do not support inflammation

as a cause of increased CD46.<sup>5</sup> Incorporation of CD46 in the viral envelope, or a possible genetic propensity in MS patients, have also been considered as causes of increased CD46.<sup>4</sup> While its origin in MS is unclear, soluble CD46 might be involved in viral pathogenesis by binding the virus in the viraemic phase and allowing another to attach to CD46 and spread from cell to cell. Both HHV6 and MV are infectious agents encountered in early childhood, and HHV6 can indeed become reactivated a few weeks after primary MV infection. On the other hand, because HHV6 and MV downregulate CD46 expression on the infected cell, they may diminish the entry of each other, delaying the time of infection. Therefore, they might produce increased antibody levels in young adults through delayed infection with, or reactivation of, each other. These suggest increased antibodies against these two viruses in MS may be interrelated.

The question remains whether a cause-effect relation exists between infectious organisms and MS, or whether viruses are just a consequence of the activation of the inflammatory-immune sequence or increased susceptibility of MS patients to infection. Studies of CD46 and other viral receptors seem warranted in MS.

**B Anlar**

Department of Pediatric Neurology, Hacettepe University, Ankara 06100, Turkey; banlar@hacettepe.edu.tr

#### References

- 1 **Hawkes CH.** Is multiple sclerosis a sexually transmitted infection? *J Neurol Neurosurg Psychiatry* 2002;**73**:439–43.
- 2 **Stewart G.** Infection and multiple sclerosis—a new hypothesis? *J Neurol Neurosurg Psychiatry* 2002;**73**:358–9.
- 3 **Enbom M.** Human herpesvirus 6 in the pathogenesis of multiple sclerosis. *APMIS* 2001;**109**:401–11.
- 4 **Soldan SS, Fogdell-Hahn A, Brennan MB, et al.** Elevated serum and cerebrospinal fluid levels of soluble human herpesvirus type 6 cellular receptor, membrane cofactor protein, in patients with multiple sclerosis. *Ann Neurol* 2001;**50**:486–93.
- 5 **Mason JC, Yarwood H, Sugars K, et al.** Induction of decay-accelerating factor by cytokines or the membrane-attack complex protects vascular endothelial cells against complement deposition. *Blood* 1999;**94**:1673–32.

### Infection and multiple sclerosis

The paper by C H Hawkes (Is multiple sclerosis a sexually transmitted infection?) has caused predictable distress to people with multiple sclerosis (MS) and their families. Living with MS is a difficult enough experience without such sudden and avoidable alarm. The UK Multiple Sclerosis Society's national helpline and local branches have been inundated with calls expressing anger and anxiety.

It is hard to understand the motive for publication when your own expert editorial commentator specifically referred to the paper's "pure speculation" and "potential to cause harm". Did the sensational nature of Dr Hawkes's hypothesis and the virtual guarantee of extensive publicity it could receive outweigh proper consideration of its scientific merit?

There is also the worrying question of what damage may have been caused to the reputation of MS research in the UK by the lay media coverage which was attracted. The MS

Society has a current forward commitment of around £12 million to nearly 70 research projects. That money is raised by voluntary donation. Anything which could discredit the quality of research here is of material concern to us.

**M O'Donovan**

Chief Executive, The Multiple Sclerosis Society, M S National Centre, 372 Edgware Road, London NW2 6ND, UK; modonovan@mssociety.org.uk

#### Reference

- 1 **Hawkes CH.** Is multiple sclerosis a sexually transmitted infection? *J Neurol Neurosurg Psychiatry* 2002;**73**:439–43.

Ed: The journal regrets any distress caused to patients with MS as a result of the widespread publicity this article received in the media. However, we wish to emphasise that the article was subject to the usual peer review process.

## BOOK REVIEWS

### Delirium in old age

Edited by J Lindesay, K Rockwood, A Macdonald (Pp 238, £55.00). Published by Oxford University Press, Oxford, 2002. ISBN 0-19-263275-2

Delirium is an extremely important condition for a number of reasons. It is very distressing and frightening for those who experience the symptoms, and descriptions of the effects on the brain as a result of high fever have been well described. There is a high mortality associated with the development of delirium, and it is often associated with behavioural disturbances that can be troublesome for carers and attendants. Finally, it presents a unique opportunity to look at the interface between psychiatric symptoms caused by organic disease and functional disorders.

Twelve years ago, the same publishers and two of the current editors produced the first edition on delirium. It was a relatively thin book but set the standards that the current edition continue. Delirium is certainly a niche market, and there appear to be no direct competitors, although textbooks on old age psychiatry usually contain chapters and notes on delirium. The new edition is greatly expanded and very much up to date.

Every aspect of delirium is included, from the history and conceptual basis of the disorder through epidemiology, neurophysiology, clinical assessment, management, prevention, and, refreshingly, the role of family caregivers and nurses in managing the disease. The core tenet of the book is that delirium is a disorder that is relatively poorly recognised (particularly the hypo-alert type) by the general clinical professions, it is relatively easy to identify people at risk of developing delirium, and that there is a real possibility of a reasonable preventive strategy for the disorder. Twelve authors have contributed and, as delirium is relatively under-researched, this probably represents a significant proportion of the leading researchers in the field internationally. There are particularly interesting sections on the conceptual basis of the disorder and how it, and its component symptoms, are defined, methods of assessment of delirium are covered comprehensively, a

summary of how evidence based management plans can be developed, and the prospects of prevention of delirium are given an adequate airing.

An interesting spin, which I discovered by accident, is that on the Oxford University Press website ([www.oup.co.uk](http://www.oup.co.uk)), one can see online updates of each individual chapter. Those present when this author last visited the website (December 2002) consisted of work that had been done from when the manuscript had been submitted to publication. It may be that reviews of the book might also appear online—this one will.

The book is a landmark in the literature on delirium, is a text of very high quality, and anyone seriously involved in the clinical management of patients with delirium or research on the subject would do very well to read this book.

**A Burns**

### Neurophysiology in neurosurgery. A modern intraoperative approach

Edited by Vedran Deletis and Jay L Shils (Pp 469, \$125.00). Published by Academic Press, California, 2002. ISBN 0-12-209036-5

This book comprises 17 chapters contributed by 24 authors. It has clearly benefited from most of the chapters being written in a more or less homogenous style and formed into seven parts based mainly on surgical procedures: motor evoked potentials/neurophysiological base; intraoperative neurophysiology (ION) of the spinal (spinal cord monitoring); ION of peripheral nerves, nerve roots and plexuses; ION of cranial nerve and brainstem; ION of supratentorial procedures; ION during stereotactic neurosurgery for movement disorders; and ION and anaesthesia management. Most of the chapters cover the background of methodology, description of the surgical procedure, and the related neurophysiological procedure, personal experience, and case reports, which gives a balanced theoretical and practical view on the topic of each chapter. The interdisciplinary approach taken in this book will ensure it has a wide range of readers across "neurosurgery, neurology, orthopaedic surgery, neurophysiology, anaesthesiology, interventional radiology, and biomedical engineering".

Chronic deep brain stimulation or neuromodulation has extended the role of clinical neurophysiology beyond its traditional diagnostic role. This new field is touched upon briefly in the part on ION during stereotactic neurosurgery. An interesting feature of this book is that it is accompanied by a CD that certainly enhances its value. Cross references are given at the end of the corresponding chapter rather than in the list of contents in the book, and at the front page of the display.

In conclusion, it is an authoritative review of intraoperative neurophysiology much weighted on the motor system for a wide range of surgical procedures. Perhaps, in its present form, those hoping for a more systematically informed discussion on intraoperative neurophysiology of the sensory system may feel slightly disappointed.

**X Liu, T Z Aziz**

## Clinical neurophysiology of the vestibular system, 3rd edition

Edited by R.W. Baloh and V. Honrubia (Pp 408, US\$98.50). Published by Oxford University Press, New York, 2001. ISBN 0-19-513982-8.

The first edition of *Clinical neurophysiology of the vestibular system*, published in 1979, had a significance beyond its content: it affirmed that neurology had a stake in the vestibular system. Here was a neurologist (Baloh) writing with an otolaryngologist (Honrubia) about semicircular canals, endolymph, audiograms, and above all the vestibulo-ocular reflex—the “VOR”. The VOR is no ordinary reflex; one can measure accurately both its input and its output and come up with a transfer function for gain—a new concept then for neurology. We have learnt a lot more about measurement of vestibular function and about disorders of the vestibular system since 1979. The 2nd edition, published in 1990, and now the third edition, incorporate these advances.

And what a terrific book it still is: based on concepts, packed with facts, lucidly written, and rigorously referenced. Its structure is logical and its language is clear, so that it is not only easy to search and browse but a pleasure to read from cover to cover. And it is comprehensive—no vestibular stone is left unturned.

There are four main parts, dealing in turn with: the structure and function of the vestibular system (four chapters); the clinical and laboratory evaluation of the dizzy patient (four chapters); specific diseases affecting the vestibular system (10 chapters); and the treatment of vertigo and vestibular loss (two, yes only two, chapters—but then that's neurology for you).

It's impossible to single out any one chapter, they are all outstanding. For example, I particularly liked the new material in chapter one on the phylogeny of the vestibular system. Now one would have to admit that familiarity with the otocyst of the sea anemone is not a lot of use in the consulting room, but this section is so clearly written and matter so interestingly explained that one happily dispenses with such utilitarian demands.

The great strength of the book and what has made it such a classic, is that although it is based on physiology, full comprehension of physiology is not a prerequisite for retrieving useful information from the disease based chapters. Although the structure is there, one can put this aside and simply delve. The chapters on the three most common vestibular diseases, benign positional vertigo, migraine, and Meniere's diseases, are absolute gems. Each could be published as a self-contained review in its own right.

The book is an elegant conceptual and factual account of the vestibular system, its disorders and diseases, rather than a self-help or how I do it manual. Some readers might miss not having, a “frequently asked clinical questions” section, or at least a “frequently encountered clinical pitfalls” section, but then

no one can have it all. Anyone who sees dizzy patients needs one dizzy book on the desk. This is the one I have on mine.

G M Halmagyi

## Role of proteases in the pathophysiology of neurodegenerative diseases

Edited by Abel Lajtha and Maren L Banik (Pp 302, £59.50). Published by Kluwer Academic/Plenum Publishers, New York, 2001. ISBN 0-306-46579-5

This volume would be an extremely useful addition to the bookshelf of anybody with an active interest in the biochemical and pathological processes that underlie some of the more common neurological diseases. In the past the role of proteolysis in these disorders has been largely neglected because it was assumed that it represented a general non-specific metabolic process. In terms of attracting research interest the field also suffered from the confusion in the literature concerning the naming of these enzymes and the fact that the same enzyme might have many different names. However, as the editors point out in their preface, this is no longer the case and they have managed to bring together an impressive array of current research on the involvement of proteases in a wide variety of disorders. From what individually might have been regarded as rather disparate studies, one can now start to see common themes not least of which is the potential therapeutic value of targeting specific proteases and the development of specific inhibitors.

If, like me, you don't have specialist knowledge of this area I would recommend going straight to the last chapter on the mammalian proteinase genes. Here you will find a clearly laid out summary of the classification and characteristics of the four main groups of proteases (serine, cysteine, aspartic, and metalloproteinases). I also found the chapter on the ubiquitin/proteasome system and the normal physiological breakdown of proteins particularly informative. Having read these two chapters you then have a wide choice of disorders and proteases to choose from. Perhaps the most widely discussed is Alzheimer's disease, undoubtedly because of the huge advances that have been made in the understanding of the biochemical processes underlying this disease over the past 15 years. Papain-like cysteine proteases (cathepsins), caspases, calpains, and a novel metallo-endopeptidase (EC 3.4.24.15) all appear to have some role in the pathology of Alzheimer's disease and may, therefore, be potential targets for drug development. There is also a group of Alzheimer's disease specific proteases that affect the processing of the amyloid precursor protein ( $\alpha$ ,  $\beta$ , and  $\gamma$  secretase) and presenilin (presenilinase). Both of these proteins are central to the development of pathology and so these enzymes in particular are key targets for current drug company research.

Apart from the interest in Alzheimer's disease, there are other chapters covering the

role of matrix metalloproteinases and calpain in the demyelination of multiple sclerosis and the key role of calpain in the pathology of traumatic brain and spinal cord injury. Further chapters describe the loss of calcium homeostasis and the subsequent pathological activation of calpain, resulting in the breakdown of key structural proteins in some neuromuscular disorders. In summary, this book has something for everyone in an area of research that holds huge promise for the future in terms of developing useful therapies for treating neurodegenerative disorders.

S Gentleman

## CORRECTIONS

The following abstract was not printed with the article by E L J Hoogervorst, M J Eikelenboom, B M J Uitdehaag, and C H Polman (One year changes in disability in multiple sclerosis: neurological examination compared with patient self report) in the April issue of *JNNP* (*JNNP* 2003;**74**:439–42).

**Objective:** To characterise the relation between one year changes in neurologist rating of neurological exam abnormalities as measured by the EDSS and changes in patient perceived disability as measured by the GNDS in patients with MS.

**Methods:** 250 patients with MS were recruited at an outpatient clinic. Disability at baseline and one year follow up was assessed using the EDSS and GNDS. Correlations between change in EDSS, GNDS-sum score, functional systems, and GNDS subcategories were studied as well as the significance of changes in EDSS associated with changes in perceived disability.

**Results:** The correlation between one year changes in EDSS v GNDS was substantially lower (0.19) than cross-sectional correlations between EDSS and GNDS, either at baseline (0.62) or at follow up (0.77). Notably, changes in functional system scores that are based on neurological examination are poorly or not at all correlated with changes in disability as perceived by the patient. Analysing the impact of a significant worsening in EDSS score we found that this was associated with significant worsening, insignificant change, and significant improvement in the patients' perceived disability in 45%, 39%, and 15% of patients, respectively.

**Conclusion:** Patients' perception of change in disability differs not only quantitatively but also qualitatively from that of an examining physician. There are true differences in change as perceived by the patient and measured by the physician and changes in many dimensions of disability are relevant to the patient and have no measurable impact on the EDSS.

The authors of the short report entitled Paraneoplastic ophthalmoplegia and subacute motor axonal neuropathy associated with anti-GQ1b antibodies in a patient with malignant melanoma, published in the April issue 2003 of *JNNP* (2003;**74**:507–9), were listed in the incorrect order. The author order should read as follows: L Kloos, C W Ang, W Kruijt, G Stoter, and P Sillevius.