The naming of parts

Many deline the journalistic trend to label well recognized 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. Unfortunately, we give a most rational and timely review of the dropped head and bent spine syndromes. Why perpetuate the unmeaning phrase head ptosis? Posis, Greek ποσις = 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 κορµος = trunk, κωρµος = trunk of a tree: but, like a few others, 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 occurrence among soldiers in World Wars I and II and 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 reactive muscle spasm or even denervation of thoracic paraspinal muscles. 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 trauma 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".

The Sandler trial of low self esteem with confusion of identity, sadomasochistic behaviour towards military authorities, and impotence 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.

Head drop and camptocormia

The article by Umapathi et al in this journal referred to the original use of the term camptocormia by Souques in 1915,3 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 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, vasmotor, electrical, visceral or X-ray disorders could be found." The application of plaster corsets 'cured' this man's deformity within six weeks.

The pollus 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, Boussey and Lhermitte recorded two subsequent cases.4 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 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 trauma 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".

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Infection and multiple sclerosis

The article by Hawkes5 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. 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. 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.6 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.

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 obvious factors such as 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", to have developed spasms or even denervation of thoracic paraspinal muscles.

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5 Hawkes CH. The role of infection in multiple sclerosis. J Neurol Neurosurg Psychiatry 2003;74:692–694...


J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.74.5.692-a on 1 May 2003. Downloaded from http://jnnp.bmj.com/ on May 20, 2022 by guest. Protected by copyright.
as a cause of increased CD4. Incorporation of CD46 in the viral envelope, or a possible genetic propensity in MS patients, has also been considered as causes of increased CD4. While its origin in MS is unclear, soluble CD46 might be involved in viral pathogenesis by binding the virus in the systemic 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.

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References

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.

In order to know the motive for publication when your own expert editorial comments specifically refer 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.

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Neurophysiology in neurosurgery. A modern intraoperative approach

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 mainly based on surgical procedures: motor evoked potentials/neurophysiological base; intraoperative neurophysiology (ION) of the spinal (spinal cord monitoring); ION of peripheral nerves, nerve roots and pleuxes; 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 methodological 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. One contribution worth noting is the ION of cerebral hemispheric involvement 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 neuro-modulation 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

PostScript
Clinical neurophysiology of the vestibular system, 3rd edition

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 physiological, morphological, audiograms, and above all the vestibulo-ocular reflex—the “VOR”. The VOR is not an 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: 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); special topics, easily skipping along 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

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 proteinolysis 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 therefore brought 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 proteasome 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 metallo-proteases). 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 (α, β, and γ 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 research company.

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

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 (J Neurol Neurosurg Psychiatry. 2003;74:439–42).

Objective: To characterise the relationship 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 Para-neoplastic ophthalmoagglutination and subacute motor axonal neuropathy associated with anti-GQ1b antibodies in a patient with malignant melanoma, published in the April issue 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 Kruit, G Stoter, and P Sillevis.