The naming of parts

Many deprole 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. Unfortunately, the anatomy gives a most valuable and timely review of the dropped head and bent spine syndromes. Why perpetuate the unangly phrase head ptoths? Tosisis, Greek \( \pi\text{το\(\theta\)ς} \) = 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.

Cintragie (arching), suggesting that it was not some persistent lumbar pain. It would be difficult to doubt the probability that psychological factors influenced these men's recuperation. To describe these soldiers as hysterical, 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 appraising 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".

The Sandler trial of low self esteem with confusion of identity, sadomasochistic behaviour toward 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.

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References


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 and even disappear with change in position, for example when supine. We would therefore prefer to reserve the phrases head drop (used interchangeably with head ptoths) 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 not necessarily relating 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, to have developed span or even denervation of thoracic paraspinal muscles.
as a cause of increased CD4+. Incorporation of CD4+ 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 CD4+ might be involved in viral pathogenesis by binding the virus in the vesicle phase and allowing another to attach to CD4+ 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 CD4+ expression on the infected cell, they may diminish the effect 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 coincidence. It is important to note that HHV6 and HHV7 may be interrelated.

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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Reference

Delirium in old age


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 worry and anxiety. It is hard to understand the motive for publication when your own expert editorial commentary 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 outweight 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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Reference

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


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 consequence 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 disturbance 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 continues. 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), 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 before 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 Miralda 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 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 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 and 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 can regularly see online updates of each individual chapter. Those present when this author last visited the website (October 20, 2003) consisted of work that had been done before 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.
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 physiology, anatomy, endocrinology, neurophysiology, and above all the vestibulo-ocular reflex—the “VOR”. The VOR is no ordinary reflex; one can measure accurately both its input and 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); specific diseases; and the treatment of vertigo and vestibular loss (two, yes only two, chapters—but then that’s neurology for you).

It is 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 the otocyst of the sea anemone is not a role of 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. 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.

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 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 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 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 proteinases (cathepsins), caspases, calpains, and a novel metalloendopeptidase (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 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 JNPP (J Neurol Neurosurg Psychiatry 2003;74:439–42).

Objective: To characterise the relation between one year changes in neurologist rating of neurological examination 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 ophthalmoplegia and subacute motor axonal neuropathy associated with anti-GQ1b antibodies in a patient with malignant melanoma, published in the April issue 2003 of JNPP (2003;74:507–9), were listed in the incorrect order. The author order should be as follows: L Kloos, C W Ang, W Kruit, G Stoter, and P Sillevis.