Familial paroxysmal tremor: an essential tremor variant

We read the letter concerning familial paroxysmal tremor by García-Albea *et al* with considerable interest.¹ The authors describe a 24 year old man with a mild 9–10 Hz postural tremor of the upper limbs, in whom intermittent exacerbations of the tremor occur. The patient's mother (aged 50) developed essential tremor in her 40s, having previously had a similar paroxysmal arm tremor in late adolescence. The patient had four brothers, two of whom (aged 21 and 22) had mild episodic tremor. The authors knew of only three cases of paroxysmal tremor and considered their patient to have an exceptional presentation of essential tremor.^{2,3}

In fact, the concept of familial paroxysmal tremor is far from new. In 1949, Critchley clearly described episodic tremor in his paper on essential (heredofamilial) tremor and cited Flatau (on page 117) for having suggested the term "intermittent tremor" in such cases.^{4,5} Furthermore, Marshall reported in 1962 that in the early stages of essential tremor the amplitude increases "in an episodic fashion, against a background tremor of the same frequency but lower amplitude".⁶

The information that we obtained during our recent study of 20 families with hereditary essential tremor showed that this type of tremor typically begins with a feeling of shakiness "inside" which progresses to an intermittent and then persistent tremor.⁷ Consequently we would consider paroxysmal tremor of the type reported by García-Albea *et al* to be characteristic of the early natural history of hereditary essential tremor rather than an unusual phenomenon, a view which John Marshall clearly held.⁶

P G BAIN
L J FINDLEY
Department of Neurology,
Radcliffe Infirmary, Woodstock Road,
Oxford OX2 6HE, UK

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García-Albea *et al* reply:

We thank Drs Bain and Findley for their interest in our report on "familial paroxysmal tremor".¹ Their recent study of 20 families with hereditary essential tremor showed that this tremor "typically begins with a feeling of shakiness inside which progress to an intermittent and then persistent tremor".² Many of the 101 patients with essential tremor evaluated in our Movement Disorders Unit reported this type of clinical

course (unpublished data). In our opinion, however, the proposed "paroxysmal tremor" has a number of differences from the classic descriptions quoted by Bain *et al*.²

The term "intermittent tremor" suggested by Flatau³ is defined literally in the report by Critchley⁴ as follows: "Rarely there may be no trace of tremor while the patient is at mental or physical repose, but as soon as he comes under emotional tension; if he believes he is being watched; or if he concentrates his attention; or it is uncomfortably cold; or if muscular tonus is raised, tremor appears." In summary, with the term "paroxysmal" we tried to define autolimited episodes of tremor lasting from 10 to 60 minutes, that appeared *once every three to six weeks*, without any apparent precipitating factor, including anxiety.¹

Marshall described a subclinical intermittent tremor that consisted of "short bursts of tremor of higher amplitude interspersed among the general background of physiological tremor".^{5,6} The electromyographical register in our patient, showing a synchronous 9–10 Hz tremor during the episodes, seems to be clearly different from the bursts of tremor showed in the figure of a revised chapter by Marshall.⁵ Out of the episodes, the patient had neither "internal" or "external" tremor or electromyographically registered tremor. In addition, the tremor of the patient's mother *decreased with the passage of time*, although she developed, at age 48, a typical essential tremor. This is not the typical evolution of essential tremor suggested by Bain *et al*.²

Based upon these differences, we think that the proposition expressed in our report is valid.¹

E GARCÍA-ALBEA
F J JIMÉNEZ-JIMÉNEZ
L AYUSO-PERALTA
J TEJEIRO
F CABRERA-VALDIVIA
A VAQUERO
Section of Neurology,
Hospital Universitario "Príncipe de Asturias",
Carretera de Meco S/N, Alcalá de Henares,
E-28805 Madrid, Spain

- García-Albea E, Jiménez-Jiménez FJ, Ayuso-Peralta L, *et al*. Familial paroxysmal tremor: an essential tremor variant. *J Neurol Neurosurg Psychiatry* 1993;56:1329.
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Bilateral carpal tunnel syndrome

The recent letter by Denišlić and Bajec describes a patient with bilateral tarsal tunnel syndrome.¹ A principal point made by the authors is that, to their knowledge, no other case of bilateral tarsal tunnel syndrome has yet been reported. Keck described such a patient and his 1962 paper contains excellent photographs of the same.² Denišlić and Bajec quote this paper but clearly did not look at it.

The rest of this letter adds nothing to

what has already been described. The surgical findings, as in many of these cases, were non-specific. The authors performed epineurectomy of the nerve, a procedure of doubtful value. The authors' brief discussion of causes of posterior tibial nerve damage at the ankle does not do justice to the literature (50 papers at my latest count).³

JOHN D STEWART
Montreal Neurological Institute and
Hospital, 3801 rue University,
Montreal, Quebec, Canada H3A 2B4

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NOTICE

The Summer Meeting of the **British Neuropsychiatric Association** will take place in Manchester, UK on 25–27 September 1994. For further information, please contact Professor M A Ron, Department of Neuropsychiatry, The National Hospital, Queen Square, London WC1N 3BG. Tel: 071-837 3611; fax: 071-829 8720.

BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom and for members of the British Forces Overseas, but overseas customers should add £2 per item for postage and packing. Payment can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.

Neurological Aspects of Substance Abuse. By JOHN C M BRUST. Publisher: Butterworth-Heinemann, Oxford 1993. (Pp 289; £65.00). ISBN 0-7506-9005-4.

Cocaine was detected in hair samples from Chilean mummies three and a half thousand years old, although "abuse" was restricted to the ruling classes. How much and how little has life changed! Professor Brust is to be congratulated for his industry in collecting together in this modest sized book a wealth of information, historical, pharmacological and neurological. The 12 chapters together boast 3715 references, 676 for opioids and 851 references for the chapter on ethanol abuse. Even the effects and side-effects of caffeine are carefully detailed. Delightful quotes preface each chapter, sometimes harshly modern ("If

coke is a lady, crack is a bitch"), sometimes more traditional (De Quincey on opium).

If you need to know about Khat and Betel nut use/abuse it is in this book, as are all thirty street names for phencyclidine. You will learn which eminent doctors in the nineteenth century (other than Sigmund Freud) were addicted to what we now call "hard" drugs and that Osler prescribed marijuana for patients with migraine. It appears that Roget (of the thesaurus) was a chum of Samuel Taylor Coleridge and Sir Humphrey Davy and all three liked sniffing the nitrous oxide that Sir Humphrey synthesised. Many strange and sometimes lethal practices of substance abuse are documented but it is emphasised that ethanol abuse is the world's most important drug problem, "... from a medical standpoint, ethanol is a far more harmful drug than heroin".

Brust's book will be valued by those seeking information on drug abuse, but it could have been improved as a reference source if more care had been taken with its layout. Diagrams to explain drug action would have helped readers of modest receptor literacy to understand drug actions, and the text should have been divided into more sub-headings. This would have made it easier to rediscover, for example, little gems like the description of lead poisoning in (environmentally unreformed) petrol sniffers or exactly which Australian toad is licked for its hallucinogenic secretions.

C M C ALLEN

Neuropsychology of Alzheimer's Disease and Other Dementias. Edited by RANDOLPH W PARKS, RONALD F ZEC, ROBERT S WILSON. (Pp 681; £50.00.) Publishers: OUP, Oxford. 1994. ISBN 0-19-506612 X.

With the increasing longevity of the population, the dementias are becoming more prominent in neurological and psychiatric practice. Although significant advances in understanding the dementias have been made in behavioural neuroscience and neuropsychology, they have not always filtered through to the clinicians managing these disorders. Behavioural neurology attempts to bridge this gap, and thus to improve clinical assessment and management of the dementias. This multi-author North American text achieves this, by providing background theory and clinical application.

The book is well structured: it deals firstly with Alzheimer's disease, focusing on anatomy, neuropsychology and behavioural studies. The chapter on neuropsychology is comprehensive and yet remains accessible to those with no prior knowledge of the subject. There is also a useful chapter on normal ageing. There then follows a lucid account of the other dementias. Although uniformly excellent, the chapters on the vascular, Parkinsonian, MS and HIV related dementias are particularly clear. There is much here to help in clinically differentiating the dementias. Investigation, by neuroimaging and neurophysiology, is next

considered. The use of MRI in diagnosis, e.g. demonstrating hippocampal atrophy in early Alzheimer's, is discussed. The PET chapter links imaging abnormalities with neuropsychological deficit, highlighting the use of functional imaging in neuro-anatomical localisation of neuropsychological function. Electroencephalography and evoked potential studies are also covered. Management of the dementias is dealt with in the final section. Pharmacological attempts at treating Alzheimer's, including manipulating the cholinergic system, are discussed. Psychosocial assessment and management, and legal and ethical issues, are addressed.

Although concentrating on Alzheimer's, this book serves as a good reference source regarding the dementias in general. The neurologist or psychiatrist will also find that it improves clinical assessment and management of the dementias. It achieves its aim of bridging the gap between behavioural neuroscience and clinical practice, and is strongly recommended.

JOHN GREENE

Head Injury/Third Edition. Edited by PAUL R COOPER. (Pp 590, Illustrated; Price: £108.00). 1993. London, Williams and Wilkins Ltd. ISBN 0-683-02108-7.

The third edition of Paul Cooper's book on head injuries maintains the excellent standard of its predecessors and is one of the most readable and authoritative texts on this prevalent and highly important subject. Some of the chapters have a certain familiarity about them. Reading Hume Adams in *Advances in Neuropathology* or in the *Handbook of Neurology* is pretty much the same as it is in Cooper. This is no criticism of Professor Adams, the man has to deal with the subject as best he can and one work will necessarily seem much the same as in another, it is really a criticism of multi-author texts, particularly those which try to take in the whole of such a wide field. It could be avoided by concentrating on the surgical aspects.

Almost all the chapters are of a consistently high quality, although the chapter on cranioplasty reads rather strangely. The disadvantages of ribs are that they absorb, as well as leaving chest wounds. Calvarial grafting makes the whole of the skull thin, iliac crest grafting is ugly and unpleasant for the patient, who has to deal with such a large and unpleasant additional wound. Techniques of preserving bone flaps in the abdominal fat or of sterilising them and reinserting them are not mentioned, nor the technique of preserving them and using them for producing titanium prostheses, which is probably preferable to either of the others. The best current method of cranioplasty is titanium plating, if necessary made on computerised models derived from CT. This doesn't even get a mention. It is to be hoped that the fourth edition updates this aspect of things to an acceptable modern standard.

The index makes sparse reading. Boxing is not mentioned, neither is boxing dement-

tia, natriuretic hormone, electrolyte balance, dizziness, giddiness or vertigo. Sodium balance however, when you can find it, is well covered in one of the best descriptions of this difficult subject, which is at least creditable. Dementia and boxing both get a mention from Professor Adams, although hydrocephalus only gets a sentence.

It might be expected that the Editor at least would proof read his own chapter carefully, if only to encourage the others. He manages however to insult both Bandari and Sarkari by spelling both their names wrongly, which might be thought at least to be impartial. To get the journal wrong as well in the same reference however looks very much like carelessness.

The section of epilepsy might be approached in the hope that there would be some more up to date and more easily comprehended data than that from Jennet, but on looking it up one of Jennet's familiarly incomprehensible histograms appears as the principal information on risk. The section on intracranial pressure monitoring is particularly good and lacks the facile optimism often found in papers which enthuse over the benefits of, or even the necessity for, this technique for the management of head injuries.

Overall it is a well produced readable book which has to be recommended for all workers in the field.

BERNARD WILLIAMS

Basic Neurochemistry: Molecular, Cellular and Medical Aspects/5th Edition. Edited by G J SIEGEL, B W AGRANOFF, R W ALBERS AND P B MOLINOFF. (Pp 1,080, Illustrated; Price: \$84.50). 1993. New York, Raven Press. ISBN 0-7817-0104-X.

In reading a multi-author book which covers such a broad spectrum, one tends to focus on one's own area of expertise. One offers mainly constructive comments to some previous colleagues at NIH in hope that the next edition will prove even better than the previous five. The anatomy of the spinal fluid reveals essentially 2/3 lymphocytes and 1/3 macrophages, clearly pointing to an immunological function. Although the daily flow rate can vary in circadian rhythm by up to a factor of 5, there is nevertheless decreased flow with increasing age with concomitant increase in the levels of all proteins. It is well established that the majority of proteins are sieved according to their molecular size, which is readily appreciated in the five-fold difference in the percentage transfer of IgM versus IgA from serum into CSF. Transferrin, (which is not strictly a "peptide"), is synthesised in part by the choroid plexus, as is transthyretin. Locally-produced cytokines have major effects on the neuroimmunological response, as contradistinct from responses in other tissues. In spite of these relatively minor comments, I would nevertheless offer my wholehearted endorsement of this book. It is a real bargain to get over 1,000 pages for just over £50.

E J THOMPSON