Deep brain stimulation for cervical dystonia

I read with interest the recent case report by Chang and colleagues on unilateral deep brain stimulation (DBS) of the globus pallidus internus (GPI) in a patient with delayed-onset posttraumatic cervical dystonia. I congratulate the authors reporting another patient with delayed-onset cervical dystonia treated with unilateral DBS of the GPI. The unique feature in their case is that unilateral stimulation only was used. They report on a 23 year old man who developed cervical dystonia with head turning to the left three years after sustaining a severe closed head injury. Magnetic resonance (MR) studies five days after the injury demonstrated focal lesions of the left pallidum, but also of the right thalamus. Six years later only the left pallidal lesion could be appreciated by MR studies. The authors chose to implant a quadripolar DBS electrode in the left GPI for chronic stimulation. They further report that during chronic stimulation the patient’s cervical dystonia improved, and he could turn his head to the midline easier than preoperatively. The improvement was not assessed by standard rating scales for dystonia, but by daily patient reports and by the authors’ impression based on contemporary imaging studies and also accumulating knowledge on the innervation of neck muscles. Magyar-Lehmann and colleagues, for example, showed that patients with cervical dystonia had higher glucose metabolism bilaterally in the lentiform nucleus in a PET study without significant differences regarding the laterality, the specific pattern, or the severity of cervical dystonia in individual cases. Naumann and colleagues also demonstrated bilateral basal ganglia involvement in cervical dystonia patients by striatal D2-receptor binding studies. In that study, there was no significant difference by intraindividual comparison of contralateral versus ipsilateral striatal epi- pride binding with regard to the direction of head rotation. In a recent transcranial magnetic stimulation study in normal subjects, ipsilateral as well as contralateral sternocleidomastoid responses were evoked by stimulation of an area of cortex near the representation of the trunk. With that regard, however, it is also important to consider that head rotation in patients with cervical dystonia is not only due to contrac- tion of the sternocleidomastoid, but also of the posterior neck muscles. In our series of patients who underwent bilateral pallidal DBS for treatment of cervical dystonia we have repeatedly observed clinical deterioration with dysfunction of stimulation on one side or when the battery on one side was depleted. It is unclear, therefore, whether or not additional benefit would have been achieved with stimulation also of the right GPI in the patient reported by Chang et al.

By the way, in the Discussion the authors cite data on the frequency of posttraumatic movement disorders secondary to severe head injury. I was surprised to see that these data were attributed to the study on post- traumatic hemidystonia by Lee and colleagues. These data, however, were reported in a later study where we investigated the frequency of posttraumatic movement disorders in the survivors of head injury who were randomised into two groups: group 1 with an ipsilateral focal basal ganglia lesion and group 2 with an contralateral lesion. Patients who underwent bilateral pallidal DBS for treatment of cervical dystonia we have repeatedly observed clinical deterioration with dysfunction of stimulation on one side or when the battery on one side was depleted. It is unclear, therefore, whether or not additional benefit would have been achieved with stimulation also of the right GPI in the patient reported by Chang et al.

In conclusion, for the moment I think it is advisable to continue with bilateral DBS in the treatment of cervical dystonia until solid evidence should be obtained that unilateral stimulation is sufficient. It would be most interesting to evaluate the different profiles of bilateral and alternating unilateral stimulation in patients who have bilateral electrodes. Whether such a study is feasible and practical, however, is open to debate.

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References

Failure of regular external ventricular drain exchange to reduce CSF infection

Dr. Wong and colleagues undertook quite a careful prospective randomised trial aiming to determine whether routine changing of external ventricular drainage catheters reduces the risk of CSF infection. Patients were randomised into two groups: group 1 (n = 51) had routine changes of the external ventricular drain at five-day intervals; in group 2 (n = 52) the ventricular drain was not changed. There was no difference with respect to the basic demographic data and the incidence of CSF infection. The authors observed four CSF infections in group 1 (7.8%) and two in group 2 (3.8%). Despite the higher CSF infection rate in group 1, this difference was not statistically significant. Based on their results, the authors concluded that “routinely changing external ventricular drainage catheters at five day intervals did not reduce the risk of CSF infection”.

The topic of ventricular catheters and the risk of CSF infection has been dealt with in numerous reports. The continuing interest in neurosurgeons is largely based on the fact that quite controversial recommendations
have been published regarding the use of external ventricular catheters.

In general, our experience with CSF infections is similar to that of Wong. We investigated which factors increase the incidence of CSF infections in a prospective study including 133 patients who underwent 152 surgical procedures for external CSF drainage. Assessed variables included basic demographic data, with special reference to the duration of surgery, diameter of the catheter used (5 F v 10 F), distance of the subcutaneous tunnel between the burr hole and cutaneous exit point, additional surgical procedures, and duration of CSF drainage.

In our study group we had a CSF infection rate of 4.5% per patient and 3.9% per surgical procedure. Whereas most of the variables assessed showed no statistically significant correlation with the incidence of CSF infection, interestingly we observed a close correlation between the length of the subcutaneous tunnel and the incidence of infection. In 88% of the patients with CSF infections the catheter was tunnelled subcutaneously for less than 5 cm, whereas in only 17% was the catheter tunnelled for more than 5 cm. This observation was associated with the fact that there was a higher incidence of CSF leakage through the cutaneous exit point with shorter tunnels despite correct operative management.

Taking into consideration that in the study by Wong et al., "all the bacteria are common

References


Author’s reply

The interest and comments of Audenino et al are greatly appreciated. In our paper, 48 patients who were suspected of being in NCSE were evaluated prospectively by neurology residents; the diagnosis of NCSE was later confirmed or ruled out on the basis of the patient’s EEG. Remote risk factors for seizures (such as previous stroke, neurosurgery, significant head trauma), impaired mental status, and other CSF abnormalities (sustained eye deviation, nystagmus, hiccups) were found significantly more often in the NCSE group. The combined sensitivity of remote risk factors for seizures and ocular movement abnormalities was 100%; there was no patient in the NCSE group who did not have either of these findings.
Hemicraniectomy for large middle cerebral artery territory infarction: do these patients really benefit from this procedure?

Pranesh et al presented a series of 19 patients undergoing decompressive hemicraniectomy for large middle cerebral artery infarction with clinical and radiological signs of tentorial herniation. Among these, 10 patients (53%) suffered from a dominant hemisphere stroke. Neurological state was assessed according to the National Institutes of Health Stroke Scale (NIHSS) initially and one week after surgery, and functional outcome at three months’ follow up using the Barthel index (BI) and Rankin scale (RS). The mean NIHSS score improved from 20.5 before surgery to 10.5 postoperatively. At last follow up mean BI was significantly better in younger patients (60.7) than in older patients (41.3). The authors conclude that hemicraniectomy may be a useful procedure on patients with large middle cerebral artery infarction.

Recently we undertook a prospective non-randomised study in 26 patients with decompressive hemicraniectomy for right sided middle cerebral artery infarction, analysing functional outcome (NIHSS, BI, RS) at one year of follow up. In contrast to all previous reports, neurological testing was also done, focusing on right hemisphere function (evaluation of visuospatial and visuconstructive abilities, attention, spatial span, and self rated mood). In 18 surviving patients at the one year follow up the functional outcome was good or fair in nine (BI >75, RS 2–3), moderate in six (BI 30–70, RS 4), and poor in three (BI 0–29, RS 5). Thus only nine of 26 patients (35%) were functionally independent and needed no or only minimal assistance for daily life activities. As was shown previously, age was identified as a significant and independent predictive factor on outcome, with better functional results in younger patients. Neuropsychological testing was possible in 14 patients, while four were too disabled to be evaluated. All patients showed profound attention deficits, and visuospatial and visuconstructive deficits was observed in those with less formal education. These disturbances led to a substantial handicap for professional activities.

On the basis of our functional and particularly neuropsychological results in patients with isolated non-dominant middle cerebral artery infarction, we would strongly discourage hemicraniectomy in patients with left sided, dominant hemisphere or multiterritory infarction, as the risk is significantly higher than the risk of dependency, hopelessness, and more severe neuropsychological deficits in such cases. In our opinion decompressive hemicraniectomy should be restricted to younger patients with non-dominant hemisphere infarction. The goal of the procedure is to operate on these patients in an early stage of the disease, before additional infarction had occurred as a result of local mass effect and herniation. Up to now we have operated on 39 patients with middle cerebral artery infarction in our institution, but our experiences do not encourage us to do anything with great enthusiasm.

Pranesh et al stated correctly that this surgical procedure can be undertaken safely, however, the main difficulty is in deciding to not operate on such patients, despite the simplicity of the surgical procedure.

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References

BOOK REVIEWS

Psychiatric genetics and genomics


At all turns we can less and less speak of medicine without qualifying it with the term molecular. Our genetic underpinnings and their consequences have assumed their rightful place as extremely important factors in the pathophysiology of most disease—in fact it seems nearly all disease (arguably) is related to the current molecular makeup. All psychiatric illness is not exempt from such genetic considerations has been clear for some time. What this new volume edited by Peter McGuffin and his colleagues shows, however, is how widely permeating this has become. There are chapters here that range from personality and cognition (an excellent one from Plomin, Happe, and Caspi) through to personality disorders, anxiety, and eating disorders, through to the more mainline genetics of schizophrenia and affective psychoses. In general they are well written and surprisingly up to date. As a source book of references alone this is well worth having and those to very recent publications including 2002 are numerous. The traditional triops of family, twin, and adoption studies is covered for most disorders before moving into linkage association, and, where relevant, other molecular analyses such as cytogenetics. The chapter on dementia naturally moves further into the field of molecular pathology and biology, and covers the transmissible encephalopathies and CJD. Contentious areas are not omitted and the chapter on ethical issues is thoughtful and avoids the tokenism (or complete omission) that was the hallmark of some previous works.

Authors’ reply

The points raised by Sandalcioglu et al are well taken. It was considered justified to undertake decompression even on the dominant side because, if such patients were left with a severe disability, the excellent family support system in India would be available. We do agree that the quality of life is poor after such a decompression. However, the recovery of speech function in our patients has been remarkable, apart from saving their lives which was the patients’ relatives’ wish.

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Are there any drawbacks? There are always some to be found and as usual these may simply reflect bias on the part of the reviewer. The more mental retardation is exceptionally short given its huge clinical importance (mental retardation and epilepsy together are the most common of all neurological conditions) and the recent explosion of interest in the genetic (and epigenetic) phenomena involved. However, this is a relatively small quibble; it is a well produced and worthwhile volume. On the reviewer's copy the edges of many chapters are already very well thumbed and grubby, which is as good a recommendation as any.

W J Muir


Psychiatric and cognitive disorders in Parkinson's disease


This comprehensive account of the common (but frequently overlooked and under treated) emotional and cognitive aspects of Parkinson's disease is thoughtfully organised and well written. The two authors have presented their material in a consistent manner, free from the difficulties (for example, redundancy) often associated with multi-authored texts. Tables and illustrative clinical vignettes are helpful. References are up to date and thorough. In general, the book is well edited (although the two figures demonstrating the cortico-subcortical connections need revision). The text itself is less than 200 pages and is relatively easy to read in its entirety, but each chapter can stand alone. The first few pages briefly highlight the content of and rationale for each of the chapters. The next 50 pages provide a useful background for the non-movement disorder specialist. Chapter two reviews motor features and their treatment. Interestingly, the discussion on surgical approaches is as long as the discussion of pharmacotherapy. This probably reflects the fact that deep brain stimulation is becoming more widely available. The third chapter provides a concise but thorough and clearly presented overview of the differential diagnosis of Parkinson’s disease, with a very relevant discussion of dementia with Lewy bodies. The discussion of Alzheimer disease is thoughtfully organised more appropriate. The fourth chapter (the one chapter devoted solely to cognition) effectively conveys the notion that dementia in Parkinson’s disease may be not be a homogeneous phenomenon. The authors make the interesting point that bradyphrenia may be accounted for solely by depression and/or incipient cognitive decline. Chapter five (Depression in Parkinson's disease) highlights how common depression is in this illness and provides evidence that untreated depression may result in impaired cognition, making a strong case for early recognition and treatment of depression.

Chapter six includes a discussion of anxiety, apathy, and the debatable concept of a distinct premorbid personality type. Chapter seven mainly focuses on dopaminergic drug induced psychosis. Chapter eight deals with the treatment of depression and psychosis. The appendix consists of several Parkinson's disease specific scales but does not include other scales commonly used to evaluate depression and anxiety in Parkinson's disease.

One would not necessarily want to use this book as a reference for specific treatment guidelines and/or dosing of medications. Dosages are not always discussed (for example, for quetiapine) and a few statements are subject to disagreement. In their discussion about unpredictable levodopa responses, the authors appropriately suggest switching from controlled release levodopa to more frequent doses of an immediate release formulation but state that one should keep the same total daily levodopa dosage. Because controlled release tends to have lower bioavailability, many neurologists would replace the total dose of levodopa when switching to immediate release preparations. The figure demonstrating the treatment of psychosis in Parkinson’s disease suggests that one should check blood and urine for infection or metabolic problems, then check a CT scan before proceeding. Except in unusual circumstances, most Parkinson's disease specialists would not embark on such an extensive diagnostic investigation. The suggestion that severe psychosis warrants mandatory admission and that one should consider stopping all anti-Parkinson’s disease medication does not reflect typical practice and could, in fact, be dangerous because of the risk of an NMS like syndrome.

In summary, this well written book will enable readers to have an up to date and well rounded knowledge base regarding the cognitive and psychiatric aspects of Parkinson’s disease and would be quite helpful to all clinicians (including neurologists and non-neurologists) who deal with Parkinson’s disease patients.

I Hegeman Richard

Surgical treatment of Parkinson’s disease and other movement disorders


The editors have assembled a panel of leading experts to produce this book, which is well referenced and its black and white figures nicely produced. The book is predominantly concerned with the role of stereotactic surgery for movement disorders and this subject is examined in depth. The book is divided into four parts. The first section recounts, in three chapters, the rationale for surgical therapy. The circuitry and physiology of the basal ganglia are reviewed along with the historical development of surgery for Parkinson's disease. The second and main part of the book describes the surgical management of Parkinson's disease and other movement disorders, including patient selection and assessment, target selection and localisation, operative techniques, neuropsychological evaluation, and in situ programming of deep brain stimulators. This section also contains separate chapters on thalamotomy, pallidotomy, subthalamic nucleotomy, and deep brain stimulation of the thalamus, globus pallidus, and subthalamic nuclei. Within these chapters there is a rich diversity of opinion, which is one of the great strengths of this book and reflects this rapidly expanding field.

The third section reviews the surgical treatment of focal and generalised dystonia. This is presently a very exciting field and the relevant chapters detail experience with thalamotomy, pallidotomy, and pallidal stimulation as well as the roles of intrathecal baclofen pumps and peripheral denervation procedures for managing dystonic patients.

The final part of the book, labelled Miscellaneous, describes the use of PET for examining the changes in activity in the cerebral circuitry of movement disorder patients undergoing surgery. Finally, there is an account of the role of fetal transplantation and future surgical therapies for the treatment of Parkinson's disease.

This book provides the reader with considerable penetration into the rapidly expanding field of movement disorder surgery. I found it fascinating and informative. It has a place in the hospital or university neuroscience library and I particularly recommend it to neurologists, neuropsychologists, neurosurgeons, and research fellows who wish to have an overview and/or develop their interest in stereotactic surgery for movement disorders.

P Bain

Concise guide to neuropsychiatry and behavioural neurology, 2nd edition


Cognitive neurology is on the up. In Britain, at least, the numbers of trainee neurologists who aim to make this their focus of interest is increasing rapidly—indeed, perhaps not only because of the attraction of the bright, kaleidoscopic lights of functional imaging! No, some neurologists in the making appreciate that perhaps there is a great deal still to be said for the careful assessment of patients with both focal and diffuse brain lesions. Not only does this offer an important insight into normal brain function, but it is critical for the development of therapies for cognitive impairments. So, is this handbook a helpful contribution to the renewed interest in cognitive function? It certainly does have several features to recommend it. It is compact, to the point, and gives references to important papers in the literature. It covers a vast amount of neurology and neuropsychiatry in a breathtaking short format. However, although brevity is often to be admired, there is a danger that some of the points being made are going to be appreciated only by those who already know what you are talking about. This surely should not be the aim of a handbook that is aimed at trainees. Moreover, attempts to make things concise come at the expense of important omissions. In this text, for example, there is a small section on simultanagnosia...
neuroprotection and the need for neuroprotection. These possibilities are not limited to a single line in a table, although progressive supranuclear palsy does get a paragraph. Sometimes conciseness also leads to a blurring of distinctions: motor neglect is said to refer to a lack of motor response in the neglected hemispace and is noted also to affect the contralateral limb, whereas most experts use the term to refer to only a limb specific neglect. Finally, the figures could be improved upon. For example, the one showing key elements of the limbic system might look good in the original book from which it is taken, but it really is not very clear or helpful in the version scanned into this handbook.

These sorts of quibble apart, this is a useful guide that serves as a gateway to fuller descriptions and discussions in the primary or review literature. It is worth dipping into to see whether it suits you. My own preference would be for a slightly larger handbook that covers some of the syndromes, conditions, and treatments in a little more detail.

Husain M

Neural stem cells for brain and spinal cord repair

For scientists, clinicians, patients, and the biotech industry, transplantation of stem cells has become one of the major hopes for repair of what are currently incurable degenerative diseases and trauma to the brain and spinal cord. The 16 contributions contained in Neural stem cells for brain and spinal cord repair deal with these questions. They provide a needed, very handy, and comprehensive review of the current state of knowledge in this rapidly moving area.

The contributions cover the many possible sources of stem cells, embryo, and adult, including brain, neurospheres, neural crest, bone marrow, and already established human stem cell lines such as human neuronal cells. Numerous chapters summarise the current methods for obtaining the various types of stem cells, the signalling pathways involved in their differentiation, and the degree to which it is known that stem cells are able or can be induced by appropriate growth factors to adopt or turn into the differentiated cell types that would be needed for functional replacement in damaged tissues.

A number of contributions deal with practical applications, such as the possibility of glial cell precursors being used for treatment of demyelinating diseases, the use of stem cells to boost a failing host dopaminergic system in Parkinson's disease, and the concept of global replacement by genetically modified cells able to replace enzymes non-functionally because of inherited genetic defects. Particularly interesting are the provocative observations (Magavi and Macklis) that transplanted stem cells are able to "detect" defects at a distance, and migrate through the host brain to repair damaged areas. A number of chapters consider the issue of stem cells for spinal cord injury and the questions of the relative contributions of local cell damage versus axonal disconnection.

Over the past few years stem cells have become something of a Holy Grail. The concept of a stem cell is one that can divide indefinitely, that will, like the genie of the lamp, become whatever the master requires, and that can be transplanted to repair virtually any affliction of the nervous system. It is, therefore, a sobering thought that although the haematopoietic stem cell has been identified and characterised for 40 years, and is readily available, many forms of leukaemia and radiation sickness can still be incurable. If we are still uncertain how to obtain beneficial effects with a tissue such as blood, which has no structural organisation, how much greater are the problems we must expect to encounter in the brain and spinal cord, the most complex tissue known in biology?

There is a gold rush feel to the stem cell area, and many of the claims currently being staked owe as much to hope as to practicality. Many basic questions remain to be solved. What range of cell types does the term stem cell include? How can we direct their development so that they become specific cell types? And, having done so, how can they then be transplanted to the nervous system in such a way that they will integrate themselves, detect deficiencies, and repair them? And to what extent do we have a clear concept of the dangers in using stem cells? But notwithstanding these unsolved issues, the concept that there exist, not only within the embryo, but also within the adult, cells with yet uncharted reparative potential offers real hope for a new way to treat injuries and diseases for which there is currently no cure.

Raisman G

Perspectives in affective disorders, Vol 21
Edited by W. P. Kaschka. Published by Karger, Basel, 2002, pp 204, €134.50. ISBN 3-8055-7439-8

This book is a summary of an international symposium held in September 2001 to celebrate the 25th anniversary of the depression unit at the Weissenau Centre for Psychiatry in collaboration with the University of Ulm. This unit was founded as the first of its kind in Germany for the treatment of affective disorders and was the start of a development that has led to there being over 60 special depression units in that country. The symposium included a survey of past work and a summary of the present position and future prospects in basic research, diagnosis, therapy, and the care of affective disorders.

Perhaps the most interesting part of this book is the first section of three chapters, which details the development of services for depressed patients in Germany. An elective admission to a purpose built and managed unit for the assessment and treatment of severe depression is extremely unusual in the UK, with its "one size fits all" inpatient care strategy. Intuitively depression units are appealing, but it is disappointing that no evidence is presented in this volume to support the statement that they result in a great improvement in outcome, apart from a claim that 90% of patients would recommend the unit to friends or relatives.

The second section covers basic research in affective disorders, with contributions on genetics, functional imaging, and autonomic control in psychopathology. The third section of eight chapters is subtitled "Therapeutic perspectives in affective disorders". There are some genuinely novel and valuable contributions here, including chapters on pharmacology, antidepressants, molecular mechanisms of action of mood stabilisers, and mechanisms and management of weight gain.

This book is likely to have a limited readership outside Germany. The layout and presentation are rather dreary and although it will attract psychiatrists and psychologists with a specialist clinical or research interest in affective disorders, general clinicians are likely to pass it by.

Bench C

Obsessive compulsive disorder: a practical guide

The stated aim of this book is to provide a practical and accessible guide to the diagnosis, assessment, and management of obsessive compulsive disorder (OCD). The 14 chapters include contributions from an international panel of expert clinicians and a final chapter, The patient's perspective, from a psychologist who also has OCD. The first four chapters present an overview of the nosology, epidemiology, psychopathology, and assessment of OCD. A chapter on quality of life is followed by three chapters that summarise the neurobiology of OCD in terms of genetic factors, neuroanatomy, and neurochemistry. The final section of the book provides chapters on pharmacological and psychological treatments for OCD, including treatment resistant cases and children and adolescents.

The strength of this book lies in the detail of the discussion of the subtleties of clinical assessment, pharmacotherapy, and psychotherapy. There are a number of clinical pearls contained in these chapters, which will help clinicians to ensure that their patients receive the most effective and appropriate treatments available. The chapter on integrated treatment approaches highlights the gap in evidence whereby it is still uncertain whether combining drug treatment with exposure therapy is any more effective than drug treatment given alone. Although busy general adult psychiatrists are unlikely to ever use the Yale-Brown Obsessive Compulsive Scale, included as an appendix, its inclusion helps to highlight the need for systematic assessment of target symptoms over the prolonged timescale of response to treatment.

As a stand alone text this has many merits and can be recommended to anyone who is involved in the assessment and treatment of OCD. For those more interested in the neurobiology it provides a stimulating introduction with good references to the more detailed literature.

Bench C