Transition from paediatric to adult neurological services

Drs Tuffrey and Pearce are timely in their comments about the importance of structured transition to adult orientated services for young people with chronic neurological disease.1 We agree that this is a significant problem, and that proper transition (including pretransition planning, planning at the appropriate time and multidisciplinary working) must be achieved to maximise the young person's integration into society. There is abundant evidence that those lost to follow up present later with treatable complications of their conditions.1 It is also known that health status and psychosocial factors have a proved impact on the social participation of young disabled adults.1

There are a number of well established and well researched models of multidisciplinary team care for young adults in transition with disabling conditions including neurological conditions, such as cerebral palsy, spinal banded, and muscular dystrophy. The consultant in rehabilitation medicine is well used to working with multidisciplinary teams, in a manner analogous to the paediatrician. An evaluation of a team approach versus ad hoc health services for young people with physical disabilities, in six areas of the UK, which was published in The Lancet in 2002,2 demonstrated that participation in society was increased in those for no additional financial cost where organised transition services were involved, compared with non-coordinated care. A team has been in place in Leeds for 14 years. It includes professionals including medical, nursing, occupational therapists, physiotherapists, clinical psychologists, and speech and language therapists, and sessions of general practitioner health promotion and family planning. The needs of the population served by the survival of disabled children medical care, and liaison with social services and education authorities is essential. Issues of access to employment and training, occupation of leisure time, relationships, sexuality, and independence are routinely addressed.

Consultants in rehabilitation medicine have the diagnostic and management skills necessary to coordinate the health care of these individuals.3 They liaise with other medical specialties, including orthopaedic, urology, gastroenterology, respiratory medicine, and neurology. Their training allows them to identify and treat many of the well recognised health problems, including secondary musculoskeletal complications, pain, spasticity, and urological as well as neurological problems. They analyse gait and posture, and prescribe orthoses and special seating. They also prescribe assistive technology. Their paradigm of treatment is not restricted to disease per se, but addresses the whole spectrum of impairment, activity (disability), and handicap (participation).

Tuffrey and Pearce are right to point out that the survival of disabled children of adulthood is a challenge to the existing organisation of adult health services. It is our duty to maintain good standards of medical and health care and to enable such individuals to lead as full a life as possible. The evidence for the value of coordinated transition services for individuals with disabling condition exists and should be widely applied.

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1 Tuffrey C, Pearce A. Transition from paediatric to adult medical services for young people with chronic neurological problems. J Neurol Neurosurg Psychiatry 2003;74:1014–15.


Authors' reply

We are grateful to Drs Kent and Chamberlain for their comments in response to our paper.1 Young adult teams (YATs) are indeed an effective way to provide coordinated medical care for some young adults with neurological conditions. Unfortunately as Chamberlain and colleagues correctly point out in their paper,2 this provision is currently only available in a few areas of the UK so far for many young people referral to such a service is not an option. Although we do not dispute that rehabilitation physicians are skilled in dealing with many of the secondary complications of longstanding neurological conditions, young adults with additional moderate or severe learning difficulties may not be so well served.

Since YATs have been shown to enhance the quality of life of these young adults as well as being cost effective, it is essential that all areas of UK should be working towards providing similar services for all these young people. Paediatricians, physicians, and the various other professionals working with the different age groups can then work together to ensure that transition care pathways are developed and appropriately evaluated.

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Effect of a multidisciplinary clinic on survival in amyotrophic lateral sclerosis

We write in relation to the article from colleagues at Beaumont Hospital, Dublin, who report that patients with amyotrophic lateral sclerosis (ALS) attending a multidisciplinary clinic had a better prognosis owing to better medical care than those attending general neurology clinics.1 In particular the survival of the bulbar onset patients was extended by 9.6 months if they attended the ALS clinic.

The inherent error in their conclusions relates to the manner in which the two populations compared were derived. Patients were recruited to the ALS clinic group up to one year after diagnosis; the general neurology clinic population was recruited immediately from the time of diagnosis. Patients who die from ALS within some months of the diagnosis are not available to the ALS clinic cohort but these are included in the general neurology clinic population. It is probable that patients living further from the ALS clinic and those who are more disabled by ALS attend their local neurologist. Thus the ALS clinic treated a group of fitter ALS patients, whereas the general neurologists saw all ALS patients regardless of medical, social, or economic factors. We wrote the study in light of the ALS clinic group there were no deaths in the bulbar onset group for 250 days, whereas in the general neurology ALS patients it seems that deaths occurred within 30 days of diagnosis. It is difficult to be exact about the latter figures because no survival tables are given in the article. If the patients at risk or the percentage alive was been reported below the graph, this would have been of use to the reader.2 It is noteworthy that the subsequent rate of decline in survival of the two groups is exactly the same; the only differences between the groups in all the measures are the patient survival. The 100% survival was the ALS clinic group for about 200 days (whole group and riluzole subgroup) and 250 days (bulbar onset subgroup). The reason for this initial survival advantage is that in order to attend the ALS clinic one must be relatively well and not requiring immediate hospital admission. As the authors indicate in their discussion, even the ability to attend the ALS clinic on one
The survival advantage of the ALS clinic group thus appears to reflect the increased mortality of the patients treated by general neurologists in the first 200–250 days; these patients are not well enough to attend the ALS clinic.

Inferior treatment by general neurologists is implied (for example, “less attention was paid to early introduction of gastrostomy feeding”—for which no evidence was produced) and is suggested as the reason for the increased mortality.

We accept that a multidisciplinary clinic is valuable in the management of ALS, but this paper is not scientific evidence for this view. The paper would have been useful if the authors had matched ALS clinic and general neurology clinic patients, even retrospectively, for age at onset, mode of onset, disability, and duration of illness. Patients should have been deemed to have entered the ALS clinic cohort only from the date of first attendance at the ALS clinic and not from the date of diagnosis, which may have been up to one year previously. A treatment effect of the ALS clinic can only be possible from the date of first attendance. Censoring early deaths in the clearly more ill general neurology cohort should also have been considered. By avoiding these biases one might have a possible estimate of the effect of attendance at the ALS clinic.

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References

Author’s reply
We welcome the opportunity to reply to the points raised by Hutchinson and his colleagues concerning our recent paper and to provide further scientific evidence that patients attending a multidisciplinary ALS clinic have improved survival compared with patients attending a general neurology clinic. The key criticism is that the survival benefit derived from attending the ALS clinic is a result of referral centre bias. Hutchinson et al maintain that the multidisciplinary ALS clinic selects for patients with milder disease, as only these patients live long enough to be referred to the ALS clinic. While we acknowledge that it is challenging to avoid referral bias when one is quantifying the effect of a referral centre, referral bias is not a prominent factor in our study, for the following reasons. First, survival analysis of patients was diagnosed exclusively in the ALS clinic (that is, not referred from other neurologists) reveals a similar beneficial effect on mortality compared with the previously published data (median survival, 644 and 468 days for the ALS clinic (n = 44) and general neurology cohorts, respectively; log-rank test, p = 0.02). Second, ALS patients who live more than a year from the time of diagnosis (and therefore have ample opportunity for referral) continue to experience a survival advantage from attending the ALS clinic.

Hutchinson et al suggest that the parallel nature of the survival curves (that is, the similar rate of decline of both cohorts) stems from the overrepresentation in the general neurology clinic of more disabled patients who die in the first 250 days and “are not well enough to attend the ALS clinic.” Consequently, the perceived difference in mortality is artefactual. In reality, the parallel nature of the survival curves provides the strongest proof of a robust improvement in survival along ALS patients attending the ALS clinic and that this would converge rather than remain parallel, if the improved survival observed in the ALS cohort reflected the early loss of sicker patients in the general neurology clinic cohort. Kaplan–Meier survival analysis demands that the death of each individual be an independent event.

Therefore, the prognosis of an individual who dies two years after diagnosis is unrelated to that of all other ALS patients who die within the first 250 days of diagnosis. The vast majority of positive intervention studies in both ALS patients and transgenic ALS mouse models show a parallel pattern of survival curves, as is seen in our paper. This common finding is thought to reflect the ability of an intervention to slow down, but not reverse, the progression of motor neuron degeneration that underlies ALS.

A single visit to the ALS clinic is associated with an improved survival of the ALS patient. The term “ALS clinic” is a misnomer. In fact, it represents a system of care that “serves the Irish ALS population by combining the existing infrastructure of community services and the services of a voluntary organisation with a hospital based setting.” The primary advantage of all “multidisciplinary clinics” is the coordination of a network of hospital and community based ancillary services (including respiratory medicine, nursing, occupational and physical therapy, speech and swallowing, nutrition, home help, counseling, and so on) that facilitate symptomatic interventions for each ALS patient, both in a hospital setting and in their home. Therefore, any patient who attends the clinic on a single occasion is enrolled in this system and is assiduously followed up.

We read with interest the report by Weigel et al on the outcome of contemporary surgery for chronic subdural haematoma, and commend the authors for attempting to review such an extensive and diverse range of publications. The paper ably demonstrates the lack of quality evidence for the management of this common condition. However, we are concerned about the description of the paper as “evidence based”. Exclusion criteria were broad, and fewer than 5% of papers found in the Medline literature search were included in the analysis. Correspondence with the original authors for further data or clarification is an acceptable and expected part of evidence based analysis, and would have increased papers and patient numbers significantly. The data examined do not appear to have been paired, as age and comorbidity will have dramatic effects on outcome, irrespective of surgical technique.

In this context, unpaired univariate analysis is unable to produce meaningful significance. Further detracting factors include limited search procedures, absent tables to present survival data is not valid. The vast majority of modern peer reviewed journals, including JNNP, do not publish survival tables, as the graphic representation of survival curves provides a greater wealth of data.

Similarly, if the baseline characteristics of patients attending the multidisciplinary clinic are solely responsible for our findings, attendance at the ALS clinic would not be independently predictive of survival in the Cox proportional hazards model. The Cox proportional hazards model is a popular mathematical model that allows estimation of hazard ratios and survival curves, even though the baseline hazard is not specified. Furthermore, it has been established that the site of onset, age, sex, and delay in diagnosis are surrogate markers of ALS disability.

The purpose of our study was to determine the optimum method of providing care to ALS patients. We agree that a randomly assigned study in which age, sex, site of onset, and disability are matched for each cohort would be ideal to demonstrate a difference between two different clinic types. However, this could only be accomplished in the setting of a formal randomised clinical trial, which would be both logistically difficult and ethically questionable.

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Outcome of contemporary surgery for chronic subdural haematoma: evidence based review

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quality assessment and weightings of individual papers, exclusion of preomined status in deciding success rates, and a burst hole diameter defined as up to 3 cm—classified by many neurosurgeons as a craniotomy. We are concerned that, on a less careful reading, this paper could serve as a reference in the realm of "evidence based medicine", when it fails to adhere to most criteria of good evidence based medicine.

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REFERENCES

Author's reply
We appreciate the comments by Brodbelt and Warnke on our recent evidence based review on the outcome of contemporary surgery for chronic subdural haematoma. We completely agree with them that one of the surprising findings of our review was that there is indeed a paucity of methodological good studies on the surgical management of one of the most common entities seen in neurosurgical clinical routine. As most studies we reviewed were retrospective and some relied solely on expert opinions, it was not possible to achieve our initial goal of carrying out a meta-analysis of the data. Nevertheless, it was possible to scrutinise the available data with the armamentarium of evidence based methodology. It is obvious, however, that the conclusions to be drawn depend on the primary data. The proposals of the quorum conference cited are concerned primarily with improving the quality of meta-analysis of randomised clinical trials. Good clinical practice is not necessarily good evidence based medicine. There are many problems in the methodology of evidence based medicine itself, and the validity of its recommendations are increasingly being questioned. Finally, the key to understanding an article or a review is always the critical appraisal of reader themselves. This is no less important for meta-analyses or evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews. Even to the "less careful reading'' it should be clear that our evidence based reviews.

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Mesodiencephalic targeting of stimulating electrodes in patients with tremor caused by multiple sclerosis
The review of deep brain stimulation (DBS) for tremor in patients with multiple sclerosis by Wishart and colleagues1 was a good summary of the current literature, its shortcomings, and the problems associated with this type of surgery. We have recently published a report on the difficulties involved2 and would like to add a comment about targeting the site of DBS implantation in the mesodiencephalon in this patient group.

An earlier review of stereotactic ablative and DBS surgery showed that a range of different thalamic subnuclei and mesodiencephalic areas has been targeted, with variable success.2 Although a target in the thalamic nucleus ventrointermedius (Vim) is often cited, we have found—like Aziz’s group3—that a more anterior and ventral electrode placement was most likely to reduce the tremor. In the 12 patients implanted in our series, the median coordi-nates of the site of optimal intraoperative suppression were 13.5 mm lateral to the midline, 2 mm behind the AC–PC (anterior commissural–posterior commissural) midline, and 8.5 mm deep to the AC–PC plane. These coordinates suggest a subthalamic–zona incerta target, which would interrupt the dentato–Vim projections. The deepest of the quadripolar electrodes was inserted at this site, suggesting that the remaining rostral electrodes straddle the Vim or nucleus ventro-oralis posterior, which lies anterior to the Vim.

Although our targets are not dissimilar to those reported by Aziz’s group,4 we have not done intraoperative microelectrode recordings or postoperative magnetic resonance imaging to confirm our intraoperative targeting. Furthermore, most patients with tremor caused by multiple sclerosis have major brain distortions because of demyelination, plaque formation, and exc vacuo hydrocephalus when they come to stereotactic surgery. It is difficult, therefore, to know where their mesodiencephalic anatomy conforms to a stereotactic atlas. This may explain why, in our experience, targeting in patients with multiple sclerosis is considerably more demanding than in patients with either Parkinson’s disease or essential tremor.

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References

Assessing tremor reduction and quality of life following thalamic deep brain stimulation for the treatment of tremor in multiple sclerosis
We read with interest the paper by Wishart et al.1 on chronic deep brain stimulation (DBS) for the treatment of tremor in multiple sclerosis. We would like to highlight two important points.

First, reduction in tremor should not be the ultimate goal of this surgery. It is a means to an end. The most important outcome for the patient must be improved function. Surgery that reduces tremor but does not improve limb function (for example, residual ataxia) is of questionable benefit for the patient, although some surgeons may claim it as "successful" if they only assess tremor. The authors’ review of the literature outlined many papers that focused on tremor but made no mention of function. In the authors’ own series of four patients, improvements in tremor “translated into improvements in aspects of daily functioning” but no details were provided on how this was measured. We addressed this point in a recent paper dealing with thalamic DBS for 12 patients with multiple sclerosis and tremor but unfortunately this was not included in the authors’ review.2

Second, the option of unilateral thalamic DBS in a patient with bilateral upper limb tremor should be discussed. We have found that, following DBS control of their dominant hand, some patients decide they do not need (or want) the other side done. If they have significant head tremor, however, bilateral surgery is required.3

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References

Author’s reply
We value Dr Berk and colleagues’ comments and their input on the relevance of assessing limb function and its implications for quality of life. Our manuscript was written before their important contribution1 appeared in our literature search, and we
regret that it was not included in our references. We targeted mainly English language outcome studies; an additional reference of note is that of Fernández-González and colleagues.

References


BOOK REVIEWS

The Oxford dictionary of medical quotations


There are Oxford dictionaries of practically everything but not until now of medical quotations. The literary minded doctor need no longer be limited to the 15 or so entries in the regular Oxford University Press Dictionary of quotations when searching for something with which to get started a medical thesis, or display learning at lectures to rotaries clubs on the short arm of chromosome 6. Here are corralled all the most apt aphorisms an after dinner medical speaker should want. Or are they?

Peter McDonald has definitely found more statements by and relating to medicine than before. Roughly half is text listed by author to which are appended birth and death dates (where applicable)—some aphorists are happily still extant and a statement of occupation. Roughly half is text indexed as keywords linking quotes on one theme to their various authors and spokespersons. Cross reference would have been easier if the text columns had been numbered or lined—occasionally it takes some searching to find the well-honed chiselled phrase that encapsulates the very essence of bubo, erysipelas, or fistula. Not all quotations are medical men or women. And not all his sources are listed as McDonald assembles this A–Z of quasi-medical sayings. But he has clearly torn many asides at medicine by outsiders. The themes

are familiar—pomposity, fake competence, and a liking for drink and cash. Quotations from the living are in the main rather banal, some laboured, and often obscure to the point of needing to be read twice in order to spot the reason for selection. Too few raise a dutiful snark or nod of approval for their sagacity. Some of the missing biographical information should have been rather easy to locate. The most quoted people are good old Anon, Hippocrates, Osler, the Mayo brothers (of clinic fame), Shakespeare, Oliver Wendell Holmes, Claude Bernard, Bernard Shaw, and the Bible. As useful is Peter McDonald’s other approach of creating a keyword index from which contributing authors are found by secondary intention. Here the longer entries—death, doctor, disease, health, life et al—are awfully generally. Readers may have some difficulty identifying a useful stem linking into a pithy stream of apt quotations on the short arm of chromosome 6.

A Compston

Mild cognitive impairment: aging to Alzheimer’s disease


It is hard to avoid the cliché ‘timely’ to describe this book. The past few years have seen very considerable upsurge of interest in the very early pre-dementia stages of Alzheimer’s disease. The editor of the present book, Ronald Peterson, has been at the forefront of this initiative and his term, mild cognitive impairment (MCI), to describe this early prodromic stage, has been adopted around the world. It is highly appropriate, therefore, that he should edit a book on mild cognitive impairment bringing together major contributors. The book is conventional in form, containing chapters on clinical features, neuropsychiatric symptoms, neuropsychology, and structural and functional brain imaging, as well as pathology, biological markers, and treatment options. All of the chapters are very well written with admirably up to date referencing. The book will be of considerable interest to behavioural neurologists, neuropsychologists, and brain imagers. I have only one or two quibbles with this otherwise excellent book, all of which reflect my own biases. There is an inherent view that MCI represents a clinically homogenous syndrome, yet the current clinical definition is sufficiently wide to encompass the early stages of many forms of degenerative or even vascular brain disease. Consideration of the features that might separate those with early Alzheimer’s disease from those with early frontotemporal dementia, dementia with Lewy bodies, or vascular dementia is largely lacking. The neuropsychological sections were somewhat disappointing in that they neglect important work from the London group on the earliest cognitive markers for Alzheimer’s disease in subjects with known gene mutations. There is also important work from Melbourne on the value of computerised tests of visual paired associate learning that is not mentioned. The book in general is rather biased towards North America, with only two of the chapters containing contributions from European workers. These are, however, minor quibbles. All multi-authored books suffer from a variable quality. Overall this is an excellent and important contribution that will be a standard reference for some time to come. It is also extremely reasonably priced.

J R Hodges

Behavioral neurology & neuropsychology, 2nd edn


With the recent growing interest in cognitive or behavioural neurology among neurology
Disorders of body image


This book has a comprehensive chapter on the neurological examination of the spine, which is not my experience from managing patients with cervical spine disorders. The authors also mention that arterial dissection is a rare event, which is in common with other studies. However, I have seen cases where patients presented with symptoms of arterial dissection, which were not initially recognized.

M. Husain


This is a good text—one that conveys information accurately and swiftly, but also gets over the limitations of the methods described. It is a useful and informative text that covers the neurological aspects of cognitive impairment in a busy clinic.

M. Husain


This is the second edition of neurological emergencies, which I imagine is aimed at clinicians who need to be aware of the common neurological emergencies that might present to them. The book provides a good overview of the various neurological emergencies, and I think most clinicians would find this a useful resource.