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, transition 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.² It is also known that health status and psychosocial factors have a proved impact on the social participation of young disabled adults.²

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 bifida, 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 2002,³ demonstrated that participation in society was increased 90% 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, surgical, occupational therapists, 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 service exceed those compared with traditional 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. They liaise with other medical specialties, occupational therapists, 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 service exceed those compared with traditional 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.

We are grateful to Drs Kent and Chamberlain for their comments in response to our paper. 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, this provision is currently only available in a few areas of the UK so 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 the 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.

References


Authors’ reply

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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. In their model the authors corrected for some factors predictive of a poor outcome in ALS which were all overrepresented in the general neurology clinic population (increased age, bulbar onset, and shorter duration of illness at presentation); they did not correct for a measure of baseline disease severity.

The effect of the recruitment bias can be seen in the survival graphs (figs 1–3) comparing the two populations. In 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 (fig 2). 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) had been reported below the graphs 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 those related to survival. 100% survival in the ALS clinic group for about 200 days (whole group and bulbar 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 care. As the authors indicate in their discussion, even the ability to attend the ALS clinic on one
occasional or one year after diagnosis conferred a significant survival advantage with a 25% reduction in mortality (p = 0.01).

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 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, 444 and 464 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 not those who 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 neurone 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 "services 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. When a patient becomes too ill to travel to the clinic, home visits are undertaken by a specialist ALS nurse who coordinates and integrates community based, hospital based, and, in the latter stages, hospice based care. The improved survival of these patients who attend the clinic on one occasion is thus a testimony to the "ALS clinic" system and contradicts the assertion that the observed difference in survival arises from the exclusion of ALS patients who are not fit enough to travel.

In our opinion, the criticism of the use of Kaplan–Meier survival curves rather than tables to present survival data is not valid. The vast majority of modern peer reviewed journals, including JN宁P, 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 finding, 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

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
quality assessment and weightings of individual papers, exclusion of premorbid status in deciding success rates, and a burr 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 review provides an inventory of the current literature, its shortcomings, and the problems associated with this type of surgery. We have recently published a report on the difficulties involved 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. Although a target in the thalamic nucleus ventromedialis (Vim) is often cited, we have found—like Aziz’s group—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 coordinates of the site of optimal intraoperative tremor suppression were 13.5 mm lateral to the midline, 2 mm behind the AC–PC (antero–posterior–posterior commissural) midpoint, and 4.5 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 quadrupolar 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, 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 vacuolar hydrocephalus when they come to stereotactic surgery. It is difficult, therefore, to know how 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. 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 surgeons may regard 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.

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 hand tremor, however, bilateral surgery is required.

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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 contribution appeared in our literature search, and we
trainees in the UK, there is also need for a good text—one that conveys information accurately and swiftly, but also gets over the excitement of the subject. The second edition of this book is well worth looking at if you’re trying to find such a text.

The editors have made a good job, at least in most cases, of keeping their contributors to the point and confined to relatively short chapters. As a result, they have been able to cover a wide range of topics, some from a clinical perspective, others from a more scientific stance. Impressively, although the chapters are short, they are usually well referenced. Whatever your particular interest, I think most clinicians would find this a useful and informative text that covers the major neuropsychological syndromes well.

Where it perhaps could do better is on the subject of how to assess patients. This is never covered very well in most such texts, but I think it deserves far more space than given here. Also, the American perspective of some of the topics may not be to everyone’s liking. There are some chapters where one might be left with the erroneous impression that research on this topic happens only in the USA. Despite such irritations, I would recommend more than a glance at this text. It certainly deserves to be on the shelves of most medical libraries.

M Husain

**Neurologic emergencies. A symptom oriented approach, 2nd edn**


This is the second edition of *Neurologic emergencies*, a book which I imagine is aimed at physicians responsible for the emergency care of neurological problems, although this is never explicitly stated by the authors. It has a refreshingly practical approach and my initial irritation at the title was quickly dispelled by a preface pronouncing that “the days of the knee jerk CT may be drawing to a close”. I live in hope. The first few chapters set out the basics of neuroanatomy and the evaluation of neurological symptoms but thereafter they are symptom based with sections on acute weakness, headache, visual disturbance and loss, psychogenic symptoms, dizziness, and seizures. I am sure this is the right approach but it does occasionally lead to problems with cross referencing. For example, pituitary apoplexy, which rightly appears as a cause of acute headache mimicking subarachnoid haemorrhage. Similarly, non-organic seizures are discussed in the very good chapter dealing with psychogenic neurological symptoms but is hardly mentioned in the epilepsy section where they might be more appropriately placed. The book has some odd omissions—no mention of the usefulness of deep tendon reflexes in determining the level of spinal cord disease or of measuring prostate specific antigen in metastatic disease of the spine. The authors also assert that arterial dissection is a rare event, which is not my experience from managing an acute neurological intake. However, these criticisms are in the detail and overall I thought this was a comprehensive account of neurological emergencies and their management. I particularly liked the chapter on psychogenic neurological symptoms, which articulates something we are perhaps rather reluctant to admit to. “It is a legitimate use of the neurological examination to communicate to the patient that the examiner does truly possess a sophisticated knowledge of the nervous system which will in turn lead to trust and confidence in the examining physician”. This book will be useful to all those who deal with neurological emergencies. In the UK I can see it appealing to casualty officers and physicians in training but I suspect a neurological readership would find it unfulfilling.

D J Dick

**Quick cognitive screening for clinicians. Mini mental, clock drawing and other brief tests**


What is the most effective way to screen for cognitive impairment in a busy clinic, or at the bedside? This is a question that most clinicians might contemplate, even if momentarily, in their busy schedules. In this brief text, Shulman and Feinstein take the reader briskly through the arguments for and against some common screening instruments.

There are several good aspects to the book. Firstly, before immersing themselves in the tests, the authors ask what one might want from an ideal screening test. Importantly, they cover—albeit rapidly—the application of signal detection analysis to clinical methods in the form of receiver operating characteristic (ROC) curves, together with the concepts of sensitivity and specificity. This lays the groundwork for much of what follows. Secondly, several of the tests that are scrutinised (for example, Mini Mental State Examination) are critically assessed, with both their potential advantages and pitfalls discussed. Thirdly, I was particularly pleased to find that the authors have included a chapter specifically on assessing frontal lobe function, because many standard screening instruments are often poor at revealing such dysfunction. It was also useful to see a chapter devoted to structured means of interviewing informants—often a critical part of the cognitive assessment. Finally, for a small text, this book has a very good list of references, directing the reader well to the primary sources.

If there are any deficiencies, perhaps one might quibble that emphasis given by the authors to clock drawing, or the relative paucity of discussion on the use of combining tests. Similarly, the neuropsychiatric interests of the authors may not quite fit those of clinical neuropsychologists. But overall this is an interesting and useful review of the methods available for screening cognitive impairment, which can be read quickly and efficiently, even by the busiest of clinicians.

Disorders of body image


This collection of review chapters has a predominantly American–Australian provenance, with further contributions from Britain and South Africa. The central emphasis is on disorders of body image as they present to clinical psychiatrists and psychologists and four of the 10 chapters deal with clinical endocrine aspects of body image. In anthropology and normative data in childhood and adolescence provide a wider perspective. The principal body image disorders, body dysmorphic disorder and anorexia/bulimia nervosa, are the most provocative. Anorectic subjects selectively restrict carbohydrate intake, leading to a fall in serotonin activity. Normal dieters respond in much the same way, but when anorexics recover and return to a normal body weight cerebrospinal fluid 5HIAA levels are raised, suggesting a permanent state of serotonergic dysregulation. A number of studies have demonstrated an association between raised 5HIAA levels and traits of obsessiveness, perfectionism, pessimism, and negative affect (in contrast to an association between low 5HIAA and aggressivity and impulsivity). Kay et al suggest that such individuals restrict their carbohydrate intake to lower brain serotonin activity so as to modify dystonic personality traits. Acetaminophen has a similar effect in normal subjects. Some authors prefer to interpret or to integrate these findings within a neurological framework.

The various contributions are, almost without exception, clear, concise, and easy to read. This is very much a mainstream offering, dealing as it does with an area of psychiatry in which cultural, psychological, and biological factors rub shoulders and it can be recommended to a wide range of trainee psychiatrists and psychologists and to quite a few of their more adventurous trainees.

M Husain

B Toone

M Husain

B Toone