

Standards of care for patients with neurological disease

A consensus

REPORT OF A WORKING GROUP

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The Government White Paper 'Working for Patients' requires that each health district have in place by 1991 some system of medical audit, defined therein as 'a systematic critical analysis of the quality of medical care, including the procedures used for diagnosis and treatment, the use of resources, and the resulting outcome for the patient.' Such critical analysis presupposes the definition of standards against which the quality of care can be assessed.

Clinical trials can be highly effective at distinguishing which drugs or operative procedures are efficacious in achieving clearly defined outcomes — a reduction in mortality, for example, or a prolongation of time to relapse in malignant disease. There are, however, many difficulties in setting standards for the care of chronic diseases, such as multiple sclerosis or Parkinson's disease, in which scientific medicine plays a comparatively small part in caring for the patients and their families during the course of what may be a long and distressing illness.

One remit of the Research Unit of the Royal College of Physicians is to research and to stimulate research into effective patterns of medical care that achieve favourable defined outcomes. Motor neurone disease, for example, cannot be cured, but effective care should achieve the correct diagnosis at minimal cost and discomfort to the patient, keep the patient and relatives informed at all stages of the illness, relieve physical symptoms and emotional distress wherever possible, and provide promptly effective physical aids and community support. The family doctor, the neurologist and other professionals involved in the

patient's care must all have a common policy, and promptly communicate their decisions and actions.

The Research Unit invited a Working Group to consider these issues in relation to multiple sclerosis, motor neurone disease, Parkinson's disease and epilepsy at a meeting at the Royal College of Physicians on July 3rd and 4th 1989. The group included neurologists, representatives of family practice, social work, nursing, and the Patient Associations concerned with these diseases. Background papers were circulated before the meeting, and, after discussion that was often brisk, this Consensus Report was agreed. The definition, epidemiology, differential diagnosis, appropriate investigation and prognosis of each of the four illnesses was first considered, and the Working Group's papers on these can be obtained by application to the Publications Department of the Royal College of Physicians (£6.00 to cover costs and postage). Here we present themes common to the systems of care of the four chronic illnesses before considering their specific management. Points of particular concern to patients are listed. Finally a number of audit measures are proposed.

The diagnosis

When, where and by whom should the patient be told about the diagnosis?

For all the conditions considered, the patient should be informed of the nature of the illness as soon as the doctor is sure of it. Some members of the Working Group felt that it is often helpful first to ask the patient what he thinks is wrong. Many have suspicions of, for example, multiple sclerosis, and it is right to address these anxieties directly. For this diagnosis in particular, it sometimes happens that a doctor is suspicious that a first neurological event is due to demyelination. In this instance we feel that, if a patient directly asks, then the possibility should be frankly discussed.

Some patients present believing that their symptoms have a psychological basis or are related to stress. Oth-

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ers may have earlier been reassured about the absence of organic disease. For these it is helpful to explain the physical nature of their symptoms. However, it is never right to 'force' a diagnosis on a patient, and for all discussions the pace should be set by the patient.

The nature of a busy outpatient clinic often militates against an unhurried discussion of the diagnosis and its implications. Furthermore, at first disclosure of a serious diagnosis, people often fail to understand what is being said, and, even if much is grasped, they will undoubtedly wish to ask further questions after a period of reflection at home. Consideration should be given to the following options. A further outpatient appointment after a short interval can be offered, at which time the consultant can see the patient alone or with his partner as they wish. For some patients, distance may make this a not very easy proposition, and these and many other patients will prefer to discuss the implications of the diagnosis with their family doctor. Some neurological clinics are so busy that it is not practicable to offer an early unhurried appointment for further discussion. It is imperative therefore that the family doctor be informed promptly of what diagnosis has been reached, what plans have been made and what has been said to the patient.

For some patients with these disorders, the diagnosis has already been reached by their family doctor; for many more, a confident diagnosis can be reached at a single neurological consultation. Most can be appropriately investigated as outpatients if this is required. However, for reasons other than diagnostic difficulty, it is sometimes helpful to admit patients with, for example, motor neurone disease and multiple sclerosis, because it is difficult to 'name' a mortal illness, such as motor neurone disease, on the basis of a single outpatient consultation. Our experience is that after only a single outpatient consultation many will seek further opinions and investigation elsewhere. Emphasis should therefore be placed on allowing adequate time for discussion, so that the patient feels that the doctor has the expertise and knowledge to be sure about the diagnosis without recourse to abstruse investigations or to treatment available only elsewhere. Another reason for admission is that poor systems of co-ordinating investigations for an outpatient mean that they are more rapidly and conveniently undertaken as an inpatient. Furthermore, the inpatient setting does offer the opportunity for discussion of the diagnosis with dignity. Here, as in the outpatient clinic, privacy must be available for discussion of a disorder which has life-long implications for the patient. Such a policy of course has to be balanced against the needs of those for whom admission is imperative.

If these guidelines are followed, the diagnosis is made and discussed fully with each patient within a few weeks of presentation. This approach leads more often to quiet acceptance of the situation than does sudden and unexpected disclosure at the first visit, or a complete lack of information. We stress that withholding information at an early stage may jeopardise the development of a trusting partnership between

patient and doctors — a relationship that is of great importance in the management of chronic illnesses.

What should be said about the implications of the diagnosis?

What needs to be said obviously differs for each of the diagnoses here considered, but there are certain common themes. Most patients assume that the diagnosis of multiple sclerosis inevitably implies progressive disability. This erroneous view, based on extrapolating from their knowledge of a few well publicised cases, should be put into perspective. It is appropriate to err on the optimistic side when discussing prognosis early in the course of the illness. For patients with motor neurone disease, we do not feel it appropriate to be too dismal at an early stage but, as the patient becomes more disabled, it is helpful to provide reassurance for example about continuing mental clarity and the preservation of continence. For patients with Parkinson's disease, emphasis should be placed upon the essentially treatable nature of the disorder, and the importance of maintaining general health and mental and physical activity. For epilepsy, the range of topics to be covered at diagnosis is rather wider, and at an early stage advice should be given about driving, employment, domestic and leisure activities, the effects of alcohol and, where appropriate, contraception, inheritance and pregnancy. It is also important to say clearly what should be done during and after a seizure, and to discuss the aims, benefits and possible adverse effects of treatment.

For all the above disorders the patients and their families should know that the medical team is able and confident of being able to provide an answer to many of the problems that arise. If undue concern remains, it may be helpful to suggest a second consultant opinion.

Systems of care

The management of all the neurological diseases under discussion involves both family doctor and consultant neurologist. Some patients with Parkinson's disease may be under the care of a consultant geriatrician, and some cases of epilepsy under a consultant paediatrician. The consultant is usually involved in confirming or making the diagnosis and deciding upon appropriate investigation.

Each health district should formulate a policy for caring for those with chronic physical disabilities, with active rehabilitation services for those who can be improved, and with organised systems of care for these, and also for those whose diseases, like motor neurone disease, are advancing. Disabled people may be less handicapped if relatively modest adjustments to their homes are made, and if physical aids are promptly provided after skilled assessment. Every district should have a hospital based assessment team, including a physiotherapist and an occupational therapist,

who can determine, together with the patient, the relatives and the family doctor, the patient's present and likely future needs.

Professional care must be shared. The neurologist and family doctor must liaise about advice on employment, eligibility to hold a driving licence, recreation, diet, and treatment of associated symptoms and diseases. Continuity of care is essential so that, even though care of the neurologically disabled patient is shared, the patient knows whom to contact, and how to make contact, for further advice. The extent to which the long-term care of each patient is hospital or community based will vary depending on the particular manifestations of the disease and the expectations held by individual patients and their relatives.

We believe that some districts should appoint and evaluate the effectiveness of 'keyworkers' assigned to individual patients. Keyworkers might have professional training in one of a number of disciplines, such as nursing, social work or physiotherapy, but would need to acquire some expertise across all relevant disciplines so that they could give 'first level' advice to their patients. Keyworkers should be appointed by and be professionally and managerially accountable to consultant members of a department of neurology. Keyworkers could be appointed within existing resources by replacing, for example, a physiotherapist. Since the Government has adopted the Griffiths report, budgetary arrangements for keyworkers' domiciliary care will need to be made with local authorities. The keyworkers should be part of the District Disability Assessment Team, and would seek advice from team members in other professions. The keyworker would devise and help maintain a record card held by the patient, on which all relevant professions made entries.

Arrangements for outpatient care should be flexible but, for the diseases considered here, consultant neurological supervision should be available at intervals that are determined by the rate of progression of the illness or by the difficulty in controlling symptoms. Patients and their relatives, their family doctors and neurologist must all define and agree on the objectives of outpatient visits. Outpatient appointments for 'old' patients may more readily be found if patients in remission with, for example, multiple sclerosis or epilepsy are not seen needlessly on a 'routine' basis. Particular problems arise in providing continuity of care for patients with epilepsy. Patients with continuing seizures are too often relegated to management by registrars, and therefore tend to suffer because of frequent, and often ill-advised, changes and additions to drug therapy. It is important that the consultant sees and reviews such patients at least once per year so as to provide other less experienced staff with a policy for ongoing management of that patient.

Neurologists and others should be more ready to advise by telephone patients known to them, and their family doctors, so that the rigours of an outpatient appointment can be avoided. Other novel systems of care that should be tried and evaluated include the use of 'walk in' clinics, similar to those now popular

with diabetic patients, at which advice can be obtained without prior appointment. Such clinics could be based upon a local neurological rehabilitation centre, or a unit with beds for the younger patient with chronic disabilities. All concerned with the management of chronic neurological illness can help the patient by counselling, focussing in particular on helping the patient's expectations adjust to the new realities brought about by his illness. All team members should also have sufficient knowledge to advise about how chronic neurological illnesses affect personal and sexual relationships.

Local branches of voluntary patient associations (for addresses, see below) should be encouraged to form partnerships with the local neurological services. At an appropriate stage in their illness, patients should be offered the opportunity to meet representatives of these associations. Members of patient associations offer valuable support to the patient, and often also to carers, by allowing both to know that there are others to share the problems created by the disease. The associations may also be a useful source of advice about obtaining appropriate welfare benefits, about employment, and about changing personal relationships. Members and volunteers may offer practical help, for example, with transport, with gardening and with collecting pension payments, and the association may also be able to help with respite care or a holiday for a carer. However, we do not believe that the voluntary sector, as exemplified by these associations, should have to make up for deficiencies in the provision of appropriate National Health Service care.

Many patients with neurological illness are concerned to know about recent advances in the management of their condition, and the results of current research. Patient associations, through newsletters, are well placed to inform patients in this way.

Appropriate pharmacological treatment, and the management of symptoms

Multiple sclerosis

Pharmacological treatment. Drug treatment in multiple sclerosis has three aims: to accelerate recovery from an acute relapse; to relieve persistent symptoms; and to influence the long-term course of the disease.

Corticosteroids, given as short pulses of high dose intravenous methylprednisolone, intramuscular ACTH or oral steroids in diminishing doses over a few weeks, improve the rate of recovery in the context of recent symptomatic change [1,2]. There does seem to be a selective effect on pyramidal function, especially spasticity. On the present evidence, their use is not limited by adverse effects. These drugs have little role in patients with slowly progressive or persistent disability. In general, there is no place for the long-term use of steroids. It must be remembered that not every change in symptoms is necessarily due to relapse. A worsening of spasticity, for example, may be due to an intercurrent infection. The rare short-lived paroxysmal events

(eg trigeminal neuralgia or dysarthria lasting a few minutes) are well prevented by carbamazepine.

Experimental treatments designed to influence the long-term course of multiple sclerosis are not usually considered until disability is well established [3,4]. The agents currently in use are of uncertain benefit and carry an appreciable risk of adverse effects. It is noteworthy that, when those researching in multiple sclerosis were asked to consider themselves as patients, many said that they would not be prepared to be randomised into trials of immunosuppression or treatment with interferon [3]. Patients and clinicians are therefore understandably cautious early in the illness, even though that may be the best time for influencing the prognosis. Very occasionally, when the disease is pursuing a rapidly advancing malignant course, it is reasonable to try and stem progression with cyclophosphamide [5].

Symptomatic treatment. Treatment can be given at any stage to relieve persistent symptoms which, although they adversely affect daily living, do not cause overt disability. The complications of multiple sclerosis which are most amenable to symptomatic treatment include involvement of the bladder and relief of spasticity and paroxysmal symptoms.

Approximately 80% of patients describe urinary urgency or frequency which modifies or completely dominates social, domestic and economic life. A combination of mild urgency with moderate immobility usually results in incontinence and disturbed sleep, as a result of which all symptoms are tolerated less well and fatigue may develop. Bladder symptoms usually arise from loss of spinal inhibition or reflex emptying, resulting in the urge frequently to empty a partially filled bladder. If the normal mechanism of detrusor contraction and sphincter opening is uncoupled, hesitancy, retention and incontinence also occur. Drugs which inhibit detrusor contractions and maintain urethral tone (anticholinergics, such as propantheline) successfully relieve these symptoms but the therapeutic window is narrow, especially if sphincter dyssynergia co-exists. Full bladder emptying can occasionally be achieved by manual abdominal pressure or other manoeuvres, but a useful and under-used technique is intermittent self-catheterisation which achieves several hours freedom from urinary symptoms and thus enhances social activities and improves sleep. This technique requires some motivation, adequate vision and the use of one good arm. It can be performed by a partner, and may obviate the need for indwelling catheterisation or urinary diversion. Urinary infections should be promptly treated. In general, urinary problems are so prominent in patients with chronic multiple sclerosis that help from a urologist with a particular interest in neurological disease is often useful.

Spasticity may be relieved by the use of muscle relaxants, acting peripherally (dantrolene) and centrally (baclofen or diazepam), by physiotherapy, and by the use of high dose intravenous methylprednisolone. Occasionally, motor end point injections with procaine

or tenotomy may be needed to release an abnormal flexed posture of the lower limbs.

Motor neurone disease

Pharmacological treatment. No pharmacological treatment influences the natural history of the disease.

Symptomatic treatment [6]. Pain may be helped by appropriate gentle exercises, support, and by carefully adjusted analgesic treatment. Dysarthria may be helped, temporarily, by speech therapy, by intra-oral devices and, at a later stage of the disease, by the provision of appropriate aids to communication such as a CANON light-writer or POSSUM equipment linked to a microcomputer, which can also be used to control some aspects of the environment, such as lighting. Drooling of saliva may be helped by anticholinergic drugs, by radiotherapy to the salivary glands, or by surgical diversion of salivary ducts. Nutrition can be maintained via a nasogastric tube or a gastrostomy. Some patients may be helped by a cricopharyngeal myotomy, but experience shows that this is only effective if some tongue movement remains. Many patients suffer from insomnia, partly due to muscle pain and inability to get comfortable in bed. An electrically operated turning bed may be very useful. Some patients suffer from night terrors associated with hypoxia, and these may be helped by nasal catheters through which air is delivered at positive pressure. Very occasionally respiratory muscles fail before limb and bulbar muscles, and for these few patients support by a cuirasse ventilator is reasonable. Emotional lability may be helped by clomipramine.

It is helpful to explain to both patient and carer that incontinence seldom occurs, that mental clarity usually persists to the end, and that although choking attacks cause concern they are usually not fatal. Most patients die peaceably of respiratory failure.

Parkinson's disease

Pharmacological treatment. Two considerations influence pharmacological treatment — the age of the patient and the severity of symptoms. Patients who develop Parkinson's disease before the age of about 60 usually show a good response to levodopa but tend to develop fluctuations of response and dyskinesias within a few years. The disease is usually only slowly progressive in this group and dementia is not common. Patients presenting over the age of 70 tend to show a moderate or poor response to levodopa but seldom show severe dyskinesias or response fluctuations. Dementia and falls are often major problems, and the older patient may progress within a few years to total dependency.

The severity of the symptoms can be conveniently separated into mild, moderate, severe, and terminal. 'Mild' is defined as with symptoms but no disability; 'moderate' is disability (that is restriction of activity) but preservation of personal independence; 'severe' is when the disease has deprived the patient of personal

Table 1. Drug treatment in Parkinson's disease

	Mild	Moderate	Severe	Terminal
Under 60	Nil, or benzhexol + amantadine	Low-dose levodopa + decarboxylase inhibitor (DCI); ? bromocriptine	Max. tolerated levodopa + DCI; ? bromocriptine ? selegiline	Symptomatic treatment only
Over 70	Low-dose levodopa + decarboxylase inhibitor (DCI)	Optimal (max. tolerated dose) of levodopa & DCI ? + selegiline		

independence in activities of daily living. The 'terminal' phase of the disease is reached when specific therapy no longer usefully relieves the patient's symptoms, and reliance has to be placed on symptomatic treatment and nursing care. In the mild case, no pharmacological treatment may be necessary. In the younger patient, levodopa should usually be reserved for when the disease becomes 'moderate' and is causing disability, in view of the high incidence of side effects. In the elderly patient with impaired cognitive function, drug treatment (Table 1) may be limited by the tendency for all anti-Parkinsonian medication to provoke confusional states.

Patients with established disease often have more problems with associated symptoms than they do with the symptoms of Parkinson's disease. Constipation is common, especially in the elderly, and especially in patients taking anticholinergic drugs. Depression is common and frequently overlooked, even though it may well be relieved by tricyclic antidepressants. The retardation of depression may be mistaken for the bradyphrenia and bradykinesia of Parkinson's disease. Equally the limited range of facial expression and slow movements of a patient with Parkinson's disease may falsely suggest that the patient is depressed.

Akathisia (pathological restlessness) due to the disease may disturb sleep for patient and household. It may be mistaken for insomnia but seldom responds to sedative drugs. Reversal of sleep rhythm may occur because the akathisia is relieved in the day-time by levodopa, so allowing the patient to sleep. Levodopa may exacerbate depressive symptoms, especially when treatment with this drug is first initiated. Levodopa induced dyskinesias, 'on-off' fluctuations in response, or wearing off effects are difficult to treat. Small frequent doses of levodopa, or the addition of doses of bromocriptine or selegiline timed to the patient's daily activities, may help. Apomorphine by subcutaneous injection or infusion may become a more generally available and useful method of controlling these symptoms. The side effects of treatment usually require hospital outpatient supervision, with occasional admission to analyse the patient's response to treatment.

The mono-amine oxidase inhibitor selegiline has been shown to prevent the establishment of damage to

the substantia nigra of experimental animals produced by 1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine (MPTP), and there is now evidence that the course of Parkinson's disease in man may also be slowed by this drug [7]. However, larger scale prospective studies are required before this type of medication should be recommended as standard.

Symptomatic management. Patients will often need advice about a new symptom arising in the course of their disease. Depression of mood responds reasonably well to appropriate drugs, or may be due to levodopa. Confusion may be an adverse effect of medication, particularly the anticholinergic drugs. Constipation is often a problem, but may be helped by bulking or by stimulant laxatives. Advice may be needed about sexual dysfunction, incontinence, driving and interpersonal relationships.

Day centre care may provide valuable relief for the home carer of a severely affected patient. Holiday relief in a nursing home or geriatric or neurological ward may also be highly desirable. Early planning of these arrangements increases the family's trust in the neurologist's sympathetic understanding of their needs.

As the disease progresses, a visit by a district nurse or occupational therapist is often helpful. Home nursing assistance, especially for bathing, is often essential in the severe or terminal case, and hospice care is appropriate for some for the terminal stages of the illness. Home help, volunteer assistance and health visitors all have an important place in aiding and assisting the main carer. The patient and family should feel that there are others with whom they can share the problems the disease creates. At present the family doctor is best placed to organise the provision of these services, but a 'keyworker', as described above, could play a useful role.

Epilepsy

Pharmacological treatment. There is little knowledge of the prognosis of untreated epilepsy. This makes it difficult to know when to begin drug treatment. In practical terms this is decided on an individual basis after

discussion with the patient. For patients with occasional seizures or with seizures causing relatively minor symptoms, social factors such as the requirement for a driving licence and types of employment are important when reaching a decision. Similar factors apply to the decision to withdraw drugs after a period of remission of seizures. Treatment should be limited to a single appropriate drug wherever possible, although, in patients whose seizures are refractory to single drugs, brief trials of drugs in combination may be undertaken. In view of the known adverse effects, treatment should not now be initiated with phenobarbitone or primidone. These drugs also seem particularly difficult to withdraw without recurrent seizures. Sodium valproate may be preferable to these drugs for the primary generalised epilepsies (simple absences, epilepsy, juvenile myoclonic epilepsy).

There is little evidence that for other adult epilepsies there is any difference in efficacy between anticonvulsant drugs. The choice is determined by considering what potential adverse effects are particularly relevant to the patient. Estimation of serum levels of anticonvulsant drugs, although performed more often than is necessary, is useful if seizures are continuing, if there is doubt about the patient's compliance, or concern about whether symptoms are due to toxicity, or whether the patient will be able to tolerate an increase in dosage.

Symptomatic management. Most patients with intractable epilepsy will be in the community. Patients with added intellectual, neurological or psychiatric disability require liaison with professionals in ancillary disciplines — social workers, day training centre staff, mental handicap teams, etc. Here again a keyworker would be valuable.

The medical role of the neurologist should be:

- to optimise pharmacological treatment of epilepsy in any given individual as quickly as possible
- to consider at an early stage (within two or three years) whether a patient's epilepsy is medically refractory, and to explore the advisability of surgical treatment. Such patients almost always have complex partial seizures. About 70% of carefully selected patients are cured by surgery [8]. Current information suggests that, of the 50 per 100,000 population developing seizures each year, 1–2 per year would benefit from surgery to the temporal lobe. Adequate assessment of so many patients has resource implications for neurology, neurophysiology, neurosurgery, neuropsychology and neuroradiology. Regional and supra-regional teams will be required to fulfil a demand for surgery.
- For patients with medically refractory epilepsy unsuitable for a surgical approach, the neurologist's role is to ensure minimisation of therapy compatible with optimal control, so as to avoid an excessive incidence of chronic adverse effects from the drugs used in treatment. There may come a time when, because of adverse effects, it is just not worthwhile pursuing new combinations of drugs,

and patients may feel better tolerating a few seizures rather than such effects. Patients who are unlikely ever to achieve a long-term remission should be told of this.

It is especially important that family doctors and neurologists are aware of the particular problems of women with epilepsy, notably the interaction between anticonvulsant drugs and oral contraceptives, the effect of pregnancy on epilepsy and on anticonvulsant drug metabolism, and the teratogenic effect of anticonvulsant drugs.

Status epilepticus continues to have a high mortality. District general hospitals should have protocols for prompt and adequate therapy.

Points of particular concern to patients

The Working Group felt that a number of aspects of care were not well handled by neurologists at present. Points raised included:

- a lack of a co-ordinated plan for management shared between family doctor and hospital
- a lack of continuity of care in hospital clinics
- a lack of imparting early information about the diagnosis and about its meaning to the patient's possibilities in life
- a lack of recognition of the importance to the patient of fatigue, sexual dysfunction and depression
- failure to inform the patient about the relevant patients' associations
- conflicting advice from professionals about prognosis and about features specific to the disease (eg driving and epilepsy)
- poor transport facilities to outpatient clinics at which attendance is often inappropriate, and felt to be inappropriate
- the slow provision of physical aids, structural alterations to the home, or supporting services such as community physiotherapy
- the lack of adequate local facilities for welfare counselling, home nursing and physiotherapy, day care and holiday relief.

Some patients feel that the most frustrating aspect of contact with some consultant neurologists is their nihilistic approach. Clinicians need to strike a balance between therapeutic optimism, based on an increasingly active research initiative, and the exposure of vulnerable individuals to improbable or overly hazardous approaches to treatment.

Priority areas for research

Some areas are common to all the diseases we considered. We need better measures of functional status and of the quality of life, so that the quality of supportive management can be measured and become more appropriate. We need to evaluate more carefully the effectiveness of our present systems of care, and of newer proposals — the effectiveness of 'keyworkers' and of special clinics such as epilepsy clinics.

Multiple sclerosis

The priority for research into multiple sclerosis is to find an effective treatment. Because there has been no serendipitous breakthrough, systematic work on the mechanisms of demyelination is still required. The disorder has a reputation for poorly conducted and unimaginative research. The literature is contaminated by elaborate hypotheses often constructed merely to justify a particular therapeutic hunch. Understanding the mechanisms of myelin injury through an integrated approach using tissue culture, animal models and analysis of human samples may eventually lead to effective treatments. These may not immediately benefit the large number of patients in whom the disease is well established but, in the long term, learning about the aetiological factors — genetic or environmental — which render some individuals susceptible to multiple sclerosis may lead to prevention of the disease.

Motor neurone disease

The issues for motor neurone disease are similar to those for multiple sclerosis. In addition, at a simpler level, we need more information about the relative merits of techniques used to deal with dysphagia, including nasogastric tubes and the relative merits of pharyngostomy versus gastrostomy. The symptomatic management of drooling is also unsatisfactory and requires further research.

Parkinson's disease

Research priorities here are the structure and formation of the Lewy body ubiquitinated proteins and their relevance to exogenous toxins and the cause of Parkinson's disease. We also need further research into the biology of striatal implants, and the more effective delivery of dopamine agonists to the central nervous system.

Epilepsy

Further research is needed into when it is appropriate both to start and to stop anticonvulsant treatment.

Auditing the care of patients

We have here laid down standards for the treatment of four neurological illnesses. Much of what we have written about systems of care is likely to be relevant to other chronic diseases, the care of which is shared by family and hospital doctors. With defined standards, the quality of care may be audited [9].

The following are examples of general measures that could be used in audit:

- Accessibility to specialist care (eg waiting times and distance to travel for a neurological consultation).
- Accessibility to other aspects of care (eg availability of counselling, community physiotherapy, day hospitals, waiting times for provision of aids etc).

- Continuity of care (eg proportion of outpatient visits at which a patient is seen by the consultant).
- Information (eg information given about patients' associations; patients' understanding of their diagnosis; the family doctor's satisfaction with information received from the neurological clinic).
- Adverse outcomes (eg drug-induced confusional states; pressure sores).
- Inappropriate provision of medication (eg long continued steroids in multiple sclerosis, patients with continuing juvenile myoclonic epilepsy in whom sodium valproate has not been tried; inappropriate polytherapy for epilepsy).
- Failure to address significant symptoms (eg urinary infections in multiple sclerosis; depression in Parkinson's disease).
- Satisfaction of patients (and if appropriate their carers) with services and interpersonal aspects of care.
- Adequate record of plan of investigation and management.

Resource implications

Data on some of these audit measures (for example, on waiting times) can be obtained by simple secretarial work, and are suitable for routine recording for which a computer would be helpful. Other measures, such as a review of the medical records to ascertain whether these contain a written plan for future management, require first the retrieval of sets of notes and then the time of a skilled person, who may or may not be medically qualified, to review their contents. Yet other measures, such as an audit of patients' understanding of their diagnosis, require specific focused research projects. Such research audit may be small scale, but the methodology must be sound. All such initiatives will require time, and hence either additional resources or the displacement of more traditional clinical work. And yet there is evidence of an underprovision of neurological care at present. There are currently 190 neurologists in the United Kingdom, including six full-time academic posts, for a population of 56.8 million — roughly one per 373,000 [10]. The Association of British Neurologists has recently recommended that there be one neurologist per 200,000.

An attempt has been made by some neurologists in the United Kingdom to judge the amount of time that they 'ought' to spend with patients [11], but the following example will suffice. With three clinics a week, a neurologist will have available about 480 hours of outpatient time each year. And yet, with the prevalence of 240 patients with multiple sclerosis in a population of 200,000, half of this time would be used up if just one hour were spent each year with each patient with this one disease. If the community wishes to improve neurological care to the standards proposed in this paper, then there should be an increase in the number of consultant neurologists, and their training in the management of epilepsy and other causes of

chronic disability should improve. Resources will also be necessary to pay keyworkers and therapists.

References

1. Milligan, N. M., Newcombe, R. and Compston, D. A. S. (1987) A double blind controlled trial of high dose methylprednisolone in patients with multiple sclerosis. 1. Clinical effects. *Journal of Neurology, Neurosurgery and Psychiatry*, **50**, 511-6.
2. Thompson, A. J., Kennard, C., Swash, M. *et al.* (1989) Relative efficacy of intravenous methylprednisolone and ACTH in the treatment of acute relapse in multiple sclerosis. *Neurology*, **39**, 969-71.
3. Weiner, H. L. and Paty, D. W. (1989) Diagnostic and therapeutic trials in multiple sclerosis: a new look. *Neurology*, **39**, 972-6.
4. Noseworthy, J. H., Vanderwoort, M., Hopkins, M. and Ebers, G. C. (1989) A referendum on clinical trial research in multiple sclerosis: the opinion of the participants at the Jekyll Island workshop. *Neurology*, **39**, 977-81.
5. Killian, J. M., Bressler, R. B., Armstrong, R. M. and Huston, D. P. (1988) Controlled pilot trial of monthly intravenous cyclophosphamide in multiple sclerosis. *Archives of Neurology*, **45**, 27-30.
6. Newrick, P. G. and Langton-Hewer, R. (1984) Motor neurone disease: can we do better? A study of 42 patients. *British Medical Journal*, **289**, 539-42.
7. The Parkinson study group (1989) Effect of deprenyl on the progression of disability in early Parkinson's disease *New England Journal of Medicine*, **321**, 1364-71.
8. Engel, J. Jr (1987) *Surgical treatment of the epilepsies*. New York: Raven Press.
9. Hopkins, A. (1990) *Measuring the quality of care — how far have we got?* London: Royal College of Physicians (in press).
10. Langton-Hewer, R. and Wood, V. A. (1988) *Neurology services in the United Kingdom*. Bristol: Frenchay Hospital, Department of Neurology.
11. Menken, M., Hopkins, A. and DeFries, G. (1988) Norms of care in British and American neurological practice. *Archives of Neurology*, **45**, 94-8.

Addresses of patients' associations

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