Letter from Cuba
A J Wills

Imagine a country where housing, health, and education are free, without drug reps or private medicine, where the average consultant neurologist earns US $30 per month and a mobile phone is rarer than hens' teeth. This is not Arcadia, but a real place, where I was privileged to spend a week as a guest of the Department of Neurology, Santiago. This is Cuba.

Cuba is an anachronism in these days of the “new world order” and clings tenaciously to the tenets of socialism, emphasising social justice at the expense of economic freedom. As a consequence, although in close geographical proximity to the USA, political relations between the two nations have been anything other than neighbourly. Although the “Bay of Pigs” invasion and the Cuban missile crisis seem long ago, the US continues to impose a punitive economic blockade. Cuban nationals working in the US have severe restrictions on their freedom of movement. Any ship, of whatever nationality, that docks in a Cuban port is quarantined from US waters for a period of 6 months. US citizens can be heavily fined and theoretically imprisoned on their return merely for visiting Cuba. The Cuban Democracy Act prohibits foreign subsidiaries of US companies from trading with Cuba.

The collapse of the Soviet Union and removal of various trade subsidies, particularly linking sugar for oil, was a disaster for Cuba and economic conditions deteriorated markedly in the 1990s. At the beginning of this period doctors’ salaries were averaging at US $600 per month (20 times higher than the current figure). The whole population saw a marked decline in living standards with an increasing reliance on the state subsistence. Students spend 6 years at medical school and general medical training follows this. A neurological residency (specialist registrar equivalent) lasts 3 years, culminating in an exit exam prior to the attainment of consultant status. In contrast to the UK there is no opportunity for a dedicated research period leading to a higher degree. In the Santiago de Cuba province there are 16 consultant neurologists for a population of 3.9 million (1 general practitioner per 2000 inhabitants). Infant mortality is low (6.2/1000 births), comparable favourably with figures from the developed world (USA 7.0/1000, UK 5.7/1000). Life expectancy also compares favourably with the industrialised nations. Cuba continues to export doctors to developing nations, particularly in Africa, and these jobs are popular as salaries are often 10 to 20 times greater than those earned domestically. Infectious diseases are unusual, HIV still relatively rare, and malaria eradicated. Cardiovascular diseases, diabetes, and cancer are major determinants of morbidity and mortality.

Economic conditions are now improving slowly, with tourism providing much needed hard currency. This has caused problems, including a resurgence of prostitution, although street crime and drug trafficking seem to be rare occurrences.

GENERAL HEALTHCARE

Before the revolution Cuba had 3.9 hospital beds per 1000 population. This figure is now 7.3 per 1000 with 1 doctor per 174 inhabitants (1 General Practitioner per 600 v UK figure of 1/1800). Infant mortality is low (6.2/1000 births), comparable favourably with figures from the developed world (USA 7.0/1000, UK 5.7/1000). Life expectancy also compares favourably with the industrialised nations. Cuba continues to export doctors to developing nations, particularly in Africa, and these jobs are popular as salaries are often 10 to 20 times greater than those earned domestically. Infectious diseases are unusual, HIV still relatively rare, and malaria eradicated. Cardiovascular diseases, diabetes, and cancer are major determinants of morbidity and mortality.

NEUROLOGICAL SERVICES

Each province has a teaching hospital, which provides neurological services for the local population. Students spend 6 years at medical school and general medical training follows this. A neurological residency (specialist registrar equivalent) lasts 3 years, culminating in an exit exam prior to the attainment of consultant status. In contrast to the UK there is no opportunity for a dedicated research period leading to a higher degree. In the Santiago de Cuba province there are 16 consultant neurologists for a population of 1 million (1/60000). The ratio of consultants to residents (specialist registrar equivalent) is 1.6:1. There is no waiting list for neurology outpatient appointments and inpatient beds seem to be easily accessible.

Within each hospital there are dedicated neurological beds as well as a stroke unit. Doctors do not carry bleeps or pagers but are contacted by telephone. Consultants are on-call from home. Modern diagnostic facilities are available, including MRI, CT, EEG/EMG, and angiography. The Helms-Burton law, which by threatening non-US intermediaries with lawsuits in US courts, discourages healthcare investors from trade with Cuba. Maintenance is therefore difficult and when equipment breaks down repairs may take many weeks. As a consequence of the Helms-Burton law Cuban neurologists have a restricted choice of treatments for conditions such as epilepsy and migraine, relying on off-patent drugs, which can be manufactured domestically.

However, most neurological conditions seem to be managed similarly in Cuba as in the UK. There are noticeable differences in a few areas. All patients with severe myasthenia are offered thymectomy, regardless of age. Acetylcholine receptor antibody status cannot be determined so the diagnosis is made clinically and neurophysiologically. Guillain-Barré syndrome is treated with domestically manufactured intravenous immunoglobulin because of the non-availability of plasma exchange.

However, it is in the field of stroke medicine that Cuban achievements seem especially impressive. Following the recommendations of a multidisciplinary commission, there is close integration of hospital and primary care and each polyclinic (consisting of 10–15 GP practices) is staffed by community neurologists. This has encouraged an intensive educational campaign aimed at healthcare workers and patients; warning of the dangers of hypertension and transient ischaemic attacks, resulting in a dramatic (30%) reduction in stroke mortality and morbidity over 5 years. All patients with stroke are admitted to a stroke unit with dedicated beds (including high dependency beds) staffed by a multidisciplinary team. CT and carotid ultrason are routine, whereas thrombolysis is not available.

CONCLUDING REMARKS

A common philosophy, based on the principle of health for all, which is free at the point of demand, underpins the NHS and Cuban healthcare system. This commonvision could form the basis for closer collaboration in future. We in the UK could benefit from this liaison, particularly while attempting to provide high quality neurological care with economic efficiency. UK neurology needs to change so that patients with acute neurological problems can be promptly seen by someone with neurological expertise. UK neurologists should be more closely involved in the management of common neurological problems such as stroke and head injury. The Cuban model of healthcare could provide a useful paradigm for change. The benefits for Cuba are self-evident, especially if our political masters have the courage to subvert the US trade embargo, and challenge Cuba’s ill-deserved portrayal as a pariah nation.
Epilepsy

Vigabatrin, tiagabine, and visual fields

MC Lawden

A retinotoxic class effect of GABAergic antiepileptic drugs seems unlikely

The paper by Krauss et al (this issue, pp 339–343) helps to settle a controversy that has been simmering in the epilepsy world for several years. Initial reports that vigabatrin use was associated with irreversible visual field defects evoked scepticism. Various voices held that such visual field defects were not uncommon in patients with epilepsy and might be associated with epilepsy itself rather than from specific drug treatment. Evidence has now accumulated to convince all but the most sceptical that the antiepileptic drug vigabatrin, an irreversible inhibitor of GABA transaminase, has a strong tendency to produce visual field constriction by a toxic effect on the retina, although the precise mechanism by which it does so has yet to be identified. It is not clear either why a small minority of patients develop visually disabling field constriction while in most subjects visual field defects are mild and asymptomatic or indeed completely undetectable. It does not appear that such visual defects are produced by the majority of antiepileptic drugs in mainstream use—most comparative studies have compared patients taking vigabatrin with those taking carbamazepine, sodium valproate or phenytoin. It remains possible that other less widely used drugs might have similar toxic effects and attention has focused particularly on those drugs whose pharmacological effect is exerted upon the GABAergic system.

A single case of visual field constriction associated with prolonged treatment with the GABA agonist drug probabine has been reported, but this drug is not in widespread use. Of all the antiepileptic drugs tiagabine is closest to vigabatrin in its mode of action. Tiagabine blocks the reuptake of GABA at the synapse, thus increasing its availability, an effect that vigabatrin achieves by reducing its breakdown. If retinal toxicity was a class effect of drugs increasing the effect of GABA at retinal synapses then tiagabine would appear the most likely candidate to exert a similar action. It was, therefore, somewhat alarming when Beran et al reported that they had detected visual field defects similar to those associated with vigabatrin in 6 of 12 patients exposed to tiagabine. Although these results were first announced at the Third European Congress on Epileptology in Warsaw in 1998, they have not yet been published in full and are therefore difficult to evaluate. Nousiainen et al, who had earlier demonstrated a high frequency of visual field defects in vigabatrin monotherapy patients, failed to find any such defects in 15 patients treated with tiagabine monotherapy.

Krauss et al, performed static perimetry, kinetic perimetry, and electroretinography (ERG) on 12 patients treated with tiagabine and compared the results with 32 vigabatrin treated and 14 control patients. None of the tiagabine treated patients displayed any abnormality of visual field and their ERG results did not diverge from normal. By contrast, 53% of the vigabatrin treated patients had field defects and all had abnormal ERG results. Although patient numbers remain small (only 11 of the tiagabine treated patients were able to produce reliable visual fields), these results, taken with those of Nousiainen et al, indicate that visual field constriction is at worst a much rarer side effect of tiagabine than vigabatrin, and at best may not occur with the drug. A class effect of GABAergic drugs causing retinal damage now seems unlikely.

J Neurol Neurosurg Psychiatry 2003;74:286

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Objective measures for the progression of Parkinson’s disease

B Snow

Objective measure of the nigrostriatal dopaminergic deficit: evidence that SPECT does the job

A persisting frustration in the diagnosis, treatment, and research of Parkinson’s disease is the lack of an objective measure of the nigrostriatal dopaminergic deficit. In particular, we need a tool to monitor the progress of the neuronal degeneration. Although PET, with markers of presynaptic dopaminergic function such as 6-fluorodopa, is an accepted measuring tool, PET is complex, expensive, cumbersome, and not widely available. The alternative is SPECT, and Winogrodzka et al on pp 294–298 of this issue provide evidence that SPECT does the job.

Winogrodzka et al took 50 patients with Parkinson’s disease and performed SPECT scans 12 months apart. For their tracer, they chose [123I]β-CIT, which binds to the presynaptic dopamine transporters. They showed an 8% decline in binding density. This matches well with previous PET, SPECT, and clinical studies. Why is this important? Because we may soon have agents that slow the underlying progression of Parkinson’s disease. Animal studies suggest that dopamine agonists, particularly the new ones, may be neuroprotective. A recent paper has suggested that Coenzyme Q10 may also slow neurodegeneration. Before we can accept these exciting possibilities, we need objective evidence for a slowing of loss of dopaminergic function. Winogrodzka et al have calculated from their SPECT data that a 30% protective effect over 2 years could be shown with 216 patients in the treatment and control groups. This would be a large but possible study.

Before we accept the results of SPECT and PET studies of neuroprotective therapy for Parkinson’s, more work must be done. In particular, it is theoretically possible for an agent such as a dopamine agonist to change the density of the presynaptic dopamine transporters. This would give a false impression that the agent was altering the number of dopaminergic neurones. Winogrodzka et al showed no effect of agonist therapy in a subgroup of patients, but the number of subjects was too low to draw a firm conclusion. Their findings are in agreement with other short term studies, but we still do not have a definitive answer to the question. We are starting to see the results of imaging studies of possible neuroprotective agents. The question of the therapeutic agent changing the tracer binding must be answered before we can interpret with confidence the results of imaging studies of neuroprotection for Parkinson’s disease.

J Neurol Neurosurg Psychiatry 2003;74:287

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Motor neurone disease

Quality of life in motor neurone disease—towards a more practical assessment tool?

J D Mitchell, M R O’Brien

The ideal QoL measure in MND needs to fulfil a range of requirements

The optimal approach to the evaluation of quality of life (QoL) in people with motor neurone disease (MND)/amyotrophic lateral sclerosis (ALS) is still unclear. The paper by Jenkinson and others (JNPN February issue, 242–245) is an important contribution to this debate.

The ideal QoL measure in MND needs to fulfil a range of requirements. It must not be an unacceptable burden on sufferers and carers. MND patients tire easily; if the tool is too long, fatigue will ensue and the responses will become increasingly unreliable. The nature of the questions may also colour the responses obtained. The Sickness Impact Profile (SIP) can come over very negatively to subjects. The process of completing this instrument can exacerbate feelings of despair, already often a major issue in MND patients.

Should it be generic or disease specific? Although generic scales such as the SIP and SF-36 have the advantage of enabling comparisons with other disorders, they do not reflect on specific issues affecting perception of QoL in MND.

The use of instruments such as the Schedule for the Evaluation of Individual QoL, which evaluate patient selected QoL relevant issues serially during the course of the illness, do, however, allow for inclusion of items of specific concern for individual patients. MND patients often yield low scores on generic scales. Such “floor” effects can make evaluation of trends of QoL in advanced disease difficult. Also, it may not be possible to measure QoL in advanced disease, because of the effort required to complete the instrument.

The Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSASQ-40) has been a very significant step towards a disease specific QoL instrument for MND and asks the subject to give one of five possible answers to a series of 40 questions. These responses give an overall score with subscores in five domains (physical mobility, activities of daily living, eating and drinking, communication and emotional functioning). Completion takes about 15 min in our department.
One criticism of the ALSAQ-40 is that it concludes with an assessment of emotional functioning, which can be upsetting to patients, focusing as it does on burden, embarrassment, and hopelessness. The ALSAQ-5, which forms the basis of this paper by Jenkinson et al, attempts to achieve a comparable evaluation to the ALSAQ-40 by using a subset of only five questions from the ALSAQ-40. If the ALSAQ-5 was shown to have comparable validity to the ALSAQ-40 this would be a major advance. We would have a disease specific instrument capable of rapid and “painless” administration that could also be used in patients with advanced MND.

Although QoL issues are important for regulatory and other bodies, including the National Institute for Clinical Excellence, QoL data have been conspicuously absent from many major studies in MND. No prospective QoL data were collected in the trials that lead to the licensing of riluzole.4–5 If validated in long term use, the ALSAQ-5 will be an important advance and facilitate the wider application of QoL measures in MND related studies to the benefit of health care planners, patients, and carers.

J Neurol Neurosurg Psychiatry 2003;74:287–288

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Competing interests: JDM and MRO’B have received support and honoraria from pharmaceutical companies involved in the development of drugs for the treatment of MND.

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*J Neurol Neurosurg Psychiatry* 2003 74: 285-286
doi: 10.1136/jnnp.74.3.285

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