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# The incidence and lifetime prevalence of neurological disorders in a prospective community-based study in the UK

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## Summary

**Over an 18-month period, all incident cases of neurological disorders were ascertained prospectively in an unselected urban population based in 13 general practices in the London area by a General Practice Linkage Scheme with the National Hospital for Neurology and Neurosurgery. In three of these practices, the lifetime prevalence of neurological disorders was also assessed. A population of 100 230 patients registered with participating general practices was followed prospectively for the onset of neurological disorders. Multiple methods of case finding were used to maintain accuracy. The age- and sex-adjusted incidence rates of neurological disorders were calculated. The lifetime prevalence of neurological disorders was surveyed in 27 658 of the patients. The age- and sex-adjusted incidence rates were calculated for major neurological conditions. [These are expressed as rates per 100 000 persons per annum, with 95% confidence intervals (CI) in parentheses]. The commonest of these were first cerebrovascular events, 205 (CI: 183, 230); shingles, 140 (CI: 104, 184); diabetic polyneuropathy, 54 (CI: 33, 83); compressive neuropathies, 49 (CI: 39, 61); epilepsy, 46 (CI: 36, 60); Parkinson's disease, 19 (CI: 12, 27); peripheral neuropathies, 15 (CI: 9, 23); CNS infections, 12 (CI: 5, 13); post-herpetic neuralgia, 11 (CI: 6, 17); and major neurological injuries, 10 (CI: 4, 11). Lifetime prevalence rates are also reported (expressed as rate per 1000 persons with 95% CI). The most prevalent conditions were: completed stroke, 9 (CI: 8, 11); transient ischaemic attacks, 5 (CI: 4, 6); active epilepsy, 4 (CI: 4, 5); congenital neurological deficit, 3 (CI: 3, 4); Parkinson's disease, 2 (CI: 1, 3); multiple sclerosis, 2 (CI: 2, 3); diabetic polyneuropathy, 2 (CI: 1, 3); compressive mono-neuropathies, 2 (CI: 2, 3); and sub-arachnoid haemorrhage, 1 (CI: 0.8, 2). Overall, the onset of 625 neurological disorders was observed per 100 000 population annually. Six percent of the population had at some time had a neurological disorder. This is the first study of the incidence and lifetime prevalence of neurological disorders in recent times; we found that these disorders give rise to significant morbidity in the community.**

**Keywords**: community-based study; incidence; lifetime prevalence; peripheral nerve disorder; CNS disorders

**Abbreviations**: GP = general practitioner; NHNN = National Hospital for Neurology and Neurosurgery; NHS = National Health Service; NHSCR = National Health Service Central Register; OPCS = Office of Population Censuses and Surveys

## Introduction

Epidemiology has an important role in elucidating patterns in the occurrence of disease and determining risk factors and aetiology (Kurtzke, 1984; Cockerell *et al*., 1993). Information on the frequency of neurological disorders in the community is lacking (Newsom-Davis and Hopkins, 1997). The health care system in the UK lends itself well to neuro-epidemiological research (Cockerell *et al*., 1993). Health care cost does not interfere with population selection as it is free at the point of access in the UK National Health Service (NHS). Each family doctor (general practitioner, GP) is responsible for the primary care and referral of all the patients on his list, and is usually the first doctor a patient sees. When patients change GPs, their health care record is sent automatically via the National Health Service Central Register (NHSCR) to their new doctor. The central register is informed of all deaths and their cause. This ensures that for every individual in the system, there is a traceable, unique record. This system is ideal for long-term population-based studies as all individuals registered in studies can be traced efficiently.

We report the age- and sex-adjusted incidence rates (in a

**Table 1** *The linkage demographics compared with the UK census 1991*

| Age bands (years) | Linkage population (%) | UK population in 1991 census (%) |
|---|---|---|
| 0–9 | 11 | 13 |
| 10–19 | 9 | 11 |
| 20–29 | 14 | 16 |
| 30–39 | 26 | 20 |
| 40–49 | 16 | 14 |
| 50–59 | 11 | 9 |
| 60–69 | 8 | 8 |
| 70–79 | 6 | 7 |
| >80 | 3 | 4 |

**Table 2** *Ethnicity of the linkage population compared with that of the UK (based on small area statistics from the 1991 census)*

| Ethnic group | Linkage population (%) | UK population (%) |
|---|---|---|
| White | 80 | 94 |
| Irish | 7 | 2 |
| Black (Caribbean, African, other) | 7 | 1 |
| Indian, Pakistani and Bangladeshi | 3 | 2 |
| Chinese, Asian and other groups | 4 | 1 |

population of 100 230) and lifetime prevalence (in a subset of 27 657 persons) of neurological conditions in an urban population in England.

## Methods and patients/subjects

### *Definitions*

#### *Incidence rate*

This is the number of persons developing a condition within a population over a set time period, expressed as the number of cases per 100 000 people in the population per year.

#### *Lifetime prevalence rate*

The lifetime prevalence is the proportion of persons manifesting a disorder during the period of their life up to the survey date, usually given per 1000 of the population at risk (Abramson, 1997).

### *Population frame*

The population frame of these studies was an unselected population covered by 13 general practices in the National Hospital for Neurology and Neurosurgery (NHNN)–GP Linkage Scheme. These practices serve 100 230 persons, an administratively defined but otherwise unselected urban population.

All the GPs in central London were identified through the Family Health Services Association. The project was proposed to them and 13 practices where all the GPs wanted to participate actively in the scheme and whose practices kept computerized age/sex registers were chosen. In three of these practices, comprising around one-quarter of this population and where facilities were such that the researcher was able to work in the practice for long periods, lifetime prevalence was surveyed. A pilot study of the system has been reported elsewhere (Cockerell *et al*., 1996).

The demographics of this population were monitored over time for migration, births and deaths using each general practice's computerized demographic register. Small area statistics, which ascribe census data to small wards, were used to stratify for ethnicity and social class (Jarman, 1983; Majeed *et al*., 1995).

### *National Hospital for Neurology and Neurosurgery*

The NHNN is a tertiary referral centre for neurology in central London. A dedicated clinic was the basis of the facilitated link between general practice and the NHNN outpatients service. This was a general neurology clinic where only patients from the scheme were seen.

### *Case ascertainment*

Multiple methods for case finding were used to ensure complete ascertainment.

(i) Patients were referred by their GP to the 'linkage clinic' at the NHNN for a specialist opinion. Patients were assessed within 2 weeks of referral. The reason for referral was noted. If, after assessment and appropriate investigation, a new neurological diagnosis was made, it was recorded as an incident diagnosis.

(ii) Doctors in the practices were asked to inform the study office of any patient with a newly diagnosed or suspected neurological diagnosis, whether the diagnosis was made in primary care, casualty or other specialist services. Neurological diagnoses were entered onto practice computers, and copies of all relevant correspondence were kept, together with 'neurology books', where short notes about patients with suspected or confirmed diagnoses were recorded. In certain practices, all casualty cards were also kept for review. The researcher (B.K.M.) maintained a high profile through regular visits and easy access via mail or mobile telephone to discuss neurological queries and referrals.

(iii) The general practices' databases were searched regularly for all records with neurological diagnoses or where medications used were specifically for neurological

**Table 3** *The age- and sex-adjusted incidence rates for common neurological conditions compared with previously reported rates*

| Conditions | NHNN linkage age- and sex-adjusted rate (95% CI)/100 000/year | Previously reported incidence rates/100 000/year |
|---|---|---|
| Stroke | | |
| First cerebrovascular episode | 205 (183, 230) | 200 (Bamford *et al*., 1988) |
| Second cerebrovascular episode | 42 (33, 55) | 28–35 (Sorensen *et al*., 1982; Walker *et al*., 1985) |
| Intracranial haemorrhage | 10 (5, 17) | 5% of stroke, i.e. 10 |
| Seizure disorders | | |
| Epilepsy | 46 (36, 60) | 24–53 (Hauser *et al*., 1993; Brewis *et al*., 1966) |
| Single seizures | 11 (7, 18) | 20 (Kurtzke, 1984) |
| Tumours | | |
| Primary CNS tumours (benign and malignant) | 10 (5, 18) | 7 (Brewis *et al*., 1966; Walker *et al*., 1985); 15 (Counsell *et al*., 1996) |
| Parkinson's disease | 19 (12, 27) | 12–18 (Brewis *et al*., 1966; Rajput *et al*., 1984*b*) |
| Compressive mononeuropathies—all except CTS* | 49 (39, 61) | 40 (Kurtzke, 1984) |
| Arm—all excluding CTS* | 24 (17, 33) | |
| Leg—all | 20 (14, 29) | |
| Polyneuropathies | | 40 (Kurtzke, 1984) |
| Diabetic polyneuropathy | 54 (33, 83) | 11 (Cockerell *et al*., 1996) |
| All excluding diabetic and alcoholic | 15 (9, 23) | |
| Shingles | 140 (104, 184) | 71 (Cockerell *et al*., 1996); 131 (Ragozzino *et al*., 1982); 400 (Kurtzke, 1984); 480 (Schoenberg and Melton, 1993) |
| Post-herpetic neuralgia | 11 (6, 17) | 13 (Ragozzino *et al*., 1982); 34 (Cockerell *et al*., 1996); 9% of shingles (Schoenberg and Melton, 1993) |

*CTS = carpal tunnel syndrome was an excluded diagnosis.

conditions (e.g. carbamazepine, phenytoin, baclofen, sinemet).

(iv) Using the patient administration system at NHNN, patients from the general practices involved who had been patients during the study period were identified; this acted as a check on the hospital-based records.

(v) A pilot search was carried out midway through the study. This consisted of a random hand search of 4% of GP-held patient records (Lloyd George notes) in all the practices. These include letters from specialists. Incident neurological cases were identified and compared with the office records.

(vi) A full search was carried out in all practices at the end of the observation period by examining all the population's (100 230) primary care notes. The age and sex of the patient whose notes were examined were recorded. The search was carried out by medical students who were trained and supervised to identify incident neurological diagnoses in the handwritten notes and correspondence (including 'over-75' and diabetes record cards). They were instructed to be overinclusive and to take note of any cases where they were uncertain. All the sets of notes identified as positives by the students were examined by a neurologist to check diagnostic criteria. All available hospital correspondence and notes were examined and the date of diagnosis established. Information necessary for disease classification was noted. This information was compared with the office records in order to estimate the accuracy of daily data collection.

(vii) The lifetime prevalence survey was based on information obtained from computer searches in general practices and the NHNN, and the hand search of all the notes in three of the practices. The notes of all identified cases were examined by one of the authors to verify diagnosis.

## *Quality control*

To check the sensitivity of the audit, a random selection of 2% of notes from the practices were examined by an independent neurology trainee who was blind to the data already collected.

## *Data collection*

Registration of patients began in 1994; this report covers the period from January 1, 1995 to July 1, 1996.

The incidence date for a diagnosis was taken as the day that the diagnosis was made either by clinic date or the date of entry in the GP notes. For example, if a patient had tingling and numbness on a number of occasions and subsequently multiple sclerosis was diagnosed, the

**Table 4** *Incidence rates of conditions of intermediate frequency*

| Condition | Age- and sex-adjusted rate/ 100 000/year | Previously reported rate/ 100 000/year |
|---|---|---|
| Bacterial CNS infection (overall) | 7 (4, 13) | 10 (Fraser *et al*., 1973); 11 (Brewis *et al*., 1966) |
| Essential tremor | 8 (4, 14) | 24 (Rajput *et al*., 1984*a*) |
| Trigeminal neuralgia | 8 (4, 13) | 2 (Brewis *et al*., 1966); 4 (Kurtzke, 1984) |
| Benign CNS tumour | 7 (3, 13) | 10 (Kurtzke, 1984) |
| Multiple sclerosis | 7 (4, 11) | 2–8 (Brewis *et al*., 1966; Shepherd and Downie, 1980; Kurtzke, 1984) |
| Severe head injury | 7 (3, 12) | 4–6 (Langton Hewer, 1993) |
| Subarachnoid haemorrhage | 7 (3, 12) | 10–15 (Brewis *et al*., 1966; Kurtzke, 1984) |
| Subdural haematoma | 6 (3, 12) | |
| Cluster headache | 6 (3, 10) | 10 (6–14) (Swanson *et al*., 1994) |
| Cranial nerve disorder (excluding II, III, IV, VI, Bell's palsy or trigeminal neuralgia) | 6 (2, 12) | |
| Disorders of II, III, IV, VI, including pupillary abnormalities but not optic neuritis | 6 (3, 11) | |
| Aseptic meningitis | 5 (2, 9) | 1 (Brewis *et al*., 1966); 11 (10, 12) (Beghi *et al*., 1984) |
| Metastatic CNS tumour | 4 (1, 9) | 14 (12, 16) (Counsell *et al*., 1996); 15 (Kurtzke, 1984) |
| Presenile dementia | 4 (2, 9) | |
| Cerebral palsy | 3 (1, 8) | 1.5 (Brewis *et al*., 1966); 2.7 (Rosen and Dickinson, 1992); 9 (Kurtzke, 1984) |
| Neonatal encephalopathy or stroke | 3 (1, 8) | |
| Other congenital CNS abnormalities | 3 (1, 8) | 7 (Kurtzke, 1984) |
| Brachial neuritis | 3 (1, 7) | 2 (Beghi *et al*., 1985) |
| Guillain–Barré syndrome | 3 (1, 6) | 1–2 (Kurtzke, 1984) |
| Myasthenia gravis | 3 (0.8, 7) | 0.25–0.8 (Aiello *et al*., 1997); 1 (Robertson *et al*., 1998) |
| Primary malignant CNS tumour | 3 (0.7, 7) | 5 (F) 6 (M) (Ryan *et al*., 1992); 5 (Kurtzke, 1984) |
| Transient global amnesia | 3 (0.5, 7) | |
| Spinal cord injury | 3 (0.9, 7) | 1.3–4 (Kurtzke, 1984; el Masry and Short, 1997); 5 (Kraus *et al*., 1975) |

incidence date was that of the diagnosis, not the preceding and probably linked symptoms.

## *Statistical methods*

The denominator population was taken as the number of NHS patients registered with the practices. Demographics were collected at 6-monthly intervals during the study. As an urban population is mobile, the details of patients registering with and leaving the practice, and those who died, were collected. Given the prospective nature of data collection and population mobility, incidence rates were calculated for the population of July 1, 1996 plus those who died and half of those who left during the period. This correction was felt to be appropriate, as because, on average, that population had been present for half the time of the incidence study, the adjustment would give a minimum incidence rate. The denominator population for the lifetime prevalence was the population on July 1, 1996 in the three practices studied.

The incidence rates per 100 000 per annum and lifetime prevalence rates per 1000 are given after adjustment to the UK population figures from the 1991 census. Confidence limits were calculated using Poisson distribution variables

**Table 5** *The incidence rates for conditions where three or fewer were affected*

| Condition | Age- and sex-adjusted rate/ 100 000/year | Previously reported rate/ 100 000/year |
|---|---|---|
| Acute cervical myelopathy related to disc | 2 (0.2, 6) | |
| Cranial nerve injury | 2 (0.5, 5) | |
| Demyelination disorders not limited to optic nerve and not fulfilling criteria for MS | 2 (0.4, 5) | 1.2 (Kurtzke, 1984) |
| HIV encephalopathy | 2 (0.8, 5) | |
| Idiopathic myelopathy | 2 (0.4, 6) | |
| Motor neuron disease | 2 (0.3, 5) | 1–2 (Brewis *et al*., 1966; Kurtzke, 1984) |
| Spondylitic myelopathy (chronic) | 2 (0.5, 6) | |
| Truncal mononeuropathy | 2 (0.6, 6) | |
| Diabetic amyotrophy | 1 (0.1, 4) | |
| Focal dystonia | 1 (0.1, 4) | 2.2 (Nutt *et al*., 1988) |
| Non-cervical disc-related cord or cauda damage (i.e. other disc or anatomical anomalies) | 1 (0.1, 3) | |
| Optic neuritis | 1 (0.2, 3) | 1.6 (Brewis *et al*., 1966); 3 (Kurtzke, 1984) |
| Spinal malformation | 1 (0.1, 2) | 3.3 (Brewis *et al*., 1966) |

**Table 6** *CNS infections in incident cohort*

| Incident CNS bacterial infections | *n* |
|---|---|
| Tuberculosis | 3 |
| Meningococcal meningitis | 3 |
| Syphilis | 2 |
| Streptococcal meningitis | 1 |
| *Streptococcus pneumoniae* brain abscess | 1 |
| *Listeria* | 1 |
| *Cryptococcus* | 1 |
| An unidentified ventriculitis in a man dying of a reticulosis | 1 |

**Table 7** *Single incident diagnoses*

| |
|---|
| Four patients with distinct cerebellar degenerations with added features (not related to alcohol) |
| Three patients with distinct degenerative conditions the main feature of which was dementia (but not attributable to Alzheimer's or vascular disease) |
| Frontal dementia with anterior horn cell disease |
| Neurosarcoid with cord involvement |
| Neurofibromatosis |
| Tuberous sclerosis |
| Communicating hydrocephalus |
| Aqueduct stenosis |
| Cerebral cyst |
| Tonsillar herniation with Chiari malformation |
| Arnold–Chiari malformation |
| Syringomyelia |
| Myotonic dystrophy |
| Myositis |
| Idiopathic neurogenic bladder |

for small numbers (Haenszel *et al*., 1962). All figures are given to one significant place. For stroke, epilepsy and Parkinson's disease, age-specific rates were also calculated.

For case definitions, see Appendix I.

### *Exclusions*

Given the study's resource constraints, we excluded the following conditions: Bells palsy, benign positional vertigo, carpal tunnel syndrome, back pain, ear conditions and idiopathic deafness (provided they were not related to other neurology), eye conditions anterior to the optic disc (although retinal stroke was included), Menière's disease, migraine, plagiocephaly, senile dementia and tension headache.

## Results

The total population consisted of 100 230 persons on July 1, 1996. The population in whom lifetime prevalence was surveyed was 27 658 persons on July 1, 1996.

The populations are broadly comparable with the UK population but showed a slight excess of adults aged from 30 to 54 years and a reduction in the older age groups, over 65 years, accounting for 16% of the UK population and 13% of the linkage population (Table 1). Taken as a whole, the linkage population's socio-economic class and ethnicity are mixed. Non-Caucasian British constituted a greater part of the linkage population than in England and Wales as a whole (Table 2).

There was a higher rate of professional classes and less unemployment than in the UK as a whole. All the practices except one (whose population on July 1, 1996 was 13 220 and was also part of the lifetime prevalence subpopulation) score on the GP scale for socio-economic deprivation according to the Jarman index—overall the practices were eligible for deprivation payments on 42% of the population (Jarman, 1983). (The Jarman index is a validated method

**Table 8** *The lifetime prevalence of common neurological diagnoses*

| Conditions | Lifetime prevalence/ 1000 population (95% CI) | Previously reported point prevalence (PP) rates or estimated lifetime prevalence/1000 |
|---|---|---|
| Stroke | 9 (8, 11) | 5 (Langton Hewer, 1993) |
| Transient ischaemia | 5 (4, 6) | 2 (Sorensen *et al*., 1982; Langton Hewer, 1993); 6 (Geddes *et al*., 1996) |
| Active epilepsy | 4 (4, 5) | 5 (Pond *et al*., 1960; Research Committee of the Royal College of General Practitioners, 1960; Langton Hewer, 1993) |
| Congenital neurological deficit | 3 (3, 4) | 3 (Kurtzke, 1984), 2/1000 between 7 and 10 years; CNS malformation 0.7, Downs 0.5 (Kurtzke, 1984) |
| Parkinson's disease | 2 (1, 3) | 1 (PP) (Kurtzke, 1984); 2 (1) (Brewis *et al*., 1966; Langton Hewer, 1993); 2 (Mutch *et al*., 1986; Chio *et al*., 1998) |
| Multiple sclerosis | 2 (2, 3) | 1 (Shepherd and Downie, 1980; Langton Hewer, 1993; Rice-Oxley *et al*., 1995); 2 (McDonnell and Hawkins, 1998) |
| Diabetic polyneuropathy | 2 (1, 3) | 3 (Savettieri *et al*., 1993) |
| Compressive mononeuropathies (except CTS) | 2 (2, 3) | 0.4 (PP) (Kurtzke, 1984) |
| Subarachnoid haemorrhage | 1 (0.8, 2) | 0.5 (Phillips *et al*., 1980; Kurtzke, 1984) |
| Polyneuropathy (excluding diabetic and alcoholic) | 1 (0.8, 2) | 0.4 (PP) (Kurtzke, 1984) |
| Single seizures | 1 0.9, 2) | |
| Bacterial meningitis | 1 (0.8, 2) | abcess 0.02 (PP), meningitis 0.05 (PP) (Kurtzke, 1984) |

for identifying factors, excluding old age, associated with increased workload in deprived areas; it correlates well with infant mortality.) The two measures are necessary as socio-economic class excludes approximately one-third of the population.

Overall, the onset of 625 neurological disorders was observed per 100 000 population per year (including all the diagnoses tabulated except shingles); 6% of the population in whom lifetime prevalence was surveyed had had a neurological disorder.

The neurological disorders ascertained are tabulated giving age- and sex-adjusted incidence rates per 100 000 per annum (common conditions in Table 3, intermediate conditions in Table 4, unusual conditions in Table 5, a breakdown of serious CNS infections in Table 6 and single incident diagnoses in Table 7). Lifetime prevalence per 1000 persons is also given (common diagnoses in Table 8, less frequent diagnoses of the CNS in Table 9, and the peripheral nervous system in Table 10). Age-specific incidence rates for stroke, epilepsy and Parkinson's disease are given in Table 11.

### *Quality control*

During the final search, 97% of the general practice Lloyd George notes were examined. The check for false negatives after the complete notes search uncovered three missed cases amongst 1655 notes searched, giving a false-negative rate of 0.2%. The check for false negatives in the lifetime prevalence subpopulation uncovered six missed cases in the complete notes search amongst 719 notes searched, giving a false-negative rate of 0.8%.

## Discussion

This is the first large prospective study measuring the burden of all serious neurological conditions in the community in the UK in over 30 years; the Carlisle study was carried out before many investigations and disease definitions currently in use were available (Brewis *et al*., 1966). Carlisle had a mainly Caucasian English population, with an age profile which differs from current demography (Perkin, 1997). Much recent neuroepidemiological work has been carried out at the Mayo Clinic, Rochester, Minn., USA. Studies such as those at Rochester, USA, whilst useful, describe a population whose ethnicity and lifestyle do not necessarily reflect that of the UK (Glista *et al*., 1977). The routine statistics generated in primary care are unlikely to be accurate. Generalists report diagnostic uncertainty in neurology (Newsom-Davis and Hopkins, 1997). GP computer systems are set up for clinical work; as they are not designed specifically for epidemiology they will often fall short of the necessary standards for data collection due to poor validity and completeness. The UK national morbidity surveys (Ebrahim, 1995) have attempted to address the frequency of complaints in general practice. However, a major limitation is that conditions with low incidence and relatively chronic courses may give false incidence rates as patients in this system will count as 'new' when they change practice and see a participating GP for the first time (Newrick *et al*., 1996). Population-based estimates from the US report point to prevalence rates of neurological conditions (excluding headache, back pain and discs, mental retardation, psychosis, non-neurological visual and hearing loss and nervous system trauma) of 3.6 per 100 (Kurtzke, 1982). In the UK, diseases of the nervous system accounted for 7.6% of all GP consultations between 1981 and 1982;

**Table 9** *The lifetime prevalence of less common CNS disorders*

| Conditions | NHNN-linkage lifetime prevalence (95% CI)/1000 | Previously reported prevalence rates/100 000 |
|---|---|---|
| Meningitis or encephalitis | 1 (1, 1) | |
| CNS infections | | |
| Aseptic meningitis | 0.9 (0.6, 1) | |
| Essential tremor | 0.8 (0.5, 1) | 3 (1) (Langton Hewer, 1993) |
| Polio | 0.7 (0.4, 1) | |
| Severe head injury | 0.6 (0.4, 1) | 1 (Langton Hewer, 1993) |
| Optic neuritis | 0.6 (0.3, 1) | 0.1 (Kurtzke, 1984) |
| Benign CNS tumours | 0.5 (0.3, 1) | 0.6 in brain, 0.1 in cord (Kurtzke, 1984) |
| Intracranial haemorrhage | 0.5 (0.2, 0.8) | |
| Other movement disorders | 0.4 (0.2, 0.7) | Hereditary ataxia 0.08 (Kurtzke, 1984) |
| Viral encephalitis | 0.4 (0.2, 0.7) | 0.1 (Kurtzke, 1984) |
| Spondylitic and compressive myelopathy | 0.4 (0.2, 0.7) | |
| Cluster headache | 0.3 (0.2, 0.6) | 0.3 (F), 1 (M, M + F) (Ekbom *et al*., 1978; D'Alessandro *et al*., 1986) |
| Subdural haemorrhage | 0.3 (0.2, 0.6) | |
| Malignant CNS tumours | 0.2 (0.06, 0.4) | Primary malignant 0.05; metastatic in brain 0.15, 0.05 in cord (Kurtzke, 1984) |
| PN or plexus injury | 0.2 (0.05, 0.4) | 0.3 (Kurtzke, 1984) |
| Demyelinating conditions not fulfilling the criteria for MS | 0.1 (0.04, 0.3) | |
| Cauda equina lesion | 0.1 (0.02, 0.4) | |
| Dystonia primary secondary | 0.1 (0.02, 0.4)<br>0.1 (0.03, 0.3) } | 0.3 (Nutt *et al*., 1988) |
| Benign intracranial hypertension | 0.1 (0.02, 0.3) | |
| Intrinsic myelopathy | 0.1 (0.02, 0.3) | Syrinx 0.07 (Kurtzke, 1984) 0.06 (Foster, 1980) |
| Spinal cord injury | 0.1 (0.02, 0.3) | 0.5 (Kurtzke, 1984), 0.8 cited in (Kraus *et al*., 1975; Kraus, 1978) |
| Narcolepsy | 0.1 (0.02, 0.3) | |
| Motor neuron disease | 0.1 (0.01, 0.3) | 0.04–0.1 (Kurtzke, 1984); 0.06 (Langton Hewer, 1993) |
| Aqueduct stenosis and hydrocephalus in adults | 0.1 (0.01, 0.3) | |
| HTLV 1 myelopathy | 0.04 (0, 0.2) | |
| Transient global amnesia | 0.04 (0, 0.2) | |

this higher rate may reflect superior case ascertainment (McCormick and Rosenbaum, 1990). However, the latter figure includes headache and diseases of the ears and eyes. The Harris report looked at all disability in private households amongst those over 16 years of age in the UK. Disabilities were divided into groups and, taking those relevant to neurology (CNS disorders, muscular dystrophies, congenital malformations of the spine and hydrocephalus, cerebral birth injury, senility as a cause of cognitive disability), 78/1000 were disabled to some extent by these disorders (Harris, 1971). The OPCS survey of disability 16 years later graded disability according to severity as well as overall frequency. The prevalence of complaints relevant to neurology was 13% for 'CNS disorders', 2% each for dementia and mental retardation, and 6% for back complaints. In a later study, 'CNS complaints' accounted for 7% of disability overall but for 16% of conditions with a high severity score (9 or 10/10; Martin *et al*., 1988).

One of the central problems in the measurement of neurological disorders is that only 'the tip of the iceberg' is known to health care professionals. This effect is more marked with conditions which are common, asymptomatic or stigmatized, such as migraine, mild asymptomatic neuropathy, dementing illnesses and parkinsonism. Case finding methods may need to be tailored to the disease's spectrum of severity and frequency. Neurology has the highest number of conditions listed in the International Classification of Diseases. The high numbers of different uncommon diseases mean that exhaustive methods of case ascertainment and audit are most appropriate, as sampling error increases with

**Table 10** *The lifetime prevalence of less common PNS disorders*

| Conditions | NHNN-linkage lifetime prevalence (95% CI)/1000 | Previously reported prevalence rates/100 000 |
|---|---|---|
| Leg mononeuropathy—all | 1 (0.8, 2) | |
| Arm mononeuropathy—all excluding CTS | 0.7 (0.5, 1) | |
| Trigeminal neuralgia | 0.7 (0.4, 1) | 0.4 (Kurtzke, 1984; Langton Hewer, 1993) |
| Post-herpetic neuralgia | 0.7 (0.4, 1) | |
| Muscular dystrophies | 0.4 (0.2, 0.7) | 0.02–0.05 (Langton Hewer, 1993); 0.6 (Kurtzke, 1984) |
| Myasthenia gravis | 0.4 (0.2, 0.7) | 0.04–0.1 (Kurtzke, 1984; Langton Hewer, 1993); 0.08 (Christensen *et al*., 1993), 0.1 (0.08, 0.2) (Aiello *et al*., 1997); 0.4 (Kurtzke, 1978) |
| Eye movement disorders | 0.3 (0.2, 0.7) | |
| Brachial neuritis | 0.3 (0.1, 0.6) | |
| Guillain–Barré syndrome | 0.2 (0.08, 0.5) | 0.08 (Langton Hewer, 1993) |
| Horner's syndrome | 0.2 (0.04, 0.4) | |
| Other mononeuropathy | 0.1 (0.04, 0.3) | |
| Pupillary abnormalities | 0.08 (0.01, 0.3) | |
| Sacral plexitis/plexopathy | 0.04 (0, 0.2) | |

**Table 11** *Age-specific incidence rates for stroke, epilepsy and Parkinson's disease*

| Age band | Incidence rates /100 000/year by age band adjusted to the UK population | | | | |
|---|---|---|---|---|---|
| | First stroke | | Epilepsy | Single seizures | Parkinson's |
| | Men | Women | M + F | M + F | M + F |
| 0–4 | | | 86 | 32 | |
| 5–9 | | | 46 | 12 | |
| 10–14 | | | 94 | | |
| 15–19 | | | 52 | | |
| 20–24 | | | 33 | 16 | |
| 25–29 | | | 19 | 19 | |
| 30–34 | | | 24 | 5 | |
| 35–39 | | | 54 | 14 | |
| 40–44 | 35 | 82 | 18 | 9 | |
| 45–49 | 194 | | 50 | 10 | 20 |
| 50–54 | 240 | 167 | 50 | | |
| 55–59 | 560 | 203 | 31 | 15 | |
| 60–64 | 1051 | 629 | 34 | 34 | 50 |
| 65–69 | 817 | 940 | 37 | | 37 |
| 70–74 | 850 | 926 | 142 | | 222 |
| 75–79 | 972 | 1271 | 50 | 25 | 100 |
| 80–84 | 806 | 890 | 32 | | |
| 85–89 | 299 | 757 | 29 | 29 | 116 } |
| >90 | | | | | |
| >40 | 467 | 446 | | | |

rarer events. Many neurological conditions have chronic courses, so long-term follow-up is important to our understanding of them.

The UK is an ideal system to study epidemiology, as health care is free at the point of access and each patient can be traced through the system, allowing complete long-term follow-up, which has proved difficult elsewhere. Seventy-eight percent of all registered patients consult their GP each year and 13% of all consultations are for conditions of the nervous system, including eye and ear complaints (Ebrahim, 1995).

The case note search in this study was unusually thorough and accurate for a study of such size. It should not be forgotten that this survey will not include the small number of patients who are in long-stay hospitals for severe neurological problems. The confidence limits are wide for the less frequent incident conditions and in the study of prevalence in the smaller population. The demographics of the linkage

population are slightly atypical for the UK as a whole, but are probably typical for conurbations.

In the general population, 0.6% had a neurological condition diagnosed in the previous year and 6% had had a neurological condition at some time. The incidence rates for certain conditions have never been reported or have only been reported in single studies, e.g. compressive neuropathies, polyneuropathies, subdural haematoma, dystonia and myelopathy. That the findings of the study are in agreement with those of the best community-based studies of epilepsy, first stroke and Parkinson's disease helps to confirm the validity of the approach. The methodological sensitivity was high. This means that the figures we report are likely to reflect minimum rates of certain neurological conditions (e.g. epilepsy). Despite this, we found lifetime prevalence rates that were higher than previous estimates for stroke, multiple sclerosis, subarachnoid haemorrhage and neuropathies (Langton Hewer, 1993).

In the UK, non-Caucasian communities tend to remain within cities. Hence, studies done in largely rural areas will have lower rates of ethnic groups, and city-based studies higher rates than the average for the UK. To date, the majority of studies have been done in rural areas which are easier to study as they are more stable; studies of city populations are needed to balance this tendency.

A high level of accuracy was achieved, and this project will be a basis for ongoing neuroepidemiological work. The study of lifetime prevalence could be improved if an assessment of disability was also made to measure neurological disability in the community; however, within the resource constraints of the study, this was not feasible. Despite these caveats, the figures generated are likely to be more accurate than current estimates used to allocate health services in the UK. Further neuroepidemiological work within this community sample would allow examination of trends over time and the exploration of the effects of deprivation and ethnicity on neurological disorders.

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## Appendix I. Case definitions

**Aseptic meningitis** was defined as a benign self-limiting condition of suspected or demonstrated viral origin with fever and meningism,

without evidence of parenchymal brain involvement, and CSF sterile for bacteria, fungi and parasites, with lymphocytic and mononuclear pleocytosis (Beghi *et al*., 1984).

**Benign CNS tumour** included all types of benign neoplasms within the cranium and spinal cord, except those confined to the pituitary sella.

**Benign intracranial hypertension** is defined as a raised CSF pressure (documented >20 cm CSF pressure in non-obese and >25 cm CSF pressure in obese patients), in an alert patient without localizing signs, normal neuroimaging including of the venous sinuses but except empty sella, in the absence of another cause of raised intracranial pressure (Radhakrishnan *et al*., 1993).

**Cluster headache** is diagnosed in the presence of at least five 'attacks of severe strictly unilateral headache orbitally, supraorbitally and/or temporally, lasting 15–180 min and occurring from once every other day to eight times a day. Associated with one or more of the following: conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, facial and forehead sweating, meiosis, ptosis, eyelid oedema. Attacks occur in series lasting for weeks or months (so-called cluster periods) separated by remission periods usually lasting months or years.' These attacks are not classified as cluster headache in the presence of trauma, cerebrovascular disease, intracranial disorders such as raised intracranial pressure or infection or neoplasia, substance use or withdrawal, non-cephalic infection, metabolic disturbance or related to the bony structure of face, skull or neck (Headache Classification Committee of the International Headache Society, 1988).

**Dystonia** is a rare condition which can be divided into focal and generalized, and primary and secondary. Focal dystonia is an abnormal movement or spasm of the eyes, oromandibular region, larynx, neck, hand or limb muscles which may be task specific; generalized dystonia involves more than one area (face, limb and axial musculature) (Nutt *et al*., 1988). Secondary dystonia was invoked where an underlying structural lesion was so placed that this was likely to provoke the dystonia. This diagnosis was only accepted if a neurologist had made the diagnosis.

**Encephalitis** was defined as a condition of acute or subacute onset, with neurological symptoms or signs indicative of brain parenchyma involvement, i.e. seizures, coma, focal neurological signs or impairment of mental function, in the absence of evidence of other conditions or non-viral infections. Mild obtundation and febrile convulsions were not considered encephalitic (Beghi *et al*., 1984).

**Epilepsy** is defined as two or more unprovoked seizures. Single and acute symptomatic seizures were coded separately. Active epilepsy was when the person was still on anti-epileptic medication or had had seizures in the last 12 months (Commission on Epidemiology and Prognosis, 1993).

**Essential tremor** causes postural or action tremor predominantly in the upper half of the body, which worsens with action in a patient who gives a history of often recurring or continuous tremor in the extremities and/or head, that could be demonstrated on clinical examination in the absence of any systemic or neurological disorder associated with tremor. The patient must not be taking a drug known to cause tremor. A positive family history is supportive, but not essential to the diagnosis (Larsson and Sjogren, 1960).

**Malignant brain tumours** included primary and secondary tumours within the cranial cavity and spinal cord, but excluded pituitary masses, congenital cerebral tumours and dermoids. These included those that were identified only on scan without histological confirmation, but excluded those diagnosed on clinical grounds only.

**Motor neuron disease** is the relentlessly progressive degeneration of motor neurons diagnosed in the presence of a progressive pure motor syndrome (atypical features are sensory signs, parkinsonism and dementia) according to the El Escorial criteria (Brooks, 1994).

**Multiple sclerosis** was defined according to the Poser criteria for clinically definite or laboratory-supported multiple sclerosis (Poser *et al*., 1983). All cases were seen by a neurologist and had had an MRI scan consistent with the diagnosis.

**Myasthenia gravis** is defined as the presence of rapid fatigue in one or more muscle groups, weakness aggravated by exercise and relieved by rest, showing a significant response to anticholinesterase drugs. Supportive data were anticholinesterase antibodies if positive, and electromyography. The exclusion of Lambert–Eaton syndrome was by clinical examination with help from EMG (Christensen *et al*., 1993; Rowland and McLeod, 1994).

**Myelopathy**. Disc-related myelopathies were cases in which an acute disc protrusion had produced a myelopathy. Spondylitic myelopathy is the chronic myelopathy associated with spondylitic encroachment on the cord.

**Myositis** is an inflammatory disorder of muscle including dermatomyositis and polymyositis (C-2.1,2,4 in World Federation of Neurology Research Group on Neuromuscular Diseases, 1994*b*); cases were included in which there was biopsy evidence of myositis or a neurologist had made the diagnosis in the presence of raised creatine kinase and inflammatory markers and the characteristic clinical picture.

**Narcolepsy** is a disorder characterized by excessive daytime sleepiness associated with cataplexy, sleep paralysis and hypnagogic hallucinations (Diagnostic Classification Steering Committee, 1990). The most specific of these is cataplexy (Parkes *et al*., 1995). The diagnosis had to be made by a neurologist or a physician in a sleep disorder clinic.

**Parkinson's disease** was defined as at least two of the cardinal signs: resting tremor, rigidity, bradykinesia and impaired postural reflexes. Artherosclerotic- and neuroleptic-induced parkinsonism were excluded. All patients were seen by a neurologist (Rajput *et al*., 1984*b*).

**Peripheral neuropathy**

(i) **Polyneuropathy**. This was taken as a patient having clinical objective signs consistent with the diagnosis in the presence of an established cause, such as diabetes. Alternatively, an EMG diagnosis was required.

(ii) **Compressive neuropathy**. This was often classical in presentation and was not necessarily EMG confirmed. Carpal tunnel syndrome was an excluded diagnosis.

**Plexopathy/plexitis** is an often painful lower motor neuron, non-dermatomal weakness with or without sensory disturbance or deficit. This was always EMG confirmed. It may affect the lumbosacral or brachial plexus. The latter is most commonly affected by an idiopathic condition: **brachial neuritis** or **neuralgic amyotrophy** (Hughes, 1995). **Diabetic amyotrophy** was defined as an acute or subacute asymmetric weakness and wasting of proximal lower limb muscles in a diabetic associated with pain at onset; nerve condition studies had to confirm that multiple roots were affected. Palsy due to obstetric injury was not included under this heading.

**Severe head injury** was defined by outcome. Only those who developed objective neurological deficit after head injury were included (Jennett, 1996).

**Shingles** is diagnosed in the presence of a vesicular rash in a dermatomal distribution often preceded by paraesthesiae and accompanied by pain. **Post-herpetic neuralgia** is a pain syndrome following shingles and is diagnosed when pain is ongoing a month after the rash.

**Stroke** is a syndrome characterized by 'rapidly developing clinical signs of focal (or global) disturbance of cerebral function, with symptoms lasting at least 24 h (**transient ischaemic attacks** are vascular episodes lasting <24 h) or leading to death, with no apparent cause other than of vascular origin', and 'any or all of a group of disorders including cerebral infarction, intracerebral haemorrhage or subarachnoid haemorrhage' (Hatano, 1976). New diagnoses of 'silent cerebral infarction' were counted as incident stroke cases on the assumption that these patients had all presented neurological symptoms requiring scanning for these 'silent' infarcts

to become apparent. Likewise, patients newly diagnosed with multi-infarct disease, atherogenic parkinsonism and small vessel disease who had not been diagnosed previously with cerebrovascular disease were considered as incident stroke cases. Patients diagnosed with subarachnoid haemorrhage who had neither neurological deficit nor infarction on scan were not considered to have experienced a stroke.

**Subarachnoid haemorrhage** was diagnosed in the presence of 'an atraumatic lumbar puncture yielding bloody CSF in the clinical setting of an acute, devastating neurological illness lacking focal signs at onset usually accompanied by headache and meningism' (Phillips *et al*., 1980).

**Transient amnesia** is defined as a witnessed episode of clear-cut anterograde amnesia with clear consciousness and preservation of personal identity without any accompanying neurological signs during or after the attack, which resolves in 24 h or less. Those with active epilepsy, or with epileptic phenomena during the attack or who had had a head injury in the last year were excluded (Hodges and Warlow, 1990).

**Trigeminal neuralgia** is defined as a painful unilateral affliction of the face, characterized by brief electric shock-like (lancinating) pains. The pain must last 1 s to 2 min, and have four of the following characteristics: a distribution of one or more divisions of the trigeminal nerve; quality of pain described as intense, sharp, superficial, stabbing or burning; precipitating triggers on the face or in the mouth; and asymptomatic between attacks. This must be in the absence of neurological deficit. Other causes of facial pain, including multiple sclerosis and brainstem infarction, were rejected by history, examination or investigation (Headache Classification Committee of the International Headache Society, 1988).

## Appendix II. Collaborators

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