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Utility of Scottish morbidity and mortality data for epidemiological studies of motor neuron disease

A M Chancellor, R J Swingler, H Fraser, J A Clarke, C P Warlow

Abstract

***Objectives*—To determine the accuracy of (1) hospital discharge data and (2) death certificates, coded as motor neuron disease (MND).**

***Design*—Comparison of data from The Scottish Motor Neuron Disease Register (SMNDR) with routinely collected Scottish Hospital In-Patient Statistics (SHIPS) and death certificate coding.**

***Setting*—Scotland UK.**

***Patients*—1) 379 adults (>15 years) discharged for the first time from a Scottish hospital in 1989–90 and (2) 281 deaths in the same period assigned to the International Classification of Diseases (ICD)–9, category 335 (MND).**

***Main outcome measures*—The sensitivity and positive predictive value of a diagnosis of MND as retrieved by (1) the Information and Statistics Division of the Common Services Agency for the Scottish Health Service for morbidity data and (2) the Registrar General's office for mortality data, using the SMNDR as the 'gold standard'.**

***Results*—(1) Thirty per cent of adult patients identified as having MND by SHIPS did not have this disease and 23% of patients with MND did not appear on SHIPS. The sensitivity of a diagnosis of MND, as retrieved by SHIPS, was 84% and the positive predictive value was 70% overall. Miscoding of patients with pseudobulbar palsy caused by cerebrovascular disease was the major source of false positive error. The incidence of adult onset sporadic MND was over estimated by SHIPS by a factor of 1·6. (2) Mortality data were more accurate, with a false negative rate of 6% and a positive predictive value of 90%.**

***Conclusions*—Coded hospital discharge data are an inaccurate record of a diagnosis of MND and cannot, in their present form, be used as a reliable measure of disease incidence in Scotland. Greater care is required in the preparation of discharge summaries and coding if these data are to be useful for health care planning and epidemiological research. SHIPS is, however, an important source of information to achieve a complete sample of patients with MND. There is also a problematic false positive rate for mortality data but this source more closely approximates true incidence.**

Department of Clinical Neurosciences, Western General Hospital, Edinburgh
A M Chancellor
H Fraser
C P Warlow
Department of Neurology, Dundee Royal Infirmary, Dundee
R J Swingler
Information and Statistics Division, Scottish Health Service, Common Services Agency, Edinburgh.
J A Clarke

Correspondence to:
Dr A M Chancellor, Department of Neurology University Hospital of Wales, Heath Park, Cardiff CF4 4XW

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National Health Service managers, administrators, and public health specialists use morbidity and mortality data to plan the appropriate allocation of health care resources. Therefore, it is important that this information is as accurate as possible. Motor Neuron Disease (MND) is a generic term for a group of system disorders, with childhood and adult types, characterised by loss of motor neurons which result in wasting and weakness of skeletal muscle while sparing cognitive, sensory, ocular, and sphincteric function. For the vast majority of patients MND is without recognised cause.[1] There is considerable interest in the worldwide distribution of MND because the identification of an environmental aetiology might be derived from studies of geographical clustering or temporal trends.[2]

The incidence of rare diseases of insidious onset is difficult to measure and monitor on a large enough scale to provide precise estimates so proxy statistics have been used. For example, if most patients are admitted to hospital, and this proportion is stable in place and over time, the first discharge rate should reflect the incidence. However, some investigators,[3] using unverified hospital discharge rates, have reported an incidence of MND approximately two to three times that measured by other means. This discrepancy suggests that a large but unknown number of patients coded as MND after hospital discharge do not actually have this disease.

More commonly, mortality statistics are used both to follow trends and as an indirect measure of the incidence of MND,[4 5] with the belief that these data have a high degree of accuracy, given the generally fatal outcome of this disease within a year or two of diagnosis. In 1979 the ninth revision of the ICD[6] introduced an amended classification of anterior horn cell disease and the clinical subtypes of MND were included under the rubric 335·2 (table I).

The accuracy of such routinely collected statistics depends on:

(1) The proportion of incident patients admitted to hospital or dying from the disease;

(2) The precision of diagnosis at discharge from hospital or at the time of death;

(3) The satisfactory completion of Scottish Morbidity Record (SMR 1) forms and death certificates;

(4) The appropriateness of the ICD coding;

(5) The accuracy of transcription of information to the Scottish Hospital In-Patient Statistics (SHIPS) and the Registrar General's computer file, and;

(6) The efficiency of the linkage programme which retrieves first, rather than all, hospital

Table I International Classification of Diseases-9 (1979); 335, anterior horn cell disorders

335·0 Werdnig-Hoffmann disease
Infantile spinal muscular atrophy
335·1 Spinal Muscular atrophy
Kugelberg-Welander
Adult spinal muscular atrophy
335·2 Motor Neuron Disease
Amyotrophic lateral sclerosis
Motor neuron disease (bulbar) (mixed type)
Progressive muscular atrophy (pure)
335·8 Other

discharges and the satisfactory retrieval of death certificates for the relevant year.

There have been no formal studies comparing the accuracy of hospital discharge data with other case finding methods or the extent to which these data will identify all patients who develop MND, yet this is an important requirement for studies which require an unbiased and complete sample of cases. There is some information on the false negative rate of death certificates[4 5 7] but much less on the false positive rate.[8] We have therefore compared the accuracy of hospital discharge data and mortality statistics with a recently established prospective register for MND in Scotland.

Methods

THE SCOTTISH MOTOR NEURON DISEASE REGISTER

The Scottish Motor Neuron Disease Register (SMNDR) was established in January 1989 with the aim of studying the incidence, distribution, clinical features, and prognosis of MND in Scotland. The methods of this prospective, collaborative study are described in detail elsewhere.[9] The SMNDR is based on a system of registration, during life, of all patients who develop MND in Scotland and considerable efforts have been made to ensure complete case ascertainment, mainly through neurologists and neurophysiologists but also using self referral through the family care officers of the Scottish Motor Neurone Disease Association; an annual approach to all general practitioners (GPs) in Scotland by letter; a review of mortality data; and inspection of all hospital records, however the patients came to our attention. A set of formal diagnostic criteria for MND has been devised for application to this study in accordance with widely accepted clinical criteria and only patients who fulfilled the SMNDR criteria after all record review and follow up to January 1992 were considered to have MND. The SMNDR was considered as the 'gold standard' for the purpose of this study, although some registrations were obtained for the first time from SHIPS and mortality returns. The 1989 crude incidence was 2·2/100 000 per year.[9]

MORBIDITY DATA RETRIEVAL

The inpatient and day case records summary sheet of hospital morbidity in Scotland (SMR1) collects biographical, administrative, and clinical information relating to every hospital death, discharge, or transfer and includes a section pertaining to diagnosis which should 'be completed by the doctor on discharge of patient'.[10] Clerical staff complete the remaining details and code the assigned diagnosis according to the ICD-9. This form is the basis for data transferred to the SHIPS computer, although it has now been replaced by a computerised patient administration system in a number of hospitals.

A request was made to the Information and Statistics Division of the Common Services Agency for the Scottish Health Service for details of all patients coded as ICD-9 335 in any part of their discharge form for 1989 and 1990. Through a record linkage system, details were restricted to the first discharge with this diagnosis. This was available in the August after the year of interest. The hospital records of all these patients in Scotland who were not already registered with the SMNDR were requested, with the permission of the consultant in charge of the patient, through the relevant medical records officer. Those patients who fulfilled the criteria for probable or definite MND, who were not already included on the SMNDR, and who had been diagnosed on or after January 1 1989 (incident) were therefore eligible for inclusion in the SMNDR and were registered accordingly. Other patients had been coded correctly but were discovered to be prevalent to the SMNDR (diagnosis made before January 1 1989), despite a first admission in that year. The diagnoses of all patients coded as ICD-335 who did *not* have MND were recorded and the records were examined to try and determine the reason for the error. The SMNDR was also used to identify all patients diagnosed as having definite MND and first discharged from hospital in 1989 and 1990 who did *not* appear in the SHIPS.

MORTALITY DATA RETRIEVAL

A list of all deaths older than 15 years coded as ICD 335 in position 1 or 2 on the death certificate, was retrieved by the Registrar General's Office for Scotland approximately one year after the end of the year of interest. This information enabled the retrieval of general practice records, which contain a copy of all hospital correspondence, through the relevant health board. This information was treated similarly to that derived from SHIPS (see above): the SMNDR records were searched to determine the false negative rate of retrieval for mortality statistics.

Results

MORBIDITY DATA

A summary of the SHIPS data in relation to the SMNDR is given in table II. Of a total of 379 patients older than 15 years discharged for the first time in 1989–90, 141 were already registered with the SMNDR and 27 were known to be prevalent from other sources, leaving 211 in whom further diagnostic details were sought from hospital records. Altogether 112 of these did not have MND; that is, the false positive rate was 30%. Fifty two MND patients (23% of incident cases) did not appear in the SHIPS in the two years examined. SHIPS overestimated the incidence on MND by a factor of 1·6 (379/229). Table III appraises SHIPS in terms of sensitivity (84%) and positive predictive value (70%) of diagnosis of MND as determined by ICD 335. Given the very large number of true negative discharges in respect of ICD-335 in Scotland an hypothetical specificity would be close to 100%.

Table IV gives the diagnosis in the 112 patients miscoded as MND. The largest group were patients with a pseudobulbar palsy caused by

Table II Summary of the Scottish Hospital In-Patient Statistics (SHIPS) in relation to the Scottish Motor Neuron Disease Register (SMNDR)

	Discharge or diagnosis date		
	1989	*1990*	*Total*
Discharged from Scottish hospitals for the first time, coded ICD 335 and aged >15 years:	171	208	379
Already registered with SMNDR as diagnosed on or after 1.1.89 (incident)	51	90	141
Already known to SMNDR as MND diagnosed before 1.1.89 (prevalent)	9	18	27
Hospital records reviewed for patients not known to the SMNDR showed:			
Coded correctly but notes indicated prevalent to SMNDR	34	14	48
Diagnosis MND (first notification source for SMNDR)	26	23	49
Diagnosis not MND (false positive, see table IV)	50	62	112
Records could not be located	1	1	2
Total records reviewed	110	99	209
Therefore SHIPS correct in diagnosis, whenever this made (true positive)	120	145	265
Diagnosis of MND, no hospital discharge in either 1989 or 1990, not on SHIPS*	10	32	42
Discharged from hospital but not on SHIPS	7	3	10
Total, ie definite or probable MND, on SMNDR, but not on SHIPS (false negative)	17	35	52
Incident patients as defined by the SMNDR ('gold standard')	111	118	229

*Diagnosis of MND made as an outpatient.

Table III Sensitivity and positive predicitive value of a diagnosis of motor neurone disease (MND) as determined by the Scottish hospital In-Patient Survey (SHIPS) compared with the Scottish Motor Neuron Disease Register (SMNDR), 1989–90.

	SMNDR		
	MND	*Not MND*	*Total*
SHIPS:			
ICD 335, 1989–90	265	112	377*
Not ICD 335	52	†	
Total	317	†	

*Two records missing.
†The presumed but unmeasured high true negative rate would produce a specificity close to 100%
Sensitivity of SHIPS=true positive/true positive+false negative×100=84%
Positive predictive value of SHIPS=true positive/true positive+false positive×100=70%

cerebrovascular disease (38%). In 18% of patients the coding accurately reflected the medical summary but the diagnosis was incorrect, either because of a failure to meet standard criteria for diagnosis or because a 'probable' or 'possible' diagnosis of MND was disproved at follow up. Twelve per cent of miscoding seemed to be the result of a transcription error, as no possible source of confusion for ICD 335 could be determined from the records.

MORTALITY DATA

Summary statistics relating the death certificates coded as ICD 335 to the SMNDR are given in table V. A total of 27 of 281 deaths (10%) contained false positive errors giving a positive predictive value for mortality returns of 90%. Again, miscoding of patients with pseudobulbar or bulbar palsy was the most common source of error. Twelve (44%) of these errors were the result of misdiagnosis, although the coding was correct; 15 (56%) were caused by the miscoding of other conditions (table VI).

The records of seven of 95 SMNDR patients who died as a result of this disease were not retrieved by the Registrar General, giving a false negative rate of 6%. Inspection of these returns however, showed that MND was recorded as a cause of death in three cases, so presumably there was a transcription error at some point in retrieval. Until the death rate of patients on the SMNDR reaches a steady state we are unable to calculate the sensitivity of a death certified as MND.

Discussion

MORBIDITY DATA

Forty eight of 229 (21%) incident patients were identified by SHIPS as the first source of referral for the SMNDR and therefore this resource was necessary for us to determine accurately the incidence of MND in Scotland.

We have identified a serious inaccuracy in SHIPS. Altogether 30% of patients coded as ICD 335 did not have MND and therefore these data cannot, in their crude form, be used as a reliable guide to the incidence of MND in Scotland. The 1·6 fold incidence overestimate by SHIPS for 1989–90 would improve, however, if this study were to be repeated over a longer period in the future as there would be fewer prevalent cases ascertained by SHIPS in relation to the SMNDR. The problem of false positive coding, however, will persist unless efforts are made to improve accuracy.

There are three major reasons for these errors. Firstly, while the importance of accurate coding *may* be appreciated by many doctors, this task seems to receive a low priority. Although medical staff make the diagnosis, they do not take primary responsibility for the supervision of diagnostic coding, which is delegated to clerical staff who may not be able to interpret adequately the nuances of ambiguous medical terminology. In some cases records staff are left extracting information from poorly worded letters rather

Table IV Classification of 112 patients miscoded as motor neurone disease (MND) by the Scottish Hospital In-Patient Statistics

	1989	*1990*	*Total (%)*
Diagnosis when coding was incorrect:			
Pseudobulbar palsy caused by:			
(a) Cerebrovascular disease	23	20	43 (38%)
(b) Multiple sclerosis	1		1
Bulbar palsy (eg carcinoma of lung with vocal cord paresis; dysphagia caused by oesophagel carcinoma; multiple cranial neuropathies; myasthenia gravis; unspecified, non-progressive)	5	7	12
Neuromuscular disease, usually with 'neuron' in diagnosis eg 'lower motor neurone weakness', "motor neuropathy"	3	12	15
Muscular atrophy as a result of polio	4	2	6
Not MND but physician refused release of further information	1		1
Coding correct, but insufficient medical evidence for MND	4	16	20 (18%)
No reason apparent for the error in coding (transcription error):			(12%)
Neurological disease (but MND not mentioned)	5	5	10
Non-neurological disease	4		4
Total	50	62	112

Table V Summary statistics relating to 1989-90 death certificates coded as MND ICD 335 (position I or II)

	1989	*1990*	*Total*
Death certificates coded as ICD 335 (as retrieved by the Registrar General)	148	133	281
Health board records reviewed:			
Died, known to have MND, Prevalent to SMNDR	61	46	107
Died, known to SMNDR, incident	21	64	85
Further records for review	66	23	89
Unable to trace records	3	0	3
Death certificate coded as ICD 335 but diagnosis incorrect or insufficient evidence for diagnosis (false positive)	14	13	27 (10%)
True positive (281–27)			254
Registered with SMNDR, died 1989–90 but not retrieved by Register House (false negative)	2	5	7* (6%)
Incident patients 1989–90 (SMNDR)	111	118	229
SMNDR patients who died	26	69	95

SMNDR=Scottish Motor Neuron Disease Register.
*Three of these patients had MND on the death certificate in position I and therefore are an error of retrieval rather than misdiagnosis. Positive predictive value of mortality certification as ICD 335: 281–27/281×100=90% The true positive death rate (as judged by SMNDR) is not yet stable to allow calculation of sensitivity.

than specifically listed discharge diagnoses and hence take medical conditions out of context. Some patients, for example with 'pseudobulbar palsy due to cerebrovascular disease' were assigned to the wrong ICD or to both ICD 335·2 and 437·9.

Secondly, the ICD classification does not allow practitioners to indicate whether a diagnosis is firmly established, requires further information, or is largely speculative. This is in contrast to clinical practice where revision is often required and the passage of time clarifies a diagnostic problem (for example, the differential diagnosis of spondylotic cervical myelopathy and MND). It is this flaw, when the diagnosis did not meet generally accepted minimal criteria for probable or definite MND,[9] which accounted for 18% of the diagnostic inaccuracy (table IV).

Finally, some of the errors arose because coding with the ICD-9 can be ambiguous; for example the alphabetical listing directs 'bulbar (progressive) (chronic)' and 'bulbar-pseudo (not elsewhere classified)' to ICD-335·2, which may be inappropriate.

Although the accuracy of a general sample of morbidity information[11] and for a number of other specific diseases in Scotland[12–15] has previously been examined, and while many epidemiological studies of MND have collected hospital discharge data as part of a case finding exercise,[16–19] some of whom have commented on their inaccuracy,[20] none have formally evaluated their utility for this disease. Neither have previous studies been able to compare accuracy with an independent case finding method such as the SMNDR.

Our results indicate that more diligent efforts to ensure correct coding are required. The nature and uses of SHIPS should be made clear to the medical staff responsible for discharge documents. Doctors should be encouraged to use clear and unambiguous terminology, preferably within a standardised discharge format. Guidelines issued to area medical committees in 1990 have emphasised that a senior member of the clinical team should be responsible for coding.[21] Regular audit of case records is important in this regard and quality assurance within the Information and Statistics Division has now been established whereby experienced staff will blindly recode a random sample of SMR records to quantitate the accuracy of existing coding. As a result of this review sources of error will be discussed with coding staff and we will continue to monitor SHIPS returns to see if accuracy improves.

Table VI Errors in mortality data (n=27) with respect to adult onset motor neurone disease (MND). Diagnosis shown is based on review of medical records and death certificates

(1) bulbar or pseudobulbar palsy as a result of cerebrovascular disease, multiple sclerosis, or unspecified (non-progressive)	11
(2) Motor neuropathies, radiculopathies, 'lower motor neurone weakness'	5
(3) Multisystem neurological disease	2
(4) Probable cervical spondylosis	1
(5) Diagnosis unknown or uncertain but insufficient evidence for MND	8

Fifteen did not have MND in the death certificate but were coded ICD 335; 12 had MND on the death certificate (ie coding correct) but MND misdiagnosed.

MORTALITY DATA

Interpretation of variations in time and place which might be used to generate aetiological hypotheses depends on an understanding of the accuracy of the data on which these comparisons are based. In this study, mortality data were much more accurate than SHIPS, as might be predicted. Some of the problems relating to discharge data, however, also apply to death certificate coding, in particular the miscoding because of other neurological conditions, (mainly bulbar palsy of various causes), which is in turn the result of difficulties with the ICD coding system.

Whether the incidence of MND can be studied adequately with mortality data alone is controversial.[22] Other studies of the accuracy of mortality data have shown that 70–90% of patients diagnosed as having MND have this condition recorded on their death certificate.[4 5 7 8] In Scotland mortality data also have a low false negative rate. Only one other study from Japan has examined the false positive rate of death certificate coding (in 1965); an extensive attempt to verify the diagnosis from other sources showed that as many as one third of men were coded as dying of MND without having the disease, but the accuracy was better for women.[8] Our mortality statistics were much more accurate than this.

These findings have important implications for epidemiologists interpreting trends in MND frequency. There is evidence from a number of countries that the mortality from MND has increased steadily over the past 30 years[5 23 27] and this is also the case in Scotland.[28] This increase may be real, rather than a result of ascertainment bias or improved diagnostic skills, but it would be helpful to confirm this with other sources such as incidence data.

From this analysis, it can be inferred that changes in the ICD coding system or alterations in coding practice are likely to have an effect on both morbidity and mortality rates, which could be mistaken for a real variation in disease incidence. Whether these results are applicable to other countries is uncertain but the anomalies in ICD-9 will apply everywhere.

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Other participants in The Scottish Motor Neuron Disease Research Group are: **Aberdeen**—Dr J E C Hern, Dr R Knight; **Dundee**—Dr D Davidson, Dr A Forster, Dr R Roberts; **Edinburgh**— Dr B Ashworth, Dr R E Cull, Dr E H Jellinek, Dr S M Holloway, Dr A McInnes, Dr B Pentland, Dr P A G Sandercock, Dr R Will; **Glasgow**—Dr J P Ballantyne, Professor P O Behan, Dr I Bone, Professor F I Caird, Dr I Draper, Dr W F Durward, Dr G Jamal, Professor P Kennedy, Dr R Metcalfe, Dr M Thomas, Dr A I Weir; **Inverness**—Dr L R Fisher.

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