



Estimating prevalence of neurological disorders in Scotland from the Scottish Burden of Disease study.

Briefing prepared for Neurological Advisory Committee on Neurological Conditions, by the Scottish Burden of Disease Study Team.

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National Advisory Committee on Neurological Conditions briefing: Estimating prevalence of neurological disorders in Scotland from the Scottish Burden of Disease study.

A. Background

Burden of disease (BoD) studies aim to estimate the total amount of illness, injury, disability and early death affecting a country at a specific point in time. To do this, they use a single measure which combines fatal burden and non-fatal burden. The combined measure is called the disability-adjusted life year (DALY). In the [Scottish Burden of Disease \(SBoD\)](#) study, we have calculated the burden of disease (i.e. DALYs) for 132 disease and injury categories, as defined by the international Global Burden of Disease Study (GBD).^{1, 2}

To calculate the non-fatal burden in BoD studies requires estimates of prevalence for all conditions used in the BoD disease classification. In GBD, these estimates are generated through modelling based on meta-regression analyses generating synthetic estimates of the disease burden in Scotland. Scotland, however, has excellent health management systems that potentially allow us to make our own estimates of prevalence, based on counts of people we can identify as having different diseases, conditions or injuries. To get these data for the whole of Scotland, we extracted information from hospital, GP and prescribing recording systems, and also disease registers. Where necessary, we also used information from surveys, research studies or expert-informed estimates.

This briefing, prepared for the Scottish Government's National Advisory Committee on Neurological Conditions, describes the methods used by SBoD team to estimate prevalence for the neurological conditions included in the study and compares these estimates with those from the GBD study, as well as other available prevalence estimates for the selected neurological conditions.

B. Classification of neurological disorders in SBoD

We have adopted the GBD2016 disease classification which includes a neurological disorders classification. This grouping is comprised of eight neurological disorders:

- Alzheimer's disease and other dementias
- Parkinson's disease
- Epilepsy
- Multiple sclerosis
- Motor neuron diseases
- Migraine
- Tension-type headache
- Other neurological disorders

Table 1 in Appendix provides the GBD case definitions for these conditions, with the accompanying ICD classification.

The classification of 'other neurological disorders' includes many diverse types of disorders (e.g. cerebral palsy (and other paralytic syndromes), neuropathies, Huntington disease, hereditary ataxia) with a range of severities and associated sequelae e.g. cerebral palsy has multiple sequelae and many diverse causes and in burden of disease studies the burden is distributed to a number of other diseases/conditions e.g. meningitis; encephalitis; neonatal preterm birth complications. Because these neurological disorders are diverse in their underlying causes and risk factors as well as in their associated health outcomes, GBD argue that that modelling them together would not produce reliable estimates of prevalence or excess mortality.³ In GBD and SBoD, the burden for 'other neurological disorders' is modelled based on premature mortality. Further information on this method can be provided.

C. Methods

Estimates for Alzheimer's and other dementias and Parkinson's disease.

To estimate prevalent cases of dementia and Parkinson's disease in 2016, the Practice Team Information dataset (PTI) was used. This dataset was collected by ISD Scotland from April 2003 to September 2013. It includes information from a nationally representative 5% sample of Scottish General Practices regarding face-to-face consultations between individuals and a member of the practice team (GPs, nurses and clinical assistants). The presence of a unique patient-identifier on the dataset allows for the grouping of consultations for each individual. The reason for each consultation was coded using Read codes].

The number of individuals that had a Read code specific to dementia and Parkinson's were used to estimate prevalence, for the period 1 April 2003 and 31 September 2013. READ codes were selected based on mapping to READ from ICD10 codes in the GBD disease classification (a full list of READ codes used in analysis is available on request):

- Dementia: F00-F03.91, F06.2, G30-G31.1, G31.8-G32.89
- Parkinson's disease: G20-G20.9

Individuals were counted once in any year in which they attended their GP and consulted for. We projected the estimated annual incidence trends, censoring for mortality, for the time period (2003-2013) to estimate the number of prevalent cases in 204, 2105 and 2016. Vital status for individuals was not available, so adjustments to account for deaths were made using age-sex specific excess mortality rates for dementia and Parkinson's as defined in GBD 2016 (see Appendix, Table 2 and 3).³

Estimates for epilepsy, multiple sclerosis, motor neuron disease

To estimate prevalent cases for each of the above conditions in 2016, the Scottish Morbidity Records 01 (SMR01) was used. This dataset contains structured data in the form ICD-10 codes relating to diagnoses made on discharge from general and acute hospitals during inpatient episodes and day cases. There are up to six individual ICD-10 codes that can be recorded, where the primary diagnosis relates to the main reason for the hospital episode of care, and the other secondary diagnoses

refer to co-morbidities that may affect care during that hospital episode of care. The SMR01 dataset has a CHI number attached to the hospital episode of care, which allows for the identification of records for an individual. This CHI number has been linked to records from the NRS register of deaths, to exclude individuals that have died from prevalence estimates that relate to a period following their date of death.

The number of individuals that had a diagnosis of epilepsy, multiple sclerosis or motor neuron disease, from 1 January 1996 to 31 December 2016 was used to estimate the number of prevalent cases. Individuals were identified using the following ICD10 codes:

- Multiple sclerosis G35.
- Motor Neuron Diseases G12.
- Epilepsy G40., G41

Estimates for Headache disorders (Migraine and Tension type headache)

Due to the poor recording of these disorders in health administrative records, SBoD have used prevalence estimates for Scotland from the Global Burden of Disease study to estimate the burden of headache in Scotland. GBD prevalence estimates are derived from a Bayesian meta-regression method.³ GBD used data from approximately 120 studies identified from a systematic review of the literature to estimate the headache burden.

D. Estimating prevalence of neurological conditions: results

The table below provides prevalence estimates for neurological conditions in the SBoD disease classification. A comparison with GBD modelled estimates is also provided.

Condition	SBOD prevalence	GBD prevalence (95% CI) ^a	GBD prevalence (95% CI) ^a
Alzheimer's disease and other dementias	66,300	72,300	60,726-85,589
Migraine	As per GBD	1,075,029	1,026,340-1,124,514
Tension Type headache	As per GBD	1,400,722	1,253,770-1,558,924
Epilepsy	43,100	20,177	4,643 - 34,545
Multiple sclerosis	9,700	12,232	11,083 - 13,352
Parkinson's disease	12,600	9,200	7,182 -11,580
Motor neurone disease	730	1,103	1,009 -1,204

^a <http://ghdx.healthdata.org/gbd-results-tool>: downloaded 11 April 2018

D1: Comparisons with other studies

Alzheimer's and other dementias

We have estimated a prevalence of approximately 66,300 in 2016. Alzheimer's Scotland estimate a prevalence of approx 90,000 in Scotland in 2017 ⁴, based on two sets of prevalence rates (EuroCoDe (2009) study for ages 60+ and Harvey et al (1998)² for the under 60 age group.⁵ Alzheimer UK published prevalence estimates in 2014 using the DELPHI method which estimated that there were 66,773 people living with dementia in Scotland in 2013. ⁶

It is estimated that dementia is under recorded in primary care settings in Scotland by 26%.⁷ Applying this correction factor to our estimate would give a prevalence population of approximately 84,000.

Epilepsy

We have estimated a prevalence of approximately 44,000 (8.0 per 1000) in 2016, compared to GBD estimate of approximately 21,000 (3.9 per 1000). Our estimate is similar to that recently published by Weatherburn et al (2017) based on primary care records in Scotland: 8.4 per 1000.⁸

The Joint Epilepsy Council estimated prevalence of 54,000 (10.0 per 1000) in Scotland in 2010 based on primary care and prescription data.⁹

Multiple sclerosis

We have estimated a prevalence of approximately 9,700 in 2016 (1.8/1000 population), slightly lower than the GBD estimate of 12,000 (2.2/1000 population).

The most recent estimate of MS prevalence in Scotland was published by the Multiple Sclerosis Society. Their study, based on research carried out by Mackenzie et al (2013)¹⁰, estimated that there were 11,310 people (2.1/1000 population) living with MS, in Scotland, in 2016.¹¹

Parkinson's disease

We have estimated a prevalence of approximately 12,600 in 2016 (3.0/1000 population aged 20 years and over). Parkinson's UK recently estimated, using primary care data, a prevalent population of approximately 11,500 in Scotland in 2015 (2.7/1000 population, aged 20 years and over, increasing to approximately 12,500 in 2018).¹² These compare to prevalence estimates from the GBD study of around 9,000 prevalent cases (2.2/1000 population, 20 years and over).

Pringsheim et al 2014, in a meta analysis of 47 studies, estimated prevalence of Parkinson's disease in Europe between 2.8 and 3.1 per 1000 population.¹³

Motor neuron diseases (MND)

We have estimated a prevalence of approximately 700 in 2016 (13.5/100,000 population) compared to GBD modelled estimate of approximately 1100 (20.4/100,000 population).

There is little published research describing prevalence of MND. Current estimates of MND prevalence in Scotland suggest between 300-450 people living with MND in Scotland at any given time (5.5 to 7.4 per 100,000).¹⁴ Imam et al (2010) estimated a standardised point prevalence of MND of 5.66 per 100,000 (95% CI 4.49-6.83) in the south west of England. Applied to Scotland, this would give an estimated prevalence of approximately 300 (240 -370).¹⁵

In the GBD study, Motor neuron diseases are defined as “a set of chronic, degenerative, and progressive neurological conditions typified by the destruction of motor neurons and the subsequent deterioration of voluntary muscle activity. The ICD-10 code corresponding to motor neuron diseases is G12.³ For comparability with GBD, we have used the same ICD classification for MND. It should be noted, however, that this includes ICD10 code G12.2 "Progressive Bulbar Palsy" which the SBoD team have been advised is not specific to MND, often being a consequence of other neurodegenerative or neurological diseases.

D2: Estimating prevalence of ‘other neurological disorders’

It is possible to extend our method to generate prevalence estimates for individual conditions included in the classification of ‘other neurological disorders’. To illustrate this, we have carried out some provisional analysis to estimate prevalence for three conditions (cerebral palsy, hereditary ataxia and Huntington’s disease) included in the GBD classification of ‘other neurological disorders’.

To estimate prevalent cases for these conditions, hospital admission data was used, as previously described, for epilepsy, multiple sclerosis and motor neuron diseases. The ICD codes used in the analysis are provided below. **Please note that these represent initial estimates as further work and consultation would be essential to develop and validate the disease model applied.**

Estimates:

- 550 prevalent cases of **Huntington's disease** (G10) in 2016. This is equivalent to 12.8/100,000 population aged >21yrs and over. Evans et al (2013) using primary care records, estimated a UK prevalence, in 2010, of 12.3/100 000 population aged>21 yrs and over.¹⁶
- 1100 (20.2/100,000) prevalent cases, in 2016, of **hereditary ataxia** (G11 which includes congenital non-progressive ataxia, early-onset cerebellar ataxia, late-onset cerebellar ataxia, cerebellar ataxia with defective DNA repair (excl.: Cockayne syndrome, xeroderma pigmentosum) , hereditary spastic paraplegia, other hereditary ataxias and unspecified hereditary ataxia).

Ataxia UK estimate that, In the UK, at least 10,000 adults and 500 children with progressive ataxia.¹⁷ Muzaimi et (2004) estimated a minimum prevalence of 10.2 in 100,000 adults with late onset cerebellar ataxia only in South Wales.¹⁸

- 5000 prevalent cases, in 2016, of **cerebral palsy** (G83 which includes spastic quadriplegic cerebral palsy, spastic diplegic cerebral palsy, spastic hemiplegic cerebral palsy, dyskinetic cerebral palsy, ataxic cerebral palsy, other cerebral palsy and unspecified cerebral palsy).

Using hospital admission data is likely to underestimate the prevalence of cerebral palsy in Scotland but by how much is unclear. Based on a cerebral palsy prevalence rate of 2.6 per 1000 live births²² , we can speculate that we are underestimating cerebral palsy prevalence by a least 25%.

Recent studies have also shown that using hospital registers could underestimate prevalence of cerebral palsy in children by between 2 and 20%.^{19 20 21} Relying on the accuracy of coding of cerebral palsy in hospital records is also problematic and subject to miscoding and misclassification. Hollung et (2016)²⁰ reported that over 10% of children in their cohort had been wrongly diagnosed with cerebral palsy on their hospital record but also reported that some of their older children in

their cohort (i.e. born from 1996 to 2001) with mild CP did not have a diagnosis of cerebral palsy on their hospital record.

Comment

We have presented prevalence estimates for a number of neurological conditions based on SBoD methods. Due to the way prevalence is calculated as part of the burden of disease study, it is not possible to estimate the unique number of people in Scotland at any given point with a neurological condition. This is because we use a variety of datasets to estimate our prevalence with only some of these being linked at an individual level.

When considering prevalence estimates from SBoD , it is also important to remember that the total 'disability' calculated for diseases/conditions in burden of disease studies is a combination of both prevalence and severity weighed to reflect the degree of severity of the disease/condition.

Future work on the SBoD study will attempt to refine the estimates of prevalence. These improvements are partly dependant on exploring other data sources and reviewing evidence from high quality research that it is relevant to Scotland. Please contact the SBoD project team (nhs.healthscotland-sbod-team@nhs.net) for enquiries and suggestions on how to improve our estimates.

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Appendix

Table 1: Neurological conditions included in SBoD, case definition and ICD10 codes

Level 3 condition	GBD case definition	ICD 10 codes
Dementia	"...a progressive, degenerative, and chronic neurological disorder typified by memory impairment and other neurological dysfunctions".	F00, F01, F02, F03, G30, G31
Parkinson's	"... a chronic, degenerative, and progressive neurological condition typified by the loss of motor mobility and control – most notably tremors".	G20, G21, G22.
Multiple Sclerosis	"... a chronic, degenerative, and progressive neurological condition typified by the damaging of the myelin sheaths".	G35.
Motor Neuron Disease	"... are a set of chronic, degenerative, and progressive neurological conditions typified by the destruction of motor neurons and the subsequent deterioration of voluntary muscle activity".	G12.
Epilepsy	used the following definitions from the "Guidelines for Epidemiologic Studies on Epilepsy": 1) Epilepsy: a condition characterized by recurrent (two or more) epileptic seizures, unprovoked by any immediate identified cause, and 2) "Active" epilepsy: a prevalent case of active epilepsy is defined as a person with epilepsy who has had at least one epileptic seizure in the previous five years, regardless of antiepileptic drug (AED) treatment	G40., G41.
Migraine	"Migraine is a class of disabling primary headache disorders, characterized by recurrent unilateral pulsatile headaches. The two major subtypes are common migraine (without aura) and classic migraine (with aura or neurological symptoms). GBD do not distinguish subtypes as most epidemiological studies report on overall migraine only"	G43
Tension type headache	Tension-type headache (TTH) is characterized by a dull, non-pulsatile, diffuse, band-like (or vice-like) pain of mild to moderate intensity in the head, scalp, or neck.	G44.2
Other neurological conditions	"..., there are many diverse types of neurological disorders with a range of severities and associated sequelae. Because these neurological disorders are diverse in their underlying causes and risk factors as well as in their associated health outcomes, modelling them together ... would not produce reliable estimates of prevalence or excess mortality".	n/a

Table 2 Predicted excess mortality ratio values for dementia by age and sex (95% CI)

	Male	Female
Age 40-59	0.008 (0.007 - 0.01)	0.008 (0.007 - 0.009)
Age 60-64	0.016 (0.014 - 0.019)	0.016 (0.013 - 0.018)
Age 65-69	0.021 (0.017 - 0.024)	0.02 (0.017 - 0.023)
Age 70-74	0.029 (0.024 - 0.034)	0.027 (0.023 - 0.032)
Age 75- 80	0.042 (0.035 - 0.049)	0.04 (0.034 - 0.047)
Age 80-84	0.058 (0.049 - 0.068)	0.055 (0.047 - 0.064)
Age 85-89	0.088 (0.074 - 0.104)	0.084 (0.07 - 0.098)
Age 90-94	0.147 (0.124 - 0.174)	0.14 (0.118 - 0.165)
Age 95+	0.226 (0.19 - 0.268)	0.216 (0.183 - 0.254)

Table 3 Predicted excess mortality ratio values for Parkinson's disease by age and sex (95% CI)

	Male	Female
Age 40-59	0.005 (0.004 - 0.006)	0.004 (0.003 - 0.005)
Age 60-64	0.01 (0.008 - 0.013)	0.008 (0.006 - 0.01)
Age 65-69	0.016 (0.013 - 0.02)	0.013 (0.01 - 0.016)
Age 70-74	0.028 (0.023 - 0.035)	0.023 (0.018 - 0.029)
Age 75- 80	0.047 (0.037 - 0.059)	0.037 (0.029 - 0.046)
Age 80-84	0.071 (0.055 - 0.089)	0.057 (0.045 - 0.07)
Age 85-89	0.093 (0.073 - 0.117)	0.076 (0.06 - 0.093)
Age 90-94	0.126 (0.099 - 0.155)	0.101 (0.08 - 0.127)
Age 95+	0.154 (0.122 - 0.191)	0.123 (0.097 - 0.153)