Glover, G., Williams, R., Heslop, P., Oyinola, J., & Grey, J. M. (2017). Mortality in people with intellectual disabilities in England. *Journal of Intellectual Disability Research*, *61*(1), 62–74. https://doi.org/10.1111/jir.12314

JIDR

Mortality in people with intellectual disabilities in England

Journal:	*Journal of Intellectual Disability Research*
Manuscript ID	Draft
Manuscript Type:	Original Manuscripts
Keywords:	Intellectual Disability, Mortality, Life Expectancy, International comparisons, Cause of Death, England, population research

Mortality in people with intellectual disabilities in England

Abstract

Background

People with intellectual disabilities (ID) die at younger ages than the general population, but nationally representative and internationally comparable mortality data about people with ID, quantifying the extent and pattern of the excess have not previously been reported for England.

Method

Data from the Clinical Practice Research Datalink database (CPRD GOLD September 2015) identified people registered with several hundred participating general practices and diagnosed by their GP as having ID. This covered roughly 5% of the population of England over the period studied, April 2010 to March 2014. Linkage to national death certification data allowed us to derive linked population and mortality data for people with and without ID, overall and by cause.

Results

Mortality rates for people with ID were significantly higher than for those without. The all-cause standardised mortality ratio (SMR) associated with ID was 3.18. Life expectancy at birth was 19.7 years lower than for people without ID. Circulatory and respiratory diseases and neoplasms were the three most common causes of death. Potentially preventable causes included epilepsy (3.9% of deaths) and aspiration pneumonitis (3.6% of deaths). Avoidable mortality analysis showed a high proportion of deaths from causes classified as amenable to good medical care but fewer from preventable causes compared to people without ID. International comparison to

areas for which data have been published in sufficient detail for calculation of directly standardised rates suggest England may have higher death rates for people with ID than areas in Canada and Finland, and lower death rates than Ireland or one US state.

Conclusions

National data about mortality in people with ID provides a basis for public health interventions. Linked data using GP records to identify people with ID provides comprehensive population-based monitoring, unbiased by the circumstances of illnesses or death. However GPs in England currently identify only around 0.5% of the population as having ID, suggesting that individuals with mild, non-syndromic ID are largely missed. Some specific causes identified suggest control of cardiovascular risk factors, epilepsy and dysphagia and screening for colorectal malignancy are important areas for health promotion initiatives.

Mortality in people with intellectual disabilities in England

Background

Publicity about deaths of individuals with intellectual disabilities (ID) and studies of deaths in comparatively small areas of England has brought attention to the major disparities faced by this population.(Mencap 2007; Heslop et al. 2014; Mazars 2015) However as yet, no nationally representative population based study has been published. There have been recent studies of specific localities, some including population-based analyses (Heslop et al. 2014; Tyrer et al. 2007; Tyrer & McGrother 2009) but the only fully national study, based on death certification data, was limited by evidently incomplete identification of relevant deaths and a lack of detailed corresponding population data.(Glover & Ayub 2010) There is currently no routine, national statistical source in England documenting the overall extent of excess mortality in people with ID, or its pattern in respect of causes or demographic sub-groups.

Analysis of mortality data can help to document some of the burden of conditions that are potentially fatal, the extent of consequent health inequalities and the effects of some public policies for population subgroups.(Lauer et al. 2015) However there is no single source where all relevant data are collected. UK death certificates only include recording of conditions considered to be associated with the death. Primary care notes, the only set of records with the scope to include details of all medical conditions for individuals, will not necessarily record causes of deaths unless the certificate is issued by the general practitioner (GP). Linkage between the two is needed.

The Clinical Practice Research Datalink is a governmental, not-for-profit research service, jointly funded by the NHS National Institute for Health Research (NIHR) and the Medicines and Healthcare products Regulatory Agency (MHRA), a part of the Department of Health. It is based on an established anonymised research database drawing clinical records regularly from participating general practices.(Khan et al. 2010) These are routinely linked at individual person level to mortality and health service records.

Using this source we set out to identify the rates and patterns of mortality of people living in England and identified by their GP as having ID in relation to age, sex and causes of death. We also explored the feasibility of analyses in relation to minority ethnic groups. We compared English findings to those of other large population-based studies using a variety of data collection or linkage approaches which had reported sufficient detail to permit comparisons using directly standardised rates. We found relevant reports from Australia, Canada, Finland and Ireland as well as an earlier study describing an English county.(Florio & Trollor 2015; Ouellette-Kuntz et al. 2015; Arvio et al. 2016; McCarron et al. 2015; Tyrer & McGrother 2009)

Methods

Study population

We used the Clinical Practice Research Datalink database (CPRD GOLD September 2015), taking data for all patients registered between 1st April 2010 and 31st March 2014 for one day or more with a general practice that met data quality and linkage requirements, and for whom the data needed for linkage to mortality records was complete. We identified patients as having ID if they had a record of one of the set of

Read codes used nationally to define GPs' learning disability registers, or any of a number of other diagnoses, such as Down's syndrome, reliably associated with ID.(NHS Primary Care Commissioning 2012) A full list of the codes used is available from the authors.

Study data

Study subjects were grouped by ID status, sex, and roughly decennial age groups (0, 1-9, 10-17, 18-24, then ten year bands to 84 and 85-99), the region in which their GP's practice was located and their ethnic group where this was known. To provide securely anonymised data for detailed study, RW and JO at the MHRA provided tabulated counts of registered patient-days and deaths in the study time-window by all these groupings, for each of the four administrative years covered. Several versions of the deaths tabulations were provided with groupings to ICD10 chapters, to three-character ICD10 codes for common individual causes and the four chapters responsible for the largest numbers of deaths (2% or more of all deaths of people with ID), and for the groups of causes considered to represent 'avoidable' causes of death in relevant age groups.(Office for National Statistics 2015) Comparison of English death rates to those for regions of other countries used age/sex specific data from the published accounts (cited above).

Data analysis

Rates, directly standardised rates and (indirectly standardised) standardised mortality ratios (SMRs) were calculated as described by Eayres for the English Public Health Observatories.(Eayres 2008) For comparisons of all-cause mortality with Australian, Canadian, Finnish, Irish, and US data as well as an earlier study

from a part of England, we used direct standardisation for age to the 2013 European standard population.(Eurostat 2013) Calculations were undertaken using Microsoft Excel and Access with customised statistical routines written in Visual Basic. Life expectancy was calculated using a spreadsheet tool developed by English Public Health Observatories.(Eayres 2004)

Results

Population

---------- Table 1 about here ----------

Altogether 11·16 million person years were covered by the study, an average of 2·79 million people in each year or 5·22% of the population of England at the time as estimated by the Office for National Statistics (ONS).(Office for National Statistics 2013) Table 1 shows the breakdown by each of the covariates studied and the prevalence of ID identified in each subgroup. Comparison with mid-2012 population estimates from ONS suggests the age profile is representative of the English population. Coverage varied substantially between Strategic Health Authority (SHA) areas reflecting patterns of use by GPs of the information system on which CPRD is based, with Yorkshire and Humber, the East Midlands, and the North East under-represented and the South East Coast, South Central and South West over-represented.

---------- Figure 1 about here ----------

The overall proportion of people with an identifiable ID was 0·53%. Figure 1 shows population pyramids for people with and without ID. The pattern for people with ID shows a narrower base, indicating that ID is often not definitely recorded until some years into a child's first decade. Attrition in adulthood is also quicker with the pyramid for people with ID collapsing more quickly after the age of 54.

Number of deaths, death rates and life expectancy

The study identified 98,035 deaths, 5·28% of the total number of deaths registered in England in the period. Table 1 shows their distribution between the covariates studied. Of the 664 deaths of people with ID, 371 (55.9%) were of males, 293 (44.1%) of females.

The crude death rate for those with identifiable ID was 11.2 deaths per 1000 population per year (95% confidence interval (CI) 10.4 to 12.1), 1·27 times the crude death rate for others (8·8 deaths per 1,000 per year – 95% CI 8.7 to 8.8).

---------- Table 2 about here ----------

Death rates were higher in those with ID at all ages for both sexes, significantly so in all cases except males aged 18 to 24, and 85 and older (Table 2). Generally, the difference was more marked for females and in younger age groups, although the actual numbers of deaths of younger people with ID was relatively small.

The SMR for people with identifiable ID was 3·18 (95% CI 2·94 to 3·43); it was higher for females (3·40 - 3·02 to 3·81) than for males (3·03 - 2·73 to 3·35). Life expectancy at birth for people with ID was 65·5 years (95% CI 61·9 to 69·2), 63·8 years (57·7 to 69·9) for males and 66·7 (63·4 to 70·0) for females. Corresponding figures for people without ID were, for persons 85·3 (85·2 to 85·4), for males 83·6 years (83·4 to 83·7), and for females 86·9 (86·8 to 87·0). This represents a shortfall for people with ID of 19·7 years, for males 19·7 and for females 20·2.

We were unable to do a satisfactory regional analysis because of the wide variations in sample sizes. Yorkshire & The Humber and the East Midlands both appeared to have noticeably lower SMRs, but the small sample sizes in these areas gave these excessively wide confidence intervals rendering these differences non-significant. The South East Coast had an SMR for people with ID that was higher, than that for the whole country, though not statistically significantly so (3.89, 95% CI 3.16 to 4.79).

Twenty-four deaths (3.6%) were of individuals with ID from minority ethnic groups, 93 (14%) of individuals of unrecorded ethnicity. No single identified ethnic group accounted for more than four deaths, thus we were unable to draw any conclusions about patterns by ethnicity.

Causes of death

---------- Table 3 about here ----------

For people with ID, in all but one of the eleven ICD10 chapters where five or more deaths were recorded, the SMR was significantly above unity (Table 3). SMRs greater than 4 with the lower bound of the 95% confidence interval greater than 2 were seen for seven chapters. There was no chapter for which significantly fewer than expected deaths of people with ID were recorded.

Conditions listed in the ICD chapter covering diseases of the circulatory system were responsible for the largest number of deaths of people with ID. Most prominent were ischaemic heart disease (57 deaths, expected 26·4, SMR 2·2, 95% CI 1·6-2·8), cerebrovascular disease (39 deaths, expected 11·8, SMR 3·3, 95% CI 2·3-4·5), phlebitis and thrombophlebitis (10 deaths, expected 1.5, SMR 6.8, 95% CI 3.2-12.5), cardiomyopathy (9 deaths, expected 1.0, SMR 8.9, 95% CI 4.1-16.9) and pulmonary

embolism (6 deaths, expected 1.1, SMR 5.5, 95% CI 2.0-12.1). A rough estimate of the annual number of deaths nationally from each of these causes in people with ID would be five times the number we observed (our sample was four years deaths for roughly 5% of the population). Thus cerebrovascular disease is probably responsible for almost 200 deaths of people with ID annually in England and phlebitis, thrombophlebitis and pulmonary embolism almost 100.

The second most important chapter numerically was that covering diseases of the respiratory system. The most common subgroup in this chapter was 'influenza and pneumonia' (57 deaths, expected 7·4, SMR 7·7, 95% CI 5·8-9·9). Within this subgroup, all but one of the deaths for people with ID were attributed to 'pneumonia, organism unspecified', as were 99.2% of those for people without ID. This was followed by the subgroup of lung diseases due to external agents (24 deaths of people with ID, expected 1·1, SMR 21·8, 95% CI 13·9-32·4). All of these deaths were attributed to a single cause - pneumonitis due to solids and liquids.

The third ICD chapter numerically was neoplasms which accounted for 87 deaths. Overall the number of deaths attributed to neoplasms was not statistically remarkable (expected 75·9, SMR 1·1 95% CI 0·9-1·4). However the distribution between types of cancer was different from what would be expected. The largest subgroup was malignant neoplasms of the digestive organs (32 deaths, expected 20·8, SMR 1·5, 95% CI 1·1-2·2). Half of these were due to cancers of the colon and rectum (16 deaths, expected 6.8, SMR 2·4, 95% CI 1·3-3·8). This was more common in males than females (SMRs - males 2.68 (1.34 to 4.80), females 1.85 (0.60 to 4.32)). Unlike earlier studies (Jancar 1990) we found no excess of stomach cancer.

Malignant neoplasms of respiratory and intrathoracic organs accounted for twelve deaths, fewer (though not to the extent of statistical significance) than the expected 16.6 (SMR 0.7, 95% CI 0.4-1.3). Malignant neoplasms of female genital organs accounted for nine deaths (expected 4.0, SMR 2.3, 95% CI 1.0-4.3), almost equally divided between cancers of the uterine body, where this represented a statistically significantly high number, and of the ovary, where it did not. Nine deaths were attributed to malignant neoplasms of lymphoid, haematopoietic and related tissue (expected 5.7, SMR 1.6, 95% CI 0.7-3.0) but numbers in individual sub-categories were too small for detailed reporting. There was no single type of cancer for which there was a statistically significant low number of deaths. For breast cancer, six deaths would have been expected. The observed number was smaller but the shortfall did not come close to statistical significance. The number of brain cancers was too low to report but close to expectation.

In addition to exploring the ICD chapters accounting for the largest numbers of deaths of people with ID we looked for any other causes at three-character level ICD codes which were responsible for 2% or more of deaths of people with ID. The three other causes identified were dementia of unspecified type (33 deaths, expected 5·5, SMR 6·0, 95% CI 4·2-8·4), epilepsy (26, expected 0·8, SMR 34·4, 95% CI 23·4-50·5) and infantile cerebral palsy (23, expected 0·2, SMR 96·3, 95% CI 64·0-144·9).

Avoidable mortality

The Office for National Statistics reported that in England and Wales in 2013, 23% of deaths (28% in men; 17% in women) were from causes that met ONS definitions of being avoidable.(Office for National Statistics 2015) Using the same set of diagnoses and age bandings, mortality patterns in people without ID in our data were roughly in

line with this: 26·0% (95% CI 25·6% to 26·4%) of deaths of men, 16·4% (16·1% to 16·8%) of women and 21·0% (20·7% to 21·3%) of persons fell into this category. However a substantially higher proportion of deaths of people with ID were classifiable as avoidable: 50·9% (45·9% to 56·0%) of deaths of men, 36·9% (31·5% to 42·5%) of women, and 44·7% (41·0% to 48·5%) of all persons.

Some of this difference arose from the different profile of ages at death. With a few exceptions, (accidents, injuries and HIV disease) deaths from relevant causes are categorised as avoidable only within specified age limits, in most cases 0 to 74. This age band covers 76·5% of the deaths of people with ID in our data but only 29·6% of deaths of others. However, avoidable causes accounted for a larger proportion of deaths of people without ID than those with ID at each age group from 10 to 74.

Avoidable deaths may be from causes considered ‘preventable’ by public health measures or ‘amenable’ to good healthcare; Some causes are considered both amenable and preventable. Deaths from ‘amenable’ causes accounted for a higher proportion of all deaths of people with ID than of people without, whilst deaths from ‘preventable’ causes accounted for a higher proportion of deaths in people without, than with ID (Figure 2). This was true both overall and at each age group up to age 74

----------Figure 2 about here ----------

The most prominent causes of avoidable deaths differed between people with and without ID. Deaths from congenital malformations, deformations and chromosomal anomalies accounted for 19·0% of all avoidable deaths, and 24·1% of ‘amenable’

deaths in people with ID, but only 0·5% and 1·0% in other people. For people with ID the other major 'amenable' causes were pneumonia, ischaemic heart disease, epilepsy and cerebrovascular disease. The only 'preventable' cause that accounted for a notably higher proportion of deaths in people with ID was deep vein thrombosis with pulmonary embolism.

By contrast, in younger people without ID, suicide, accidents and alcohol related disorders were prominent 'preventable' causes, whilst in those older than 45, ischaemic heart disease, lung cancer, alcohol related diseases, chronic obstructive pulmonary disease, breast cancer and strokes predominated in this category.

The prominence of chromosomal abnormalities as causes of deaths classified as avoidable in people with ID raises the question of whether this category, as defined, can be considered a reliable marker of causes amenable to good care for this group. Of the 57 deaths in this category, 25 were of people with Down's syndrome aged between 45 and 74. It seems likely that not all of these would have been amenable to treatment. Excluding these reduced the amenable and avoidable proportions (amenable to 34.9% (31.4 to 38.6); avoidable to 44.7 (41.0 to 48.5)), but still left fractions highly significantly greater in people with ID than for people without ID.

---------- Table 4 about here ----------

Comparison of English data with other published studies

For comparison with the published data from other countries, and earlier data from Leicestershire in England, we calculated all-cause directly standardised death rates. As a majority of potentially includable studies did not have data for children and

young people we confined this analysis to the age groups in each study most neatly approximating to age 18 and older. The figures for our data were 26.6 deaths per thousand population per year for people with ID (95% confidence interval 24.0 to 29.4) and 11.2 (11.1 to 11.2) for other people, giving a comparative mortality index of 2.4. Figures for other regions for which reportable data have been published are shown in Table 4. The table also summarises the data sources and sample sizes. The Leicestershire and Ireland register figures gave higher rates than our, more recent England figures, as did those from the Massachusetts state monitoring system which are contemporary. The Finnish national figures and the data from the two Canadian provinces gave lower rates. We have not included data from a similar study of New South Wales in this table because the authors report that their data source gave substantially less complete identification of people with ID in those aged 70 or older. The much lower death rate produced is therefore probably not comparable.

Discussion

This study provides population-based evidence about mortality of people with ID in England. Compared with previous English population-based studies of this subject, the CPRD sample on which it is based is likely to be reasonably representative of the country as a whole and it is sufficiently large to provide data with usefully narrow confidence intervals from a fairly brief window in time (four years). The overall death rate is in line with national mortality data although the different English regions are unevenly represented.

The study has a number of specific limitations. We had no information about the subjects' level of ID. The overall prevalence of ID identified in the population studied (0.53%) is similar to that found in most service use registers, but substantially lower than found in English schools (more than 2%).(Emerson & Glover 2012) It is most likely that the individuals identified by teachers but missed from GP registers are those with mild ID and no associated syndromic cause. The findings of Arvio and her colleagues suggest that more mild levels of ID are likely to be associated with lower levels of excess mortality.(Arvio et al. 2016) At present, we are unable to determine this.

Studies of causes of death for people with ID that depend on death certification are inevitably affected by the likelihood that the accuracy of recording of causes is poorer or less complete for this group than for others.(Landes & Peek 2013; Tyrer & McGrother 2009; Heslop et al. 2013)

The numbers of deaths of people with ID identified as being from ethnic minority groups or individual regions proved too small for satisfactory investigation. This

indicates that understanding mortality patterns for members of minority ethnic groups in England is likely to require either specially targeted studies or whole population monitoring. Whilst the CPRD dataset provided too little coverage of some regional areas to permit a regional analysis of death rates for this study, its coverage is currently being extend in ways that should remedy this. Regional analyses of this topic may thus become feasible using CPRD data in the near future.

With these caveats, our findings indicate that after adjustment for age and gender, death rates for people with ID were more than three times those for the general population in England in the period to which the data relate. Correspondingly life expectancy for this group was two decades shorter. The level of excess mortality was greater for women than for men. This is a higher overall level of excess than reported in recent UK (Heslop et al. 2014; Glover & Christie 2014) and international studies.(Lauer et al. 2015; Ouellette-Kuntz et al. 2015; Florio & Trollor 2015). The gender imbalance reflects that reported by Arvio.(Arvio et al. 2016) Our comparison of these and other studies using directly standardised rates suggests that English mortality rates for people with ID may have been higher than those in some other countries. The earlier UK studies which reported SMRs of 1.92 (1.68-2.18)(Heslop et al. 2014) and 2.13 (1.09-2.83)(Glover & Christie 2014) focused respectively on a limited geographical area and very incomplete national reporting.

In relation to specific causes, the ICD chapter responsible for the largest number of deaths, as for the general population was circulatory diseases. Myocardial infarction and chronic ischaemic heart disease caused the most deaths and after adjusting for age and gender, death rates for these in people with ID were double those in the general population. Epidemiological research on age-related rates of cardiovascular

disease in people with ID is scarce and inconclusive.(Jansen et al. 2013; Patja et al. 2001) Cerebrovascular disease, pulmonary embolism and thrombophlebitis caused fewer deaths but the extent of excess was greater. We have not found studies reporting either epidemiology or death rates for these conditions in people with ID. The high death rate for cardiomyopathy may possibly be related to the treatment of leukaemia in patients with Down's syndrome, (O'Brien et al. 2008) but these deaths all occurred at ages above 35.

The second most important ICD chapter was respiratory diseases. Half of these related to pneumonia and a further 21% to external agents (largely aspiration of food or fluids). Literature about the epidemiology of dysphagia in people with ID is sparse, but suggests that the prevalence is high (around 8%) and people with ID and dysphagia often experience recurrent respiratory tract infections.(Chadwick & Jolliffe 2009) It is possible that aspiration problems also account for some pneumonia deaths as well as deaths from choking (here classified only as external causes of morbidity and mortality). A growing number of publications have advocated the need for improved practice in supporting people with IDs who have dysphagia.(Guthrie & Stansfield 2015)

The overall cancer death rate was not significantly different from expectation, but the distribution of deaths between cancer sites was different. A recent Australian study of cancer incidence in people with intellectual disability identified the stomach, colon and rectum, corpus uteri and brain as important sites for this group along with leukaemias and myelomas.(Sullivan et al. 2004) Our findings for cancer deaths identify some of these sites but not the stomach or the brain. Unlike their incidence data we also found colorectal cancer deaths more common in males than females.

Colorectal cancer has recently become the subject of a national screening programme in the UK so these figures may improve in the foreseeable future. A fuller understanding of the burden of cancer morbidity and mortality needs to consider the epidemiology in the light of exposure to known risk factor and influences on speed of diagnosis and uptake of treatment. Clarifying these issues will require more than simply death data because some forms of cancer are relatively treatable. However the importance of cancer as a cause of death does underline the urgent need for UK cancer registries to develop a way to flag for individuals with ID to allow proper monitoring of the disease burden and the effectiveness and equity of screening and treatment services.

Eight percent of deaths were given an underlying cause of a congenital malformation, deformation or chromosomal abnormalities, half of these Down's syndrome. This is relatively uninformative without more detail about the mediating conditions leading to their death. This raises an important issue for the design of future analyses of the topic. Deaths of people with Down's syndrome from heart disease, leukaemia, epilepsy or other conditions will not appear in the figures relating to those conditions if they are ascribed to the underlying chromosomal condition. This will change the epidemiological appearance of those conditions and understate the importance of planning and providing services for people with ID.

The international comparisons showed wide variations between areas. These need to be interpreted with caution. Recognition thresholds in the various countries and regions are likely to vary given the range of purposes served by the various data capture mechanisms used. Unfortunately most of the sources quoted did not provide data on the identified prevalence of ID in their source. A high threshold for

recognition is likely to be give rise to a more severely disabled group of individuals whose death rate is likely to be higher. Some differences may be attributable to period effects. The earlier English study gave a higher death rate. This may reflect changes in the overall death rates in England in the period separating it from our data, alternatively the county it studied may have a higher death rate for people with ID. It would be helpful if future publications in this area could state the identified prevalence of ID in the population studied.

The study indicates a number of areas where failings in health and social care are probably responsible for premature deaths. However the nature of the data sources means that whilst it is can indicate patterns at a national level, it cannot provide detailed evidence about how well more local areas are performing. In recent years in England there has been, what is in effect, a national register of people with ID in the form of general practice records. There is the means to extract and process data from these, linking them to mortality data regularly on a national basis. The government has a commitment to monitoring premature mortality in people with intellectual disability as a National Health Service outcomes target. However with all these elements it has not yet proved possible to surmount the information governance constraints and commercial considerations standing in the way of proper monitoring of mortality for people with ID. The finding of this study underline the urgency of getting around these obstacles and establishing a proper, regular, national monitoring system for mortality in people with ID.

References

Arvio, M., Salokivi, T., Tiitinen, A. & Haataja, L., 2016. Mortality in individuals with intellectual disabilities in Finland. *Brain and Behavior*, 6(2), p.n/a–n/a. Available at: http://onlinelibrary.wiley.com/doi/10.1002/brb3.431/epdf Accessed 8th April 2016 [Accessed January 27, 2016].

Chadwick, D.D. & Jolliffe, J., 2009. A descriptive investigation of dysphagia in adults with intellectual disabilities. *J Intellect Disabil Res*, 53(1), pp.29–43.

Eayres, D., 2008. *Commonly used public health statistics and their confidence intervals.*, Association of Public Health Observatories. Available at: http://www.apho.org.uk/resource/item.aspx?RID=48457.

Eayres, D., 2004. Life expectancy template. Available at: http://www.sepho.org.uk/viewResource.aspx?id=8943.

Emerson, E. & Glover, G., 2012. The “transition cliff” in the administrative prevalence of learning disabilities in England. *Tizard Learning Disability Review*, 17(3), pp.139–143.

Eurostat, 2013. *Revision of the European Standard Population. Report of Eurostat’s task force*, Luxembourg. Available at: http://ec.europa.eu/eurostat/web/products-manuals-and-guidelines/-/KS-RA-13-028.

Florio, T. & Trollor, J., 2015. Mortality among a Cohort of Persons with an Intellectual Disability in New South Wales, Australia. *Journal of applied research in intellectual disabilities : JARID*, 28(5), pp.383–93.

Glover, G. & Ayub, M., 2010. *How people with learning disabilities die.*, Improving Health and Lives: Learning Disabilities Observatory. Available at: http://www.improvinghealthandlives.org.uk/gsf.php5?f=8586.

Glover, G. & Christie, A., 2014. *Joint Health and Social Care Self-Assessment Framework 2013 Detailed report on Number Questions*, Cambridge, UK. Available at: http://www.improvinghealthandlives.org.uk/gsf.php5?f=312890.

Guthrie, S. & Stansfield, J., 2015. Teatime Threats. Choking Incidents at the Evening Meal. *Journal of applied research in intellectual disabilities : JARID*. Available at: http://onlinelibrary.wiley.com/doi/10.1111/jar.12218/pdf [Accessed April 4, 2016].

Heslop, P., Blair, P., Fleming, P., Hoghton, M. & Marriott, A., 2013. *Confidential Inquiry into premature deaths of people with learning disabilities (CIPOLD). Final report.*, Available at: www.bris.ac.uk/cipold/fullfinalreport.pdf.

Heslop, P., Blair, P.S., Fleming, P., Hoghton, M., Marriott, A. & Russ, L., 2014. The Confidential Inquiry into premature deaths of people with intellectual disabilities in the UK: a population-based study. *Lancet (London, England)*, 383(9920), pp.889–95.

Jancar, J., 1990. Cancer and mental handicap. A further study (1976-85). *Br J Psychiatry*, 156, pp.531–533.

Jansen, J., Rozeboom, W., Penning, C. & Evenhuis, H.M., 2013. Prevalence and incidence of myocardial infarction and cerebrovascular accident in ageing persons with intellectual disability. *Journal of intellectual disability research : JIDR*, 57(7), pp.681–5.

Khan, N.F., Harrison, S.E. & Rose, P.W., 2010. Validity of diagnostic coding within the General Practice Research Database: a systematic review. *The British journal of general practice : the journal of the Royal College of General Practitioners*, 60(572), pp.e128–e136.

Landes, S.D. & Peek, C.W., 2013. Death by mental retardation? The influence of ambiguity on death certificate coding error for adults with intellectual disability. *Journal of intellectual disability research : JIDR*, 57(12), pp.1183–90.

Lauer, E., 2016. *2012 & 2013 Mortality Report*, Charlestown, Ma. Available at: http://shriver.umassmed.edu/sites/shriver.umassmed.edu/files/2012-13 DDS Mortality Report Final_v2.pdf.

Lauer, E., Heslop, P. & Hoghton, M., 2015. Identifying and addressing disparities in mortality: US and UK perspectives. In C. Hatton & E. Emerson, eds. *International review of research in developmental disabilities*. International Review of Research in Developmental Disabilities. London: Elsevier, pp. 195–245.

Mazars, 2015. *Independent review of deaths of people with a learning disability or mental health problem in contact with Southern Health NHS Foundation Trust April 2011 to March 2015*, London. Available at: https://www.england.nhs.uk/south/wp-content/uploads/sites/6/2015/12/mazars-rep.pdf.

McCarron, M., Carroll, R., Kelly, C. & McCallion, P., 2015. Mortality Rates in the General Irish Population Compared to those with an Intellectual Disability from 2003 to 2012. *Journal of applied research in intellectual disabilities : JARID*, 28(5), pp.406–13.

Mencap, 2007. *Death by Indifference*, Available at: http://www.mencap.org.uk/document.asp?id=284.

NHS Primary Care Commissioning, 2012. QOF Business rules v21.0. , (21st January 2012). Available at: www.pcc.nhs.uk/business-rules-v21.0.

O'Brien, M.M., Taub, J.W., Chang, M.N., Massey, G. V, Stine, K.C., Raimondi, S.C., et al., 2008. Cardiomyopathy in children with Down syndrome treated for acute myeloid leukemia: a report from the Children's Oncology Group Study POG 9421. *Journal of clinical oncology : official journal of the American Society of Clinical Oncology*, 26(3), pp.414–20.

Office for National Statistics, 2015. *Avoidable Mortality in England and Wales, 2013*, Newport, South Wales. Available at: http://www.ons.gov.uk/ons/rel/subnational-health4/avoidable-mortality-in-england-and-wales/2013/stb.html.

Office for National Statistics, 2013. Statistical Bulletin Annual Mid-year Population Estimates , 2011 and 2012. , (August), pp.1–18. Available at: http://www.ons.gov.uk/ons/dcp171778_320900.pdf.

Ouellette-Kuntz, H., Shooshtari, S., Balogh, R. & Martens, P., 2015. Understanding Information About Mortality Among People with Intellectual and Developmental Disabilities in Canada. *Journal of Applied Research in Intellectual Disabilities*, 28(5), pp.423–435.

Patja, K., Mölsä, P. & Iivanainen, M., 2001. Cause-specific mortality of people with intellectual disability in a population-based, 35-year follow-up study. *J Intellect Disabil Res*, 45(Pt) 1, pp.30–40.

Sullivan, S.G., Hussain, R., Threlfall, T. & Bittles, A.H., 2004. The incidence of cancer in people with intellectual disabilities. *Cancer Causes Control*, 15(10), pp.1021–1025.

Tyrer, F. & McGrother, C., 2009. Cause-specific mortality and death certificate reporting in adults with moderate to profound intellectual disability. *J Intellect Disabil Res*, 53(11), pp.898–904.

Tyrer, F., Smith, L.K. & McGrother, C.W., 2007. Mortality in adults with moderate to profound intellectual disability: a population-based study. *J Intellect Disabil Res*, 51(Pt) 7, pp.520–527.

Funding

The work was made possible by funding from NHS England, Public Health England, and the Department of Health (England). The funders played no role in the design, data collection, analysis or interpretation of the data; nor in the writing of the report or the decision to submit the paper for publication.

Conflicts of Interest

None

Table 1. Composition of population and deaths studied. The table shows, by age, gender, region of residence, and year of study, the total person-years exposure to risk (PYER), the proportions (%) identified as having an intellectual disability and the number of deaths.

Grouping	Total PYER (% of total)	Proportion with ID	Total deaths (% of total)
Total	11,163,234.1	0.53%	98,035
Age groups			
00-09	1,250,702.6 (11.2%)	0.27%	226 (0.2%)
10-17	1,014,574.2 (9.1%)	0.67%	113 (0.1%)
18-24	891,445.0 (8.0%)	0.93%	269 (0.3%)
25-34	1,461,908.7 (13.1%)	0.66%	579 (0.6%)
35-44	1,584,597.4 (14.2%)	0.60%	1,505 (1.5%)
45-54	1,636,034.0 (14.7%)	0.65%	3,647 (3.7%)
55-64	1,326,571.6 (11.9%)	0.47%	7,796 (8.0%)
65-74	1,046,218.1 (9.4%)	0.32%	15,184 (15.5%)
75-84	662,177.9 (5.9%)	0.17%	28,671 (29.2%)
85-99	289,004.5 (2.6%)	0.08%	40,045 (40.8%)
Genders			
Male	5,518,004.6 (49.4%)	0.62%	46,806 (47.7%)
Female	5,645,185.1 (50.6%)	0.45%	51,229 (52.3%)
Indeterminate	44.3 (0.0%)	0.00%	0 (0.0%)
Region			
East Midlands	171,895.0 (1.5%)	0.55%	1,299 (1.3%)
East of England	1,202,749.8 (10.8%)	0.49%	9,778 (10.0%)
London	1,775,876.7 (15.9%)	0.48%	12,054 (12.3%)
North East	248,520.3 (2.2%)	1.02%	2,369 (2.4%)
North West	1,681,188.1 (15.1%)	0.60%	15,649 (16.0%)
South Central	1,478,524.6 (13.2%)	0.53%	13,170 (13.4%)
South East Coast	1,548,890.8 (13.9%)	0.44%	13,572 (13.8%)
South West	1,412,266.3 (12.7%)	0.65%	14,579 (14.9%)
West Midlands	1,336,093.5 (12.0%)	0.47%	12,546 (12.8%)
Yorks / Humber	307,228.9 (2.8%)	0.47%	3,019 (3.1%)
Year			
2010/11	2,968,677.5 (59.2%)	0.53%	26,206 (26.7%)
2011/12	2,880,416.9 (1.0%)	0.53%	25,269 (25.8%)
2012/13	2,797,156.5 (0.5%)	0.53%	25,229 (25.7%)
2013/14	2,516,983.2 (0.4%)	0.53%	21,331 (21.8%)
Intellectual disability status			
With evidence of ID	59,279.7 (0.5%)	-	664 (0.7%)
Without evidence of ID	11,103,954.4 (99.5%)	-	97,371 (99.3%)

Table 2 Age/sex specific death rates per 1000 population, all causes for people with and without intellectual disability, and ratio of observed deaths of those with ID to number expected assuming overall age / sex specific death rate. 95% confidence intervals in brackets.

Age group	Age specific death rates per 1000 population With intellectual disability	Without intellectual disability	Observed / expected deaths
Females			
01-09	8.3 (4.2 to 14.9)	0.1 (0.1 to 0.2)	49.9 (24.9 to 89.4)
10-17	3.6 (1.5 to 7.0)	0.1 (0.0 to 0.1)	41.2 (17.7 to 81.2)
18-24	1.6 (0.5 to 3.8)	0.2 (0.1 to 0.2)	8.5 (2.7 to 19.8)
25-34	3.0 (1.6 to 5.3)	0.2 (0.2 to 0.3)	11.6 (6.0 to 20.2)
35-44	4.4 (2.7 to 6.8)	0.7 (0.7 to 0.8)	6.0 (3.7 to 9.3)
45-54	8.2 (5.8 to 11.1)	1.9 (1.8 to 2.0)	4.2 (3.0 to 5.6)
55-64	21.6 (16.6 to 27.6)	4.8 (4.6 to 5.0)	4.4 (3.4 to 5.7)
65-74	35.1 (26.4 to 45.6)	11.7 (11.4 to 12.0)	3.0 (2.2 to 3.9)
75-84	85.7 (63.6 to 113.0)	37.2 (36.6 to 37.8)	2.3 (1.7 to 3.0)
85-99	222.4 (147.8 to 321.5)	131.6 (130.0 to 133.3)	1.7 (1.1 to 2.4)
Total	11.6 (10.3 to 13.0)	9.1 (9.0 to 9.1)	3.4 (3.0 to 3.8)
Males			
01-09	3.8 (1.7 to 7.6)	0.2 (0.2 to 0.2)	19.8 (8.5 to 39.0)
10-17	1.3 (0.5 to 2.9)	0.1 (0.1 to 0.2)	9.7 (3.6 to 21.2)
18-24	1.0 (0.3 to 2.2)	0.4 (0.3 to 0.5)	2.3 (0.8 to 5.5)
25-34	3.5 (2.1 to 5.4)	0.5 (0.5 to 0.6)	6.6 (4.0 to 10.1)
35-44	4.4 (2.8 to 6.7)	1.1 (1.1 to 1.2)	3.8 (2.4 to 5.7)
45-54	10.8 (8.3 to 13.9)	2.4 (2.3 to 2.5)	4.3 (3.3 to 5.6)
55-64	26.7 (21.5 to 32.8)	6.8 (6.6 to 7.0)	3.9 (3.1 to 4.8)
65-74	45.7 (36.4 to 56.8)	17.3 (17.0 to 17.7)	2.6 (2.1 to 3.3)
75-84	104.3 (79.0 to 135.2)	50.9 (50.1 to 51.7)	2.0 (1.5 to 2.7)
85-99	221.0 (136.8 to 337.9)	152.0 (149.6 to 154.5)	1.5 (0.9 to 2.2)
Total	10.9 (9.9 to 12.1)	8.5 (8.4 to 8.5)	3.0 (2.7 to 3.3)
Persons			
01-09	5.6 (3.4 to 8.7)	0.2 (0.1 to 0.2)	30.4 (18.3 to 47.5)
10-17	2.1 (1.1 to 3.4)	0.1 (0.1 to 0.1)	17.3 (9.4 to 29.0)
18-24	1.2 (0.6 to 2.2)	0.3 (0.3 to 0.3)	3.7 (1.8 to 6.8)
25-34	3.3 (2.3 to 4.7)	0.4 (0.3 to 0.4)	7.8 (5.4 to 11.1)
35-44	4.4 (3.2 to 6.0)	0.9 (0.9 to 1.0)	4.6 (3.3 to 6.2)
45-54	9.5 (7.8 to 11.6)	2.2 (2.1 to 2.3)	4.3 (3.5 to 5.2)
55-64	24.3 (20.6 to 28.5)	5.8 (5.7 to 5.9)	4.1 (3.5 to 4.8)
65-74	40.8 (34.2 to 48.2)	14.4 (14.2 to 14.7)	2.8 (2.3 to 3.3)
75-84	94.7 (77.6 to 114.5)	43.2 (42.7 to 43.7)	2.2 (1.8 to 2.6)
85-99	221.8 (164.1 to 293.3)	138.5 (137.1 to 139.9)	1.6 (1.2 to 2.1)
Total	11.2 (10.4 to 12.1)	8.8 (8.7 to 8.8)	3.2 (2.9 to 3.4)

Table 3. Standardised mortality ratios (SMRs) associated with intellectual disability by ICD Chapter of underlying cause of death

ICD10 Chapter	Observed/ expected deaths	SMR (95% Confidence interval)
I Certain infectious and parasitic diseases	7/2·2	3·2 (1·3 to 6·5)
II Neoplasms	87/75·9	1·1 (0·9 to 1·4)
III Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	0/0·5	-
IV Endocrine, nutritional and metabolic diseases	15/3·0	5·1 (2·8 to 8·3)
V Mental and behavioural disorders	43/7·9	5·4 (3·9 to 7·3)
VI Diseases of the nervous system	85/8·7	9·8 (7·8 to 12·1)
VII Diseases of the eye and adnexa	0/0·0	-
VIII Diseases of the ear and mastoid process	0/0·0	-
IX Diseases of the circulatory system	152/53·3	2·8 (2·4 to 3·3)
X Diseases of the respiratory system	114/23·3	4·9 (4·0 to 5·9)
XI Diseases of the digestive system	52/13·1	4·0 (3·0 to 5·2)
XII Diseases of the skin and subcutaneous tissue	*/0·6	-
XIII Diseases of the musculoskeletal system and connective tissue	*/1·4	-
XIV Diseases of the genitourinary system	16/3·0	5·4 (3·1 to 8·7)
XV Pregnancy, childbirth and the puerperium	0/0·0	-
XVI Certain conditions originating in the perinatal period	0/0.0	-
XVII Congenital malformations, deformations and chromosomal abnormalities	56/0·8	72·9 (55·1 to 94·7)
XVIII Symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified	*/2·2	-
XX External causes of morbidity and mortality	25/12·2	2·0 (1·3 to 3·0)
Cause Unknown	0/0·5	-
All causes	664/208·7	3·2 (2·9 to 3·4)

* - Between 1 and 5 deaths

Table 4. Comparison of directly standardised, all-causes mortality rate for people with ID with comparable figures from other national or sub-national studies. Rates are standardised to 2013 European standard population.

Location	Dates	ID PYER	Directly Standardised mortality rate for people with ID	Comments	Reference
Leicestershire, England	1993-2005	23,077	34.4 (30.4 to 38.7)	Intellectual disability register, Age 20+	Tyrer et al. 2007
Finland	1996-2007	333,041	19.5 (18.9 to 20.1)	National social security linked to mortality Age 15+	Arvio et al. 2016
Manitoba, Canada	2000-2005	(Age 20+ Less than 29000)	22.5 (CIs not calculable)	Established mortality / social security / health data warehouse Age 20+	Ouellette-Kuntz et al. 2015
Republic of Ireland	2002-2012	173,964	42.2 (39.7 to 44.7)	Intellectual disability register, Age 20+	McCarron et al. 2015
South East Ontario, Canada	2004-2011	14,598	19.0 (15.9 to 22.6)	Intellectual disability register, Age 20+	Ouellette-Kuntz et al. 2015
England	2010-2014	49,061	26.6 (24.0 to 29.4)	Primary care data linked to mortality Age 18+	This study
Massachusetts, US	2012-2013	46,308	33.6 (30.9 to 36.5)	State monitoring system, Age 18+	(Lauer 2016)

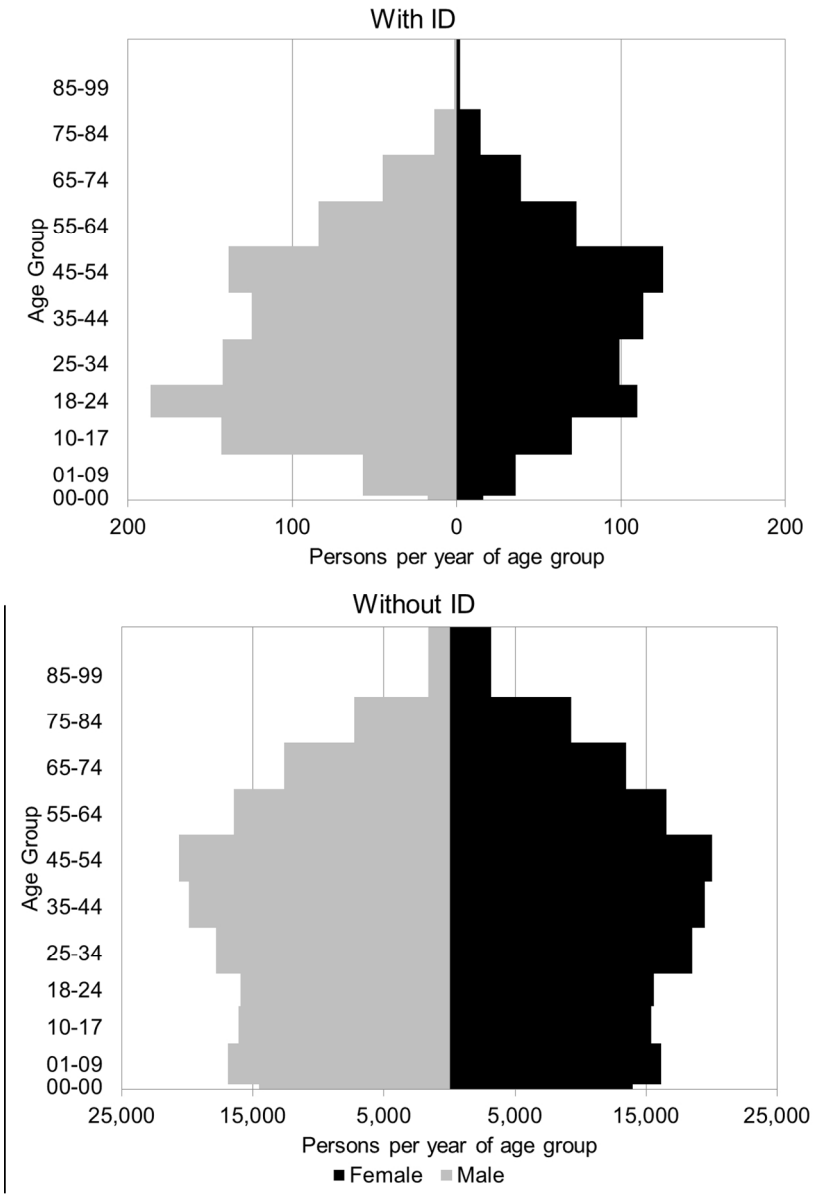

Figure 1. Population pyramids, people with and without intellectual disabilities

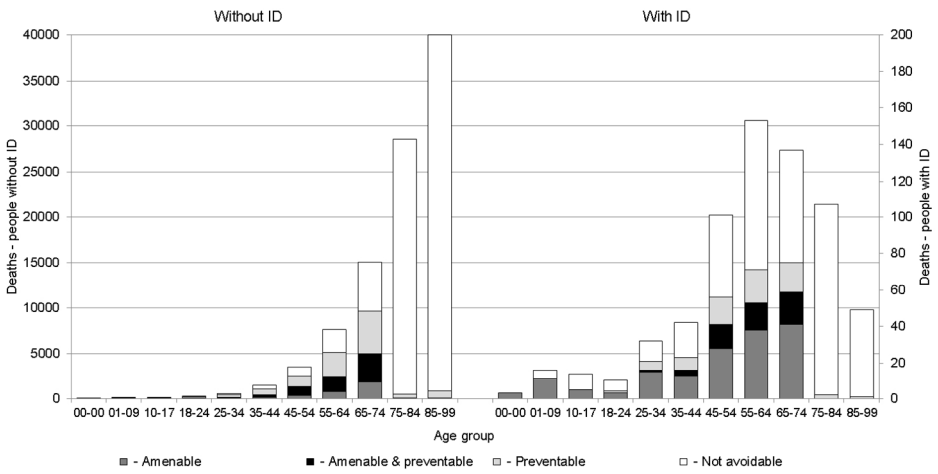

Figure 2 Deaths attributed to causes categorised as preventable, amenable to medical care, both preventable and amenable and not avoidable on the basis of underlying cause and age at death, by age group and intellectual disability status