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Mortality in people with intellectual disabilities

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Introduction

In 2013 a group of international collaborators met to discuss what we know about the age and causes of death of people with intellectual disabilities (ID), the limitations of our existing data and how we could improve our knowledge in this respect. The collaboration has since been supported by a UK Economic and Social Research Council (ESRC) grant for a series of webinars and meetings from 2014 – 2017 to bring together academics, practitioners and policy-makers to discuss the development of internationally comparable mortality data in relation to people with ID. In this Special Edition, some of the contributors to those discussions set out the best evidence we have to-date about mortality data relating to people with ID.

But first, let us review why an understanding of mortality data in general, and in relation to people with ID in particular, is such an important area of concern.

Surveillance of patterns of mortality in vital statistics

Analysis of mortality data to improve the health of the population has a long history. In the UK, William Farr used mortality data in the 1850s to stimulate debate about why

some cities were less healthy than others (Szreter 1991). At about the same time in the US, the first mortality report by the Massachusetts Sanitary Commission highlighted differences in age at death between farmers and mechanics, urging doctors to make more inquiries into the causes of death which prematurely shortened lives (Shattuck 1850). During the 19th and 20th centuries, as mortality data became available in industrialised countries, public health became transformed (Jha 2012). Doll & Hill (1950) used mortality data to highlight a substantial increase in male deaths from lung cancer and hypothesised the causal association between smoking tobacco and lung cancer. In the US, Gottlieb et al. (1981) used routine mortality data from San Francisco to signal the start of the American HIV epidemic. Reliable and timely information on cause-specific mortality has now become a fundamental component of evidence-based health policy development, implementation, and evaluation. In short, Jha (2012) argues that: '*counting the dead is one of the world's best investments to reduce premature mortality worldwide as it is one of the most robust ways to measure accurately the effectiveness of investments aimed at reducing child and adult mortality*' (p. 1.

Reducing premature mortality has been a priority nationally and internationally (World Health Organisation 2013; United Nations 2014) over recent years, but there are a number of ways in which what constitutes a 'premature death' might be determined, including:

- Life expectancy at birth - the average survival time that members of a population can expect to live from the time of birth.

- Life expectancy at age 75 – the additional number of years a person can expect to live if they reach the age of 75. This takes account of the fact that as people age, their life expectancy increases because they will have already have survived a myriad of potential causes of death.

- Standardised Mortality Ratio (SMR) - the ratio of deaths observed in a particular group to those expected in a reference population of identical age structure. This provides a measure of how many more deaths than expected (if the death rate in the general population were applied) are observed in the population of interest. SMRs can be compared over time, between sexes and with equivalent statistics for other countries.

- Years of life lost - the total number of life years lost due to premature death. It is calculated by subtracting age at death from a threshold age, most commonly a threshold age for prematurity such as age 65 or 70, but the upper limit could also be life expectancy at birth or other variants.

These measures of premature death rely on population-based mortality data and in many developed countries, national vital registration systems are the major source of such data. These systems provide universal coverage of the population and operate continuously; official notification to the registration office that a death has occurred is usually a legal requirement before burial or cremation can take place, and information about age at death, gender, place and time of death and cause of death is collected about each death and then collated centrally at population level. Once such data has been collated, surveillance of patterns of mortality data can take place.

Surveying patterns of mortality data is a reflexive process, identifying and responding to emerging trends and indicating where further analysis and action is required. Sutton (2012)

suggests that mortality surveillance should consist of two key elements. The first is the ongoing, systematic monitoring and analysis of mortality data. This requires that the entire population of a particular geographic area is included, and that the mortality data is accurate in terms of the characteristics of the people who have died and their causes of death. The second is that timely analysis of the data and dissemination of its findings will lead to actions being taken – either in relation to public health concerns raised by an understanding of the patterns of mortality, or in relation to the quality of the data itself.

Mortality data about people with ID

While information about the age and cause of death of the general population is routinely reported at national and cross-national levels, it is not usually possible to extract data about people with ID from this information. As a result, we know little at national and international levels about the age and cause of death of people with ID and are missing an important source of information about the health of this vulnerable population. There are a number of reasons why this is the case:

- A lack of consistency in the identification of people with ID. Definitions used to describe people with ID vary across countries and over time. At a local level in many countries, people with ID are identified via administrative criteria – so they are classed as having ID if they are eligible for ID services. However, eligibility criteria for such services vary, and many people, particularly those with mild ID, are unlikely to be included in such definitions.

- Multiple coding options for the existence of ID. Glover & Ayub (2008), for example, identified 48 ICD10 codes for medical conditions usually associated with ID, and 76 ICD10 codes for conditions sometimes associated with ID.

- Incomplete and incompatible registers of people with ID. In Canada and the USA, for example, funding and provision of most services (education, health, and social services) is under provincial or state jurisdiction and there is no adequate national dataset(s) of people with ID. In England, registers are held by Local Authorities about the number of children with special educational needs whose primary need is because of ID, and by primary care doctors (GPs) of people with

ID registered with their practice, but there is a considerable disjoint between the registers (Emerson & Glover 2012).

- A lack of recording of ID on Cause of Death certificates. World Health Organisation rules for completing the Cause of Death certificate requires a record of the sequential train of events leading to death, plus a record of any other diseases, injuries, conditions or events that contributed to the death but were not part of the direct sequence of events leading to death. In many cases, one's ID may not cause or contribute to the person's death, so would not therefore be recorded. In England, Tyrer & McGrother (2009) noted that only 41% of Cause of Death certificates of people known to have ID mentioned ID or an associated condition. The proportion in the English Confidential Inquiry into premature deaths of people with ID (CIPOLD) was 23%; here people with profound and multiple ID were identified on a Cause of Death certificate more frequently (58%) than people with mild ID (9%) (Heslop et al. 2013).

- The rate of coding errors on Cause of Death certificates for people with ID. An additional factor

precluding the identification of people with ID from

Cause of Death certificates is the rate of coding errors

on these certificates for people with ID (Ouellette-

Kuntz 2005; Landes & Peek 2013). Landes & Peek

(2013) identified 2,278 adults (aged 21 and over) who

had ID recorded on their Cause of Death certificate

but reported that 20% of these were coded

erroneously by stating 'mental retardation' as the

underlying cause of death. They concluded that

diagnostic overshadowing may be obscuring the true

cause of death of some people with ID.

Even if we were to overcome each of these issues, the

challenge of identifying premature death in people with ID

remains. For example, life expectancy at birth measures

using 'average' life expectancy are problematic in relation to

some people with ID because they do not allow

consideration of some early deaths given the presence of

particular conditions associated with ID. As an illustration, it

is unusual for someone born with Batten disease to live

beyond their 30s, so all people with Batten disease would be

identified as dying prematurely if the 'average' age of death

of the general population was used as a threshold of

premature death, irrespective of the person's potential life expectancy. Similarly, life expectancy at age 75 measures are inappropriate for people with moderate and severe ID who are less likely to reach the age of 75 than those without ID (Patja et al. 2000). In addition, life expectancy calculations can only be performed with confidence with population-years at risk of 5,000 or more (Williams et al. 2005), and unless internationally comparable data is used, the small number of people with ID likely to be in some subgroups is problematic.

As most of the authors in this Special Edition indicate, using SMR provides a better measure of excess deaths in people with ID, although a comparatively small number of deaths of people with ID could give rise to a large amount of random variation, necessitating the aggregation of data over a number of years, or the combination of data from more than one geographic area.

Responding to the challenges faced

The challenge to obtain robust data about mortality of people with ID is considerable, but this Special Edition presents contributions from leading academics in a range of countries

about the data that is available in each country and how they are addressing the gaps in our knowledge about mortality of people with ID. We move from data relating to smaller geographical areas, through to national studies, and then to overviews of multiple studies within the same country.

The first paper from Dieckmann et al. presents data about life expectancy of people with ID in **Germany**. The authors start with a helpful overview of international studies about mortality of people with ID from the USA, Finland, Western Australia and England. Overall, Dieckmann et al. suggest that although caution should be used when comparing the results of the studies, taken together they suggest that the life expectancy of people with ID has increased during the last two decades even though it remains below the life expectancy of the general population, gender-specific differences appear to be less significant than in the general population, and the severity of ID seems to be a strong influential factor in life expectancy.

In Germany, to-date, there has been very little or no empirical research about the life expectancy and mortality of people with ID. Germany has a particular age profile of

people with ID because of the systematic killing of disabled

people, including people with ID, in the Nazi era. As a result,

the number of people with ID of retirement age in Germany

is relatively small, and they are likely to be fairly resilient – or

"healthy survivors" as Dieckmann et al. describe them, which

can cause an overestimation of survival rates in older-age

groups of people with ID.

Dieckmann et al. report data from two regional population

samples in Germany and calculated age-group-specific

mortality rates by exponential regression analyses. They

report an average life expectancy of men with ID to be 70.9

years in one area, and 65.3 years in the other. The life

expectancy of women with ID was 72.8 years in one area

and 69.9 years in the other. The mortality rates of the

younger age groups of people with ID were significantly

higher than those of the general population, particularly up to

the age of 45-52 for men (according to the sample

population), and 52-53 for women. In the older populations

at age 84 or more, the rates were almost the same as in the

general population.

The second paper by Florio & Trollor compares mortality for people with ID to the general population in **New South Wales (NSW), Australia.** The authors have linked two sources of data: retrospective data (from 2005 onwards) from an administrative register of the NSW Department of Ageing Disability and Home Care which identifies people of all ages who are known to have an ID, and mortality data from the NSW Register of Births, Deaths and Marriages for the period June 1, 2005 to December 31, 2011. By using record linkage in this way the authors have been able to determine age and sex specific counts and rates of mortality for the cohort of people with ID aged 5-69 years old, who have previously received a state-funded ID service.

Using the World Health Organization (WHO) standard age distribution for standardisation, Florio & Trollor report an Age Standardized Death Rate of 4.04 deaths per 1,000 in the ID cohort, compared with 1.58 deaths per 1,000 for the rest of the NSW population. The Age-Adjusted Death Rates are greater for the ID Cohort at all ages between 5 and 69 years old. The SMR for the ID Cohort was 3.15, but a significant difference was found between males (SMR 2.52) and females (SMR 4.26). The greater relative mortality in

females compared to males is noted to be different from the pattern in the general population of many countries where it is males that have greater relative mortality. It is an issue that the authors suggest should be explored further in future work and other cohorts.

The third paper by Lauer & McCallion utilises information from four **USA** state disability service systems (Massachusetts, Connecticut, Ohio and Louisiana) that support people with intellectual and developmental disabilities (I/DD) to calculate average age of death and crude mortality rates. Each of these states has sufficient, well-established mortality reporting systems and similar I/DD service systems in order to facilitate aggregation and comparison of data. Contrasting information is provided by de-identified Medicaid claims data from the US State of New York.

The authors suggest that people with I/DD continue to experience life expectancies that are approximately 20 years lower than the general population of the USA, crude mortality rates that are between 20% and 50% higher than the general population, and age-adjusted rates that are

about 80% higher. A notable gender difference was found, with females with I/DD experience greater disparities from the general population than males with I/DD (19.1 years difference in average age at death for females compared with 16.8 years difference for males). Such a gender difference, as Florio & Trollor (ibid.) also contend, requires greater investigation, and Lauer & McCallion suggest that a potential way forward in the USA would be for closer examination of Medicaid and medical claims data to better understand the influence on mortality of a range of possible contributory factors.

The fourth paper in this collection is from McCarron and colleagues in **Ireland**. Ireland is interesting because it has a National Intellectual Disability Database (NIDD), an administrative database that includes details about all people with ID who are eligible for or receive services in the Republic of Ireland. Data is updated annually, and is available for almost 32,000 people from 2003-2012.

The authors report an overall mortality rate in people with ID of 8.35%, but that this depended on age and severity of ID. Overall mortality in people with ID was four times higher than

in the general population, and they were, on average, dying 19 years earlier than peers in the general population. In the youngest age group mortality was almost seven times higher in the ID population than the general population but this decreased with increasing age. The overall SMR for females was larger than for males, but gender differences diminished with increasing age. The average age at death was lowest for those with profound ID and increased with decreasing severity of ID.

The authors suggest that between 2001 and 2003 there had been an increase in average age at death of approximately nine years and that although the increase had been sustained over the following decade, no further increases were noted during that time period. Some of the initial apparent improvements are likely to have been due to improved case finding, particularly of people with milder ID who are more likely to live longer lives. However, greater efforts are needs to more fully understand this pattern and the actions needed to sustain improvements in the life expectancy of people with ID.

The final two papers in this Special Edition provide overviews of a number of different studies within the same country. We start with a paper about mortality of people with ID in **England** by Heslop & Glover. They present the findings from four very different, recent sources of data about mortality of people with ID in England.

From piecing this information together, and reanalysing some data in the light of information provided by another of the data sources, the authors draw conclusions that are similar to those of the other studies already reported in this Special Edition: that people with ID have a reduced life expectancy of 13-20 years in England when compared with the general population, and that the differences are greatest for women (compared with women in the general population) and for those with the most severe ID (compared with people with mild ID), although people with mild ID also died significantly younger than people in the general population. The SMRs of people with ID were twice that of the general population in England, and there was little indication of any reduction in this over time.

The authors comment that although each of the data
sources contributes some information, none provide
comprehensive national data about mortality of people with
ID that is able to take account of the age and sex distribution
of the population. Such data is currently lacking in England,
but is urgently needed in order to better monitor the mortality
of people with ID and the impact of any actions that are
being taken to address the health inequalities faced by this
population. In England, the most promising way forward will
be to link data from GP registers of people with ID with
national mortality data held by the Office for National
Statistics but to-date, delays over the past two years have
not yet seen this initiative come to fruition.

The final paper is by Ouellette-Kuntz and colleagues in
Canada who review three studies about mortality of people
with intellectual and developmental disabilities in the
provinces of Ontario and Manitoba. Taken together, the
studies suggest that there is evidence of excess mortality in
Canadians with intellectual and developmental disabilities,
and that while excess mortality occurs at all ages, it is most
pronounced in childhood and early adulthood. The authors
attribute some of the excess mortality to comorbidities more

commonly seen in people with intellectual and developmental disabilities, and suggest that women may have a greater risk of death than men.

The recent approach to link data mortality data with data that identifies people with intellectual and developmental disabilities has been a promising development in Manitoba by Shooshtari and Martens, and the development of similar work in Ontario holds exciting promise for better understanding the patterns of mortality in this population. It is an approach that is increasingly being recognized as offering significant potential in identifying the extent of premature deaths in people with intellectual and developmental disabilities, and changes over time.

Together these papers present the current 'state of play' in each country of what we know about mortality of people with ID. They present the limitations of our existing data, and proposals for improving our knowledge in this area. With better information about mortality in this vulnerable population, we may be able to better understand how life expectancy and the health of the population can be

improved. The study of mortality in the general population has moved far beyond the examination of demographic factors through innovative new models and methods to improve our understanding of sources of health risk and pathways through which health outcomes can be improved (Rogers & Crimmins). Without better mortality information, the people with ID are unlikely to benefit from these methods and we may continue to struggle with little reliable evidence of health and social inequities. It is imperative that disability researchers and public health departments work together to improve access to and the quality of mortality information to facilitate monitoring of this population.

The collection also focuses our intention, as ultimately, what we are working towards is the availability of evidence that will lead to measures being introduced to reduce premature and avoidable deaths in people with ID and ultimately improve the health of the population. It is this ambition that acts as the driver for much of the work being undertaken. It is also the hope of this group that the information and methodologies presented in this collection may serve as a model for other countries to begin or improve their mortality surveillance of people with ID.

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