Causes of mortality in older people with intellectual disabilities: results from the HA-ID study

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Abstract

We aim to provide insight into the cause-specific mortality of older adults with ID, with and without Down syndrome (DS), and compare this to the general population. Immediate and primary cause of death were collected through medical files of 1050 older adults with ID, five years after the start of the Healthy Ageing and Intellectual Disabilities (HA-ID) study. During the follow-up period, 207 (19.7%) participants died, of whom 54 (26.1%) had DS. Respiratory failure was the most common immediate cause of death (43.4%), followed by dehydration/malnutrition (20.8%), and cardiovascular diseases (9.4%). In adults with DS this was respiratory disease (73.3%), infectious and bacterial diseases (4.4%), and diseases of the digestive system (4.4%). Diseases of the respiratory system also formed the largest group of primary causes of death (32.1%; 80.4% was due to pneumonia), followed by neoplasms (17.6%), and diseases of the circulatory system (8.2%). In adults with DS the main primary cause was also respiratory diseases (51.1%), followed by dementia (22.2%).

Keywords

Intellectual disabilities, older adults, cause-specific mortality

Introduction

During the last decades, life expectancy of people with intellectual disabilities (ID) has increased as a result of improved healthcare (Patja, Molsa, & Iivanainen, 2001). The mean mortality age for in-clinic people with ID has increased from 19 years in the 1930s (Carter & Jancar, 1983) to the late 50s - mid-60s in the late 1990s (Janicki, Dalton, Henderson, & Davidson, 1999). In 2013, a mean age at death of 64 years was found in adults with mild to severe ID, with a decrease in the median age at death with increasing severity of ID (Heslop et al., 2014). The life expectancy of people with Down syndrome (DS) has increased as well, however their life expectancy is lower than that observed in the population of ID as a whole (Coppus, 2013).

Although life expectancy of people with ID has increased, it still remains lower than that of the general population. This difference may be explained by the high prevalence of neurological disorders, congenital disorders and other chronic comorbidity in people with ID (Patja et al., 2001). These “ID-specific conditions” are likely to contribute to the high prevalence of cardiovascular risk factors, depression, sleep disorders, and dysphagia (Bastiaanse, Kamp, Evenhuis, & Echteld, submitted; de Winter, Bastiaanse, Hilgenkamp, Evenhuis, & Echteld, 2012; Hermans, Beekman, & Evenhuis, 2013; van de Wouw, Evenhuis, & Echteld, 2013) in later life, which could be causes of premature death. Besides the lower life expectancy, the higher prevalence of these health conditions could also cause the distribution of causes of death in older adults with ID to be different than in the general population.

One of the few studies that evaluated cause-specific mortality (Tyrer & McGrother, 2009) investigated adults with moderate to profound ID aged 20 years and over (*n*= 2995), and found a relatively high mortality caused by congenital abnormalities, diseases of the nervous system, mental disorder, and pneumonia. However, no distinction was made for different age groups or older adults specifically. Another study regarding cause-specific mortality in people with ID aged between 2 and 97 years, found cardiovascular diseases, respiratory diseases, and neoplasms to be the three most common causes of death in both men and women with ID aged 40 years and over (Patja et al., 2001). In the general population the three most common causes of death were cardiovascular diseases, neoplasms, and external causes (for men) or respiratory diseases (for women). Thus in the study of Patja et al. (2001)

the cause-specific mortality differed significantly from the general population, with reduced mortality from cardiovascular diseases, neoplasms, and external causes, and increased mortality from respiratory diseases in adults with ID (Patja et al., 2001).

So far, no studies have been performed regarding the cause-specific mortality of specifically the ageing (≥ 50 years) population with ID. Hardly any knowledge about mortality patterns in adults with ID past middle age is available, while the prolonged life expectancy creates a growing population of older adults with ID (Patja, Iivanainen, Vesala, Oksanen, & Ruoppila, 2000). Furthermore, previous studies have been performed several years ago and healthcare and therapies have improved since. Hence, there could have been changes in the cause-specific mortality.

Therefore, the aim of this study is to provide more insight into the cause-specific mortality of older adults with ID aged 50 years and over. Cause-specific mortality will be assessed for the group as a whole, and for participants with and without DS separately. The secondary aim is to compare the cause-specific mortality of this group to the cause-specific mortality in the general older population.

Methods

Study design and participants

This study was part of the Healthy Ageing and Intellectual Disability (HA-ID) study, a prospective cohort study performed in the Netherlands. In the HA-ID study, the health status of older adults with ID (≥ 50 years) was investigated within three themes: 1) physical activity and fitness, 2) nutrition and nutrional state, and 3) mood and anxiety. Extensive measurements were taken for each theme, such as an extensive physical fitness assessment, physical activity measurements with pedometers, physical examination, swallowing observations, laboratory assessment, depression and anxiety screening, questionnaires regarding daily functioning and falls, in addition to obtaining information from medical files.

Participants were recruited through three participating care organizations in the Netherlands. All clients aged 50 years and over receiving care from one of the participating organizations, ranging from residence with complete care to only ambulatory support without residence, were eligible to participate, without any other exclusion criteria. At the start of the study, in 2008, the care organizations provided care and support for 2322 clients aged 50 years and over, which were all invited to participate. Clients who were capable to decide themselves if they wanted to participate provided informed consent themselves, for clients who were not capable to do so the legal representative provided informed consent. Eventually 1050 clients participated in the HA-ID study. This was a near-representative sample of older adults with ID receiving formal care in the Netherlands, with an underrepresentation of 80 to 84 year-olds, and a slight overrepresentation of women. A more detailed description of the study design, recruitment, and representativeness of the sample has been published elsewhere (Hilgenkamp et al., 2011).

The extensive baseline data collection on the three themes took place between November 2008 and July 2010. All data were stored at and accessible from the server of the Erasmus MC, which has high standards regarding back-ups, safety, and privacy. After a 5-year follow-up period, mortality data were collected up to March 2015. The follow-up period ranges from 5 to 6.3 years, because we collected the follow-up data 5 years after the last baseline data collection, and the baseline data collection was performed over a period of one year and eight months. The study protocol was approved by the Medical Ethics Committee of the Erasmus MC, University Medical Center Rotterdam (MEC 2008-234 and MEC

2011-309) and the ethics committees of the participating care organizations. The study adheres to the guidelines of the Declaration of Helsinki.

Procedures

In March 2015, the client administration departments of the three participating care organizations were contacted to identify which of the participants were deceased and which of the participants were deregistered at the care organizations. Only the medical files of deceased participants who had an ID-physician as their primary physician were available, and therefore used to collect information on deaths. An ID-physician is a specialized physician who has received a 3-year postgraduate university training combining learning and working in the field of care for people with ID. In the Netherlands, this is a recognized medical specialization. Medical files of the deceased participants who received primary care from a general practitioner were not available at the care organizations.

Manner and cause of death (as explained below) were collected from the medical files by a medical student. If there were doubts about how to code and categorize a cause of death, this was discussed with a local panel of two ID-physicians until consensus was reached. The circumstances of the death and any contributing factors, were reviewed and discussed in this panel in order to determine the cause of death.

For comparing the causes of death to the general population, cause-specific mortality data of the general 50+ population were obtained online from Statline, Statistics Netherlands (Centraal Bureau voor de Statistiek, 2014).

Measurements

Personal characteristics

Information on age and sex were collected from the administrative systems. Level of ID was retrieved from psychologists' and behavioral therapists' files and was classified as borderline (intelligence quotient (IQ) = 70-80), mild (IQ = 55-70), moderate (IQ = 35-55), severe (I IQ = 25-35) or profound (IQ <25). The presence of DS, multimorbidity (presence of four or more chronic conditions (Hermans &

Evenhuis, 2014) and polypharmacy (use of five or more chronic medication) were collected from the medical files.

Mortality data

Manner of death

The manner of death was either natural or non-natural. Natural death was defined as death solely caused by spontaneous disease. A non-natural death was defined as every death directly or indirectly caused by an accident, violence, suicide, misconduct or intent by another person or any other external cause (Cohen, 2004). When a person was found dead, and death was completely unexpected and of unknown cause, the manner of dead was noted as unclear.

Causes of death

Both the immediate and primary causes of death were written down. The immediate cause of death was defined as the disease, injury or complication of which the patient died, and is the ultimate consequence of the underlying cause of death (Patja et al., 2001). The primary cause of death was defined as the disease or injury which initiated the train of morbid events leading directly to death, and is also called the underlying cause of death (World Health Organization, 2015b). For example, when a patient had colon carcinoma with hepatic metastases resulting in fatal liver failure, the immediate cause of death is liver failure, while the primary cause of death is colon cancer.

This information was collected into a standardized form. Causes were coded according to the 10th revision of the International Statistical Classification of Diseases and related health problems (ICD-10), a coded list made by the World Health Organization (WHO) (World Health Organization, 2015a). When the cause of death was unclear, which was the case in people who had died and/or been found dead unexpectedly, the cause of death was noted as unknown.

Statistical analysis

To provide insights into the differences between the deceased and deregistered participants, because we could not assess if they had deceased and with what cause, we used *t*-tests for continuous

variables, and Chi-square tests for dichotomous variables. Differences between deceased and survived participants, and participants whose medical file was available and whose medical file was not available were also assessed with independent sample *t*-tests for continuous variables, and Chi-square tests for dichotomous variables. Causes of death were categorized based on the main categories in the ICD-10. The frequencies of causes of death according to these main categories were ranked using percentages, for the total group and for participants with and without DS separately. Primary causes of death were compared against those of the general population, for the total group and per 5-year age category. Analyses were performed using the Statistical Package for Social Sciences version 21.0 (IBM Corporation, New York). A p value <0.05 was indicative for statistical significance.

Results

Characteristics of the study population

The baseline characteristics of the total study sample, the deceased, and the deregistered participants are described in Table 1. At baseline, the mean age of the total study population (n = 1050) was 61.6 ± 8.0 years (range 50-93), and 51% was male.

During the follow-up period (4.7 ± 1.4 years, 0 - 6.3 years), 207 (19.7%) participants died, and 56 (5.3%) were deregistered during the follow-up period. Of the 149 participants with DS, 54 (36.2%) died during the follow-up period, which is 26.1% of the total group of participants that died during the follow-up period. The mean age at baseline for the deceased was 65.4 ± 9.5 years, and 59.7 ± 6.2 years for the deregistered group. Deregistered people were significantly younger (t = -5.3, p < 0.001), more often female (χ^2 = 4.9, p = 0.026), had less often multimorbidity (χ^2 = 47.5, p < 0.001), and less often polypharmacy (χ^2 = 31.9, p < 0.001) than deceased participants (Table 1). When comparing deceased participants to those who survived, deceased participants were significantly older (t = 6.5, p < 0.001), more likely to have DS (χ^2 = 33.7, p < 0.001), multimorbidity (χ^2 = 47.5, p < 0.001), and polypharmacy (χ^2 = 31.9, p < 0.001).

Of the 207 participants who died within the follow-up period, 159 medical files were available to study the cause of death. Of these 159 participants, the mean age at time of death was 68.2 ± 9.8 years

(range 53-95). The participants whose files were available had more severe levels of ID ($\chi^2 = 6.59$, $p = 0.037$) and more multimorbidity ($\chi^2 = 25.2$, $p < 0.001$) compared to the people whose file was not available. Characteristics of both subgroups are presented in Appendix A.

[Table 1]

Cause-specific mortality

Manner of death

From the 159 studied deaths, autopsy was performed in 3.8% (n=6) of the deceased participants. The manner of death was natural in 139 participants (87.4%), non-natural in five participants (3.1%) and unclear in 15 participants (9.4%). Of the non-natural deaths, four deaths were due to complications after a hip fracture caused by a fall, and one due to suffocation after choking on food. The 15 participants of whom the manner of death was unclear, had died suddenly and unexpected, with no clear cause of death.

Immediate cause of death

The immediate causes of death are presented in Table 2. For the total group, the largest group of immediate cause of death was respiratory failure (43.4%). Causes of respiratory failure were pneumonia, heart failure, lung cancer or neurological disease. The second largest group was dehydration/malnutrition (20.8%), either caused by the inability or unwillingness to eat or drink. The third largest group was cardiovascular diseases (9.4%), mostly acute myocardial infarction or arrhythmias. Of the group infectious and bacterial diseases, the main cause was sepsis (83.3%). With regard to diseases of the digestive system the main cause was liver failure, and for diseases of the urinary system the main cause was kidney failure. In 15.8% of the participants the immediate cause of death was unknown.

For older adults with DS, respiratory failure also formed the largest group of immediate cause of death (73.3%), followed by infectious and bacterial diseases (4.4%), and diseases of the digestive system (4.4%). For the group of older adults with ID by other causes than DS the largest group of immediate

cause of death was also respiratory failure (31.6%), followed by dehydration/malnutrition (28.1%), and cardiovascular diseases (12.3%) (Table 2).

Primary cause of death

The primary causes of death are presented in Table 3. Respiratory diseases formed the largest group of primary cause of death (32.1%). In 80.4% (*n* = 41) this was due to pneumonia, 17.6% (*n* =9) suffered from chronic obstructive pulmonary diseases, and one person had an acute upper respiratory infection. Neoplasms (17.6%) formed the second largest group. Out of all neoplasms, 50% (*n* =14) were digestive, 14.3% (*n* = 4) were urogenital, and 7.1% (*n* = 2) respiratory. Circulatory diseases formed 8.2% of causes of death. In 12.6% of the participants, the primary cause of death was unknown.

For older adults with DS, respiratory diseases also formed the largest group of primary cause of death (51.1%), followed by dementia (22.2%). For the group of older adults with ID by other causes than DS, again respiratory diseases formed the largest group of primary causes of death (24.6%), followed by neoplasms (21.9%), and diseases of the circulatory system (11.4%) (Table 3).

[Table 2]

[Table 3]

Causes of death in the general population

In the general 50+ population, the three largest groups of primary causes of death are neoplasms (31%), circulatory diseases (28%), and respiratory diseases (9%) (Centraal Bureau voor de Statistiek, 2014). An overview of causes of death in the general population compared to our study population is presented in Figure 1. Appendix B shows the distribution of primary causes of death per 5-year age category for older adults with ID and the general population.

[Figure 1]

Discussion

In this study, the cause-specific mortality of 1050 older adults with ID was investigated. During the 5-year follow-up period, 19.7% of participants died. Of the participants with DS, 36.2% died during the follow-up period, which is 26.1% of the total group of participants that died during the follow-up period. Our findings show that older adults with ID are most likely to die from respiratory diseases, followed by neoplasms and circulatory diseases as a primary cause of death, with respiratory failure as the most common immediate cause of death, followed by dehydration/malnutrition, and cardiovascular diseases. In the general population, comparatively more deaths from neoplasms were found as a primary cause, followed by circulatory diseases and respiratory diseases respectively.

Our findings on cause-specific mortality differ to some extent from findings of earlier studies. We found a greater proportion of deaths from respiratory diseases as the primary cause of death (32.1%) compared to Heslop et al. (2014), who found 15% of the deaths caused by respiratory diseases (Heslop et al., 2014). Our study population being older and having more severe levels of ID may explain this difference. Increasing age and severity of ID is linked with more neurological damage – resulting in abnormalities in swallow function and cough reflex - and medication use (Chadwick & Jolliffe, 2009). These factors, along with increasing age, are shown to be significant factors associated with dysphagia, thereby increasing the risk of respiratory tract infections (Marik & Kaplan, 2003; Park et al., 2013).

Many people died from respiratory diseases, especially pneumonias, which is a major concern. Dysphagia, leading to aspiration, is an important etiologic factor that could lead to pneumonia (Marik & Kaplan, 2003). Earlier findings in our study cohort showed that 77.4% of participants had dysphagia, of which 51.7% had moderate to severe dysphagia (Bastiaanse et al., submitted). In 89.5% of the people with dysphagia this diagnosis had not been registered in medical files (Bastiaanse et al., submitted). In adults with ID, risks associated with having dysphagia include aspiration, choking and asphyxiation, poor nutritional status, and dehydration (Chadwick & Jolliffe, 2009). These can have many negative health consequences, such as respiratory infections, asphyxia, and death (Chadwick & Jolliffe, 2009). Creating awareness of dysphagia among direct caregivers, and proactive screening by regular swallowing risk assessments is necessary to detect dysphagia. When diagnosed, an individual feeding plan and possible

treatments should be considered for each client, to prevent respiratory tract infections and life-threatening situations.

Furthermore, we found less deaths from circulatory diseases compared to earlier studies in adults with ID (Heslop et al., 2014; Patja et al., 2001; Tyrer & McGrother, 2009). This may be explained by the great proportion of unknown causes of death we found in our study. It has been shown that the majority (80%) of sudden unexpected deaths in the general population are caused by cardiovascular problems (de la Grandmaison, 2006; Naneix, Perier, Beganton, Jouven, & Lorin de la Grandmaison, 2015); hence it may be that most sudden unexpected deaths in our cohort were related to cardiac dysfunction. Indeed, there are indications that myocardial infarctions are more often missed in people with ID than in the general population (de Winter, van den Berge, Schoufour, Oppewal, & Evenhuis, 2016; Jansen, Rozeboom, Penning, & Evenhuis, 2013). If we were to classify 80% of unknown deaths in the group of circulatory diseases, we would have percentages similar to the earlier studies.

We found fewer deaths from neoplasms (17.6%) than in the general population (31%). This is partly caused by the relative low number of deaths from neoplasms found in older adults with DS (6.7%). This may be due to their lower life expectancy. For the group older adults with ID by other causes than DS, death from neoplasms were more prevalent (21.9%) but still lower than in the general population. Because older adults with ID often have multimorbidity (Hermans & Evenhuis, 2014), there is a lot of awareness and often a careful consideration regarding the burden of the diagnostic examination in relation to the possible health gains and gains in quality of life with treatment, which may result in some underdiagnoses of neoplasms.

More than a quarter of the participants that died during our follow-up period had DS. This can be explained by the lower life expectancy of people with DS (Coppus, 2013). Of the 149 participants with DS, 54 died during our follow-up period (36.2%). In an earlier study regarding mortality in adults with DS aged 45 years and over, 21.8% of the participants died within a mean follow-up period of 4.5 years (Coppus et al., 2008). This percentage is lower than the percentage found in our study, which may be caused by our longer follow-up period and slightly older study sample.

The main primary cause of death in older adults with DS was diseases of the respiratory system, similar to the older adults with ID by other causes than DS. Dementia was the second largest primary

cause of death in older adults with DS. Dementia becomes increasingly prevalent in people with DS in later life, and was found to be associated with survival (Coppus, 2013; Coppus et al., 2008). We did not find diseases of the circulatory system to be a primary cause of death in older adults with DS. This is in line with Coppus et al. (2008), who did not find diseases of the circulatory system to be related to survival in people with DS. In previous studies within our cohort we also found that older adults with DS had a lower prevalence of cardiovascular risk factors (de Winter et al., 2012), and we found that cardiovascular risk factors were predictive for 3-year all-cause mortality in older adults with ID (de Winter et al., 2016). This lower prevalence of risk factors in people with DS may be due to a protective mechanism related to the genetic syndrome (Coppus et al., 2008).

Another important finding of this study is the great proportion of deaths with an unknown primary (12.6%) or immediate (15.8%) cause. Autopsy was performed in 3.8% of the deceased participants. This number is comparable to that found in a study regarding deaths in 17 Dutch general practices (3%) over a two year period, in which 841 deaths occurred (Oppewal & Meyboom-de Jong, 2004). With autopsy, the cause of death can be traced in around 90% of cases (Lundberg & Voigt, 1979; van Ingen & Meijer, 1994), and it can be made clear whether or not death was amenable or non-natural, and should thus be further managed by a forensic team. Often, emotional issues, administrative, and financial barriers are the main reasons for physicians not to perform autopsy (Oppewal & Meyboom-de Jong, 2001). However, it might have added value to obtain the cause of death for the medical team and caregivers in order to improve their knowledge and future care for other patients, and for the family to learn about possible hereditary disorders and help in their mourning process.

Strengths of this study are the fairly long follow-up time and the large study population. Nevertheless, our results need to be interpreted with caution. First, despite our nearly representative study sample of older adults with ID, there might have been selection bias when determining the cause-specific mortality. Older adults with borderline or mild ID who do not use any form of formal care have not been included in this cohort (Hilgenkamp, et al., 2011). Additionally, we were only able to include the medical files of participants who received care from an ID-physician; these participants had more severe levels of ID and more often multimorbidity than those receiving care from a general practitioner, and a higher percentage was female (Appendix A). Therefore, these people may have different mortality rates

and possibly other causes of death than those receiving care from a general practitioner, and these results may not be representative for those receiving care from a general practitioner. This may also be a reason for the differences in results with other studies as discussed previously. Second, information on causes of death in medical files was sometimes limited. This was either because very little was written down, or physicians did not know the cause of death because no diagnostic examination had been performed. In order to collect information on causes of death, we read the full medical files of the deceased participants, and if the cause of death was not clear, this was discussed with a panel of ID-physicians.

Implications of the study

In the Netherlands, following a death of a client the treating physician fills out two forms, form A concerning the death certificate and form B regarding the cause of death. Form B is anonymously collected by the Statistics Netherlands (Centraal Bureau voor Statistiek) to collect data on causes of death. This form is not linked to the medical files, and the cause of death is thus not always also noted in the medical files. It is recommended that a copy of this form is added to the medical file, so the registration of the causes of death is improved.

In order to reduce the most common cause of death, diseases of the respiratory system, a proactive attitude from caregivers towards screening, monitoring and treating dysphagia is needed. Chronic obstructive pulmonary diseases was also found as a cause of diseases of the respiratory system, which may be associated with the increasing number of adults with ID smoking tobacco nowadays, and the second hand smoking with the staff of the care organizations, which was often the case before this was forbidden by law. This may also be of influence on the high prevalence of respiratory diseases as a cause of death.

Another important issue this study raised is the great number of sudden unexpected deaths with unknown cause. It is important to consider autopsy in sudden unexpected deaths, because this can improve the knowledge of the medical team and caregivers and improve future care.

Conclusion

In conclusion, the cause-specific mortality in older adults with ID was investigated in this study. Diseases of the respiratory system were the main primary cause of death, with respiratory failure as the main immediate cause of death. Due to this great number of deaths from respiratory diseases, and a high prevalence of dysphagia in this population, further studies to explore the potential link between dysphagia and risks, including mortality, in older adults with ID is required. Additionally, it is important to have special attention for groups at risk for dysphagia by close monitoring and proactive care. The large number of deaths with unknown cause shows that autopsy should be considered more often, and the registration of cause of death should be improved, since this both may improve future care. Future research is required to acquire more knowledge regarding cause-specific mortality in older adults with borderline of mild ID who do not use any form of formal care.

Conflict of interest

None.

References

Bastiaanse, L. P., Kamp, v. d. A., Evenhuis, H. M., & Echteld, M. A. (submitted). *Dysphagia in older people with intellectual disabilities: results of the HA-ID study*. Submitted.

Carter, G., & Jancar, J. (1983). Mortality in the mentally handicapped: a 50 year survey at the Stoke Park group of hospitals (1930-1980). *J Ment Defic Res, 27 (Pt 2)*, 143-156.

Centraal Bureau voor de Statistiek. (2014, 2014-12-16). Open data Statline. Retrieved from http://opendata.cbs.nl/dataportaal/portal.html#_la=nl&tableId=81628NED&_theme=134

Chadwick, D. D., & Jolliffe, J. (2009). A descriptive investigation of dysphagia in adults with intellectual disabilities. *J Intellect Disabil Res, 53*(1), 29-43. doi:JIR1115 [pii]
10.1111/j.1365-2788.2008.01115.x

Cohen, B. A. J. (2004). *Forensische geneeskunde : raakvlakken tussen geneeskunst, gezondheidszorg en recht*. Assen: Van Gorcum.

Coppus, A. M. (2013). People with intellectual disability: what do we know about adulthood and life expectancy? *Dev Disabil Res Rev, 18*(1), 6-16. doi:10.1002/ddrr.1123

Coppus, A. M., Evenhuis, H. M., Verberne, G. J., Visser, F. E., Oostra, B. A., Eikelenboom, P., . . . van Duijn, C. M. (2008). Survival in elderly persons with Down syndrome. *J Am Geriatr Soc, 56*(12), 2311-2316. doi:JGS1999 [pii]
10.1111/j.1532-5415.2008.01999.x

de la Grandmaison, G. L. (2006). Is there progress in the autopsy diagnosis of sudden unexpected death in adults? *Forensic Sci Int, 156*(2-3), 138-144. doi:S0379-0738(05)00028-9 [pii]
10.1016/j.forsciint.2004.12.024

de Winter, C. F., Bastiaanse, L. P., Hilgenkamp, T. I., Evenhuis, H. M., & Echteld, M. A. (2012). Cardiovascular risk factors (diabetes, hypertension, hypercholesterolemia and metabolic syndrome) in older people with intellectual disability: results of the HA-ID study. *Res Dev Disabil, 33*(6), 1722-1731. doi:S0891-4222(12)00102-3 [pii]
10.1016/j.ridd.2012.04.010

de Winter, C. F., van den Berge, A. P., Schoufour, J. D., Oppewal, A., & Evenhuis, H. M. (2016). A 3-year follow-up study on cardiovascular disease and mortality in older people with intellectual disabilities. *Res Dev Disabil, 53-54*, 115-126.

Hermans, H., Beekman, A. T., & Evenhuis, H. M. (2013). Prevalence of depression and anxiety in older users of formal Dutch intellectual disability services. *J Affect Disord, 144*(1-2), 94-100.

Hermans, H., & Evenhuis, H. M. (2014). Multimorbidity in older adults with intellectual disabilities. *Res Dev Disabil, 35*(4), 776-783. doi:S0891-4222(14)00036-5 [pii]
10.1016/j.ridd.2014.01.022

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, 383*(9920), 889-895.

Hilgenkamp, T. I., Bastiaanse, L. P., Hermans, H., Penning, C., van Wijck, R., & Evenhuis, H. M. (2011). Study healthy ageing and intellectual disabilities: recruitment and design. *Res Dev Disabil, 32*(3), 1097-1106. doi:S0891-4222(11)00019-9 [pii]
10.1016/j.ridd.2011.01.018

Janicki, M. P., Dalton, A. J., Henderson, C. M., & Davidson, P. W. (1999). Mortality and morbidity among older adults with intellectual disability: health services considerations. *Disabil Rehabil, 21*(5-6), 284-294.

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, 57*(7), 681-685. doi:10.1111/j.1365-2788.2012.01567.x

Lundberg, G. D., & Voigt, G. E. (1979). Reliability of a presumptive diagnosis in sudden unexpected death in adults. The case for the autopsy. *Jama, 242*(21), 2328-2330.

Marik, P. E., & Kaplan, D. (2003). Aspiration pneumonia and dysphagia in the elderly. *Chest, 124*(1), 328-336.

Naneix, A. L., Perier, M. C., Beganton, F., Jouven, X., & Lorin de la Grandmaison, G. (2015). Sudden adult death: An autopsy series of 534 cases with gender and control comparison. *J Forensic Leg Med, 32*, 10-15. doi:S1752-928X(15)00019-0 [pii]
10.1016/j.jflm.2015.02.005
Oppewal, F., & Meyboom-de Jong, B. (2001). Family members' experiences of autopsy. *Fam Pract, 18*(3), 304-308.
Oppewal, F., & Meyboom-de Jong, B. (2004). Mortality in general practice--an analysis of 841 deaths during a two-year period in 17 Dutch practices. *Eur J Gen Pract, 10*(1), 13-17.
Park, Y. H., Han, H. R., Oh, B. M., Lee, J., Park, J. A., Yu, S. J., & Chang, H. (2013). Prevalence and associated factors of dysphagia in nursing home residents. *Geriatr Nurs, 34*(3), 212-217. doi:S0197-4572(13)00063-3 [pii]
10.1016/j.gerinurse.2013.02.014
Patja, K., Iivanainen, M., Vesala, H., Oksanen, H., & Ruoppila, I. (2000). Life expectancy of people with intellectual disability: a 35-year follow-up study. *J Intellect Disabil Res, 44 (Pt 5)*, 591-599.
Patja, K., Molsa, 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), 30-40. doi:jir290 [pii]
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), 898-904. doi:JIR1201 [pii]
10.1111/j.1365-2788.2009.01201.x
van de Wouw, E., Evenhuis, H. M., & Echteld, M. A. (2013). Objective assessment of sleep and sleep problems in older adults with intellectual disabilities. *Res Dev Disabil, 34*(8), 2291-2303.
van Ingen, G., & Meijer, C. J. (1994). Autopsies for general practitioners and for clinical specialists; analysis of indications. *Ned Tijdschr Geneeskd, 138*(15), 767-770.
World Health Organization. (2015a). ICD-10 Version. Retrieved from http://apps.who.int/classifications/icd10/browse/2015/en
World Health Organization. (2015b). WHO Mortality Database. Retrieved from http://www.who.int/healthinfo/cod/en/

Table 1. Baseline characteristics of the study population

	Total, n = 1,050 (100%)	**Deceased, n = 207 (19.7%)**	**Deregistered, n = 56 (5.3%)**
Age [a] (*m* ± *sd*)	61.6 ± 8.0	65.4 ± 9.5	59.7 ± 6.2*
n (%)			
50-59 yr	493 (47.0%)	61 (29.5%)	29 (51.8%)
60-69 yr	370 (35.2%)	81 (39.1%)	23 (41.1%)
70-79 yr	162 (15.4%)	50 (24.2%)	4 (7.1%)
80+ yr	25 (2.4%)	15 (7.3%)	0 (0.0%)
Sex, n (%)			
Male	538 (51.2%)	116 (56.0%)	22 (39.3%)*
Female	512 (48.8%)	91 (44.0%)	34 (60.7%)
Level of ID, n (%)			
Borderline	31 (2.9%)	2 (1.0%)	3 (5.7%)
Mild	223 (21.2%)	37 (18.5%)	10 (18.9%)
Moderate	506 (48.2%)	102 (51.0%)	29 (54.7%)
Severe	172 (16.4%)	35 (17.5%)	7 (13.2%)
Profound	91 (8.7%)	24 (12.0%)	4 (7.5%)
Down syndrome, n (%)			
No	724 (69.0%)	122 (58.9%)	27 (48.2%)
Yes	149 (14.2%)	54 (26.1%)	13 (23.2%)
Unknown	177 (16.8%)	31 (15.0%)	16 (28.6%)
Multimorbidity [b], n (%)			
No	557 (53.0%)	64 (30.9%)	37 (66.1%)*
Yes	490 (46.7%)	142 (68.6%)	18 (32.1%)
Unknown	3 (0.3%)	1 (0.5%)	1 (1.8%)
Polypharmacy [c], n (%)			
No	557 (53.0%)	79 (38.2%)	32 (57.1%)
Yes	366 (34.9%)	110 (53.1%)	11 (19.6%)
Unknown	127 (12.1%)	18 (8.7%)	13 (23.2%)

[a] Age at time of inclusion in study.

[b] Defined as the presence of ≥ 4 chronic conditions.

[c] Defined as the use of ≥ 5 chronic medication.

* Indicating a significant difference between deregistered and deceased participants, $p < 0.05$.

Table 2. Immediate causes of death, *n* (%).

Cause of death (category)	Total group (*n* = 159)	DS (*n* = 45)	ID no DS (*n* = 114)
Follow-up time, mean and range	4.7 ± 1.4, 0 – 6.3 years	4.6 ± 1.5, 0 – 6.3 years	4.8 ± 1.3, 0 – 6.3 years
Respiratory failure	69 (43.4%)	33 (73.3%)	36 (31.6%)
Dehydration / malnutrition	33 (20.8%)	1 (2.2%)	32 (28.1%)
Unknown	25 (15.8%)	5 (11.1 %)	20 (17.5%)
Cardiovascular diseases	15 (9.4%)	1 (2.2%)	14 (12.3%)
Infectious and bacterial diseases	6 (3.8%)	2 (4.4%)	4 (3.5%)
Other	5 (3.1%)	1 (2.2%)	4 (3.5%)
Diseases of the digestive system	3 (1.9%)	2 (4.4%)	1 (0.9%)
Diseases of the urinary system	3 (1.9%)	-	3 (2.6%)

ID = intellectual disability; DS = Down syndrome.

Table 3. Primary causes of death.

Total group (*n* = 159) *n* (%)	Total group (*n* = 159)	DS (*n* = 45)	ID no DS (*n* = 114)
Diseases of the respiratory system	51 (32.1%)	23 (51.1%)	28 (24.6%)
Neoplasms	28 (17.6%)	3 (6.7%)	25 (21.9%)
Unknown	20 (12.6%)	3 (6.7%)	17 (14.9%)
Diseases of the circulatory system	13 (8.2%)	-	13 (11.4%)
Dementia	13 (8.2%)	10 (22.2%)	3 (2.6%)
Diseases of the nervous system	11 (6.9%)	3 (6.7%)	8 (7.0%)
Other	11 (6.9)%	2 (4.4%)	9 (7.9%)
External cause	7 (4.4%)	1 (2.2%)	6 (5.3%)
Diseases of the digestive system	5 (3.1%)	-	5 (4.4%)

ID = intellectual disability; DS = Down syndrome.

Appendix A. Characteristics of the deceased participants.

	Deceased participants whose file was available[a] (n=159)	**Deceased participants whose file was not available (n=48)**
Age (in years) at time of death, mean ± SD	68.2 ± 9.8	67.5 ± 8.2
Sex		
Male, n (%)	81 (50.9%) *	35 (72.9%) *
Female, n (%)	78 (49.1%) *	13 (27.1%) *
Level of ID		
Mild, n (%)	26 (16.7%) *	13 (29.5%) *
Moderate, n (%)	78 (50.0%) *	24 (54.5%) *
Severe, n (%)	52 (33.3%) *	7 (15.9%) *
Diagnosed with Down syndrome, n (%)	45 (31.2%)	9 (28.1%)
Presence of multimorbidity, n (%)	123 (77.8%) *	19 (39.6%) *
Presence of polypharmacy, n (%)	91 (60.3%)	19 (50.0%)

*** Indicating a significant difference between the two groups, with $p < 0.05$.**

[a] Medical files of deceased participants who received medical care by a specialized ID-physician were available for the study. Medical files of participants who received care from a general practitioner were not available.

Appendix B. Percentages of primary causes of death per 5-year age category for older adults with ID and the general population.

Cause of death (category)	50 – 54 yrs		55 – 59 yrs		60 – 64 yrs		65 – 69 yrs		70 – 74 yrs		75 – 79 yrs		80 – 84 yrs		85– 90 yrs		90 – 95 yrs		95+ yrs	
	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen	ID	Gen
%	*n* = 2	*n* = 3728	*n*= 34	*n* = 5558	*n* = 35	*n* = 18180	*n* = 23	*n* = 11839	*n* = 21	*n* = 13265	*n* = 16	*n* = 17582	*n* = 19	*n* = 24019	*n* = 3	*n* = 25564	*n* = 5	*n* = 18146	*n* = 1	*n* = 7083
Diseases of the respiratory system	100	3.3	26.5	4.7	51.4	6.0	30.4	6.7	23.8	8.6	12.5	9.4	26.3	10.3	-	9.9	40.0	10.4	100	10.9
Neoplasms	-	50.7	17.6	52.4	8.6	53.7	8.7	51.5	42.9	46.5	12.5	37.1	15.8	27.8	66.7	18.9	20.0	12.4	-	6.8
Diseases of the circulatory system	-	16.9	5.9	16.6	8.6	18.0	8.7	21.0	4.8	22.8	18.8	26.8	5.3	30.1	-	33.7	40.0	34.6	-	33.9
Mental & behavioral disease (incl. dementia)	-	1.9	17.6	2.2	11.4	1.7	4.3	1.9	-	2.8	12.5	5.2	-	8.1	-	11.5	-	14.0	-	15.1
Diseases of the nervous system	-	3.0	5.9	3.2	11.4	3.2	8.7	3.2	9.5	4.0	-	5.1	5.3	5.8	-	5.6	-	5.2	-	4.8
Other	-	11.4	8.7	10.8	-	10.0	8.6	9.9	4.8	10.2	6.2	10.7	15.9	11.6	33.3	13.5	-	15.6	-	21.3
External cause	-	9.3	2.9	6.6	2.9	3.8	4.3	2.5	4.8	2.3	6.2	2.8	10.6	3.0	-	3.8	-	4.6	-	4.5
Diseases of the digestive system	-	3.6	5.9	3.5	-	3.5	4.3	3.3	4.8	2.8	-	3.0	5.3	3.2	-	3.3	-	3.2	-	3.1

ID = Intellectual disabilities; Gen = General population.