PEER REVIEW HISTORY

ARTICLE DETAILS

TITLE (PROVISIONAL)	The Obscuring Effect of Coding Developmental Disability as the Underlying Cause of Death on Mortality Trends for Adults with Developmental Disability: A Cross-Sectional Study Utilizing U.S. Mortality Data from 2012 to 2016
AUTHORS	Landes, Scott; Stevens, James; Turk, Margaret

VERSION 1 – REVIEW

REVIEWER	Professor Pauline Heslop University of Bristol, England
REVIEW RETURNED	29-Oct-2018

GENERAL COMMENTS	This is an interesting and well-written paper describing a well-conceived study about potential inaccuracies in recording causes of death of people with developmental disabilities. In my view it makes an original and valuable contribution to the literature. Overall, the authors should be commended for the strength of the paper. There are a few ways in which I think it could be improved ready for publication: 1. it would be helpful for the authors to describe more fully the definition of 'developmental disability'. The definition given is drawn from one medical textbook, yet there are several different ways that developmental disabilities can be described and categorised, so some further brief clarification about this would be helpful. 2. More information is required about the sequential UCOD revision process and how the revisions to the death certificates were made, with exemplars to illustrate process. In the discussion, the authors comment that 'Despite best efforts and available data, it is not possible to verify the accuracy of the revised UCOD without access to the medical records of the decedent and the medical personnel that completed the death certificate.' What are the best efforts and available data that the authors are referring to? A more detailed description of the revision process would be beneficial here. 3. The study population is drawn from across the USA, so one of the limitations to be acknowledged is that there may be some geographical variations in conventions about reporting deaths that still need to be identified and addressed. 4. The role of the World Health Organisation in providing international standardisation of medical certificates of causes of deaths and how they are completed could usefully be added and reflected on in the discussion about the way forward for addressing the findings.

REVIEWER	PHILIP MCCALLION Temple University Philadelphia USA
REVIEW RETURNED	04-Nov-2018

GENERAL COMMENTS	There is a growing issue and concern that the ability to identify people with developmental disabilities is declining in large public datasets so at first glance the desire to reduce identification of developmental disabilities as UCOD does present some concerns given data on the differences in mortality and morbidity for people with intellectual and developmental disabilities found in other studies. However I believe the authors have addressed how developmental disabilities may be more appropriately noted. That said I think there is a need in the discussion to more strongly reinforce and encourage the noting of developmental disabilities. There have been a series of articles in the last few years addressing mortality and premature death in people with intellectual and developmental disabilities not cited here that would strengthen the introduction The methods are not described at a level here that would permit others to replicate the findings (as required by the journal)

VERSION 1 – AUTHOR RESPONSE

Reviewer: 1
Reviewer Name: Professor Pauline Heslop
Institution and Country: University of Bristol, England
Please state any competing interests or state 'None declared': None declared

Please leave your comments for the authors below

This is an interesting and well-written paper describing a well-conceived study about potential inaccuracies in recording causes of death of people with developmental disabilities. In my view it makes an original and valuable contribution to the literature.

Overall, the authors should be commended for the strength of the paper. There are a few ways in which I think it could be improved ready for publication.

1. It would be helpful for the authors to describe more fully the definition of 'developmental disability'. The definition given is drawn from one medical textbook, yet there are several different ways that developmental disabilities can be described and categorised, so some further brief clarification about this would be helpful.

• We added language from the CDC's description of developmental disability to our definition. As our manuscript is focused on ICD-10 coding of underlying cause of death, we also summarized a more nuanced, but brief, definition of developmental disability from the WHO on page 4.

"Developmental disabilities comprise a diverse array of conditions that originate at birth or during the early developmental part of life - intellectual disability, cerebral palsy, Down syndrome, autism, as well as other chromosomal abnormalities.15 These disabilities are attributable to physical, learning, language, or behavioral impairments, directly impact daily functioning, and extend across a person's life course.15,16 Developed by the World Health Organization (WHO) to facilitate global medical communication and research, the International Statistical Classification of Diseases and Related Health Problems, Tenth Revision (ICD-10) classifies developmental disabilities variably as mental and behavioral disorders in the cases of intellectual disability, cerebral palsy, and autism, or as chromosomal or congenital abnormalities in the cases of Down syndrome and spina bifida.17"

2. More information is required about the sequential UCOD revision process and how the revisions to the death certificates were made, with exemplars to illustrate process. In the discussion, the authors comment that 'Despite best efforts and available data, it is not possible to verify the accuracy of the revised UCOD without access to the medical records of the decedent and the medical personnel that completed the death certificate.' What are the best efforts and available data that the authors are referring to? A more detailed description of the revision process would be beneficial here.

• To address concerns with the robustness of the description of our revisions, we expanded on our description of our sequential UCOD revision process and provided illustrative examples of this process on pages 7-8.

"If the UCOD listed on the death certificate was not coded as a developmental disability, it was accepted as valid and retained for analysis. In the event that the developmental disability was identified as the UCOD, it was revised by the study team utilizing a sequential UCOD revision process.8,12 We chose to identify a singular UCOD code to construct a straightforward methodology, recognizing some recent studies contend that multiple morbidity data could be helpful, especially in understanding complex conditions with co-morbidities, as in dementia and Alzheimer disease.33,34 In the instances when the UCOD was revised, we identified a valid UCOD by working sequentially from the last line to the first line of Part I of the death certificate, moving from the first to the last listed code per line. ICD-10 codes that the U.S. Centers for Disease Control and Prevention (CDC) states are not to be used as the UCOD were not considered valid options for the revised UCOD.35 In addition, ICD-10 Chapter 18 R-codes were only utilized for the UCOD if no other valid UCOD was listed in Part I of the death certificate.2 As they record co-morbidities present at the time of death that were not part of the sequence of events leading to death, ICD-10 codes in Part II of the death certificate were not considered in the revision process.

The following examples demonstrate our sequential UCOD revision process. The first example is a decedent who had J96.9 (Respiratory failure, unspecified) recorded on Line 1, Position 1; J69.0 (Pneumonitis due to inhalation of food and vomit) recorded on Line 2, Position 1; and F79 (Unspecified intellectual disability) on Line 3, Position 1; with F79 identified as the UCOD. In this instance, working sequentially from the last line (Line 3), we would dismiss F79 as it is not a valid UCOD, and identify J69.0 on Line 2 as the first listed valid UCOD. A second example, involving a more complex revision process, is for a decedent who had T17.9 (Foreign body in respiratory tract, part unspecified) recorded on Line 1, Position 1; W80 (Inhalation and ingestion of other objects causing obstruction of respiratory tract – commonly termed choking) recorded on Line 1, Position 2; G80.9 (Cerebral palsy, unspecified) recorded on Line 2, Position 1; and R56.8 (Other and unspecified convulsions) listed in Line 3, Position 1; with G80.9 identified as the UCOD. In this instance, we would initially identify R56.8 as the UCOD. However, as R-codes are unspecified causes and not useful for public health,2 we would continue looking for a more valid UCOD. We would dismiss G80.9 as it is not a valid UCOD, dismiss T17.9 as CDC rules do not allow this ICD-10 code to be an UCOD,35 and identify W80 as the valid UCOD, superseding R56.8. In instances where no valid UCOD was present in Part I of the death certificate, as in the cases where the death certificate only listed an ICD-10 code for a developmental disability, we identified the UCOD as 'Unknown.' "

• In the discussion section, we dropped the term "best efforts" and "available data" and more clearly detail the long-term limitation of our strategy page 14:

"Although sequentially revising the UCOD for U.S. death certificates that identify a developmental disability as the UCOD increased the reliability of mortality trends for this population, it is not possible to verify the accuracy of the revised UCOD without access to the medical records of the decedent and the medical personnel that completed the death certificate."

3. The study population is drawn from across the USA, so one of the limitations to be acknowledged is that there may be some geographical variations in conventions about reporting deaths that still need to be identified and addressed.

• We added information regarding possible state level variation in the certification process, and mentioned that our study does not account for this variation to the discussion section on pages 14-15.

“Instead, focus should be on ensuring that at the time of death, the medical certifier of the death certificate, the individual completing the medical portion of the death certificate, accurately identifies the disease process or injury, other than developmental disability, that initiated the chain of events leading to death. In all U.S. states, this would be wither the attending physician present at the time of death or the decedent’s personal physician, with some states allowing the chief medical officer of medical facilities to certify.43,44 In instances when death occurs without an attending physician present, cause of death is unknown, or death occurs by accident, suicide, or homicide, the individual responsible for certifying the cause of death would be the medical examiner or coroner, with responsibility varying by U.S. state.43,45 In addition, it is unlikely that the death certificates of all decedents with a developmental disability included an ICD-10 code for developmental disability. Based on inconsistencies regarding the proper location to code developmental disability on death certificates we observed in this study, it is obvious that there is confusion regarding where to record a developmental disability on the death certificate. This confusion may result in the medical certifier not recording the developmental disability on the death certificate at all. Thus, the results from this study only describe mortality trends for decedents with developmental disability who had their disability recorded on their death certificate, and do not account for possible state level variation in the cause of death certification process.”

4. The role of the World Health Organisation in providing international standardisation of medical certificates of causes of deaths and how they are completed could usefully be added and reflected on in the discussion about the way forward for addressing the findings.

• We now detail WHO’s development of ICD standards in the introduction on page 4.

“Developed by the World Health Organization (WHO) to facilitate global medical communication and research, the International Statistical Classification of Diseases and Related Health Problems, Tenth Revision (ICD-10) defines developmental disabilities variably as mental and behavioral disorders in the cases of intellectual disability, cerebral palsy, and autism, or as chromosomal or congenital abnormalities in the cases of Down syndrome and spina bifida.17”

• In addition, we added description of the WHO’s role in: encouraging the continued practice of coding developmental disability as UCOD (page 13); providing guidelines for recording cause of death on the death certificate (page 15); and argue that change is needed at a national and international level (page 15).

“Unfortunately, current CDC instructions and WHO guidelines for completing death certificates permit identifying a developmental disability as the UCOD in at least two known instances.41,42 ICD-10 codes for intellectual disability (F70-79) are permitted as UCOD in instances that the actual UCOD is unknown. In addition, Down syndrome is suggested as the UCOD in the event the death certificate indicates unspecified dementia or Alzheimer disease.”

“In order to fully address these limitations, it is necessary to move beyond retrospective remedies, and formally change instructions for coding a developmental disability on the death certificate. As CDC instructions on cause of death coding are developed based upon and in cooperation with WHO

guidelines,46,47 change is needed at both a national and international level. Due to concerns that population representative datasets rarely include adults with developmental disability, which limits surveillance of morbidity and mortality trends among this population,48,49 it is imperative to continue recording developmental disabilities on death certificates. Per CDC and WHO guidelines, Part II of the death certificate is intended for the recording of co-morbidities that were present at the time of death, but were not part of the sequence of events leading to death.41,42 To ensure that developmental disabilities are recorded on the death certificate, but not identified as the UCOD, instructions for completing death certificates should specify that developmental disabilities should not be recorded in Part I or permitted as the UCOD. Instead, developmental disabilities should be recorded in Part II of the death certificate.10,11,13 Evidence from this study supports this recommendation. Among the death certificates that recorded a developmental disability in Part I (with or without also recording a developmental disability in Part II), 82% coded developmental disability as the UCOD. In contrast, among the death certificates that recorded a developmental disability only in Part II, 19% coded developmental disability as the UCOD. This proposed change would preserve the ability to surveil mortality trends for this population by ensuring the recording of the developmental disability on the death certificate while minimizing the possibility a developmental disability is coded as the UCOD. Changing the instructions for coding developmental disability would also increase the accuracy of mortality data for adults with developmental disability by deterring the coding of a developmental disability as an UCOD. As a result, the death certificates of this population would more accurately represent actual mortality trends, allowing for better-informed public health and preventive care efforts to reduce premature mortality for this population."

Reviewer: 2
Reviewer Name: PHILIP MCCALLION
Institution and Country: Temple University Philadelphia USA

Please state any competing interests or state 'None declared': None declared

Please leave your comments for the authors below

1. There is a growing issue and concern that the ability to identify people with developmental disabilities is declining in large public datasets so at first glance the desire to reduce identification of developmental disabilities as UCOD does present some concerns given data on the differences in mortality and morbidity for people with intellectual and developmental disabilities found in other studies. However I believe the authors have addressed how developmental disabilities may be more appropriately noted. That said I think there is a need in the discussion to more strongly reinforce and encourage the noting of developmental disabilities.

• To emphasize this important point, we added the following text on page 15.

"Due to concerns that nationally representative datasets rarely include adults with developmental disability, which limits surveillance of morbidity and mortality trends among this population,48,49 it is imperative to continue recording developmental disabilities on death certificates."

2. There have been a series of articles in the last few years addressing mortality and premature death in people with intellectual and developmental disabilities not cited here that would strengthen the introduction.

• We now briefly summarize recent research detailing increased longevity, but continued premature mortality among adults with developmental disability on page 5.

Longevity for individuals with developmental disabilities improved since the early 1960s, resulting in a larger percentage of adults in this population now living into their 60s.18-22 Despite these gains, on average, adults with developmental disability die 20-25 years earlier than those in the general population.18,19,21

3. The methods are not described at a level here that would permit others to replicate the findings (as required by the journal)

• To address concerns with the robustness of the description of our revisions, we expanded on our description of our sequential UCOD revision process and provided illustrative examples of this process on pages 7-8.

“If the UCOD listed on the death certificate was not coded as a developmental disability, it was accepted as valid and retained for analysis. In the event that the developmental disability was identified as the UCOD, it was revised by the study team utilizing a sequential UCOD revision process.8,12 We chose to identify a singular UCOD code to construct a straightforward methodology, recognizing some recent studies contend that multiple morbidity data could be helpful, especially in understanding complex conditions with co-morbidities, as in dementia and Alzheimer disease.33,34 In the instances when the UCOD was revised, we identified a valid UCOD by working sequentially from the last line to the first line of Part I of the death certificate, moving from the first to the last listed code per line. ICD-10 codes that the U.S. Centers for Disease Control and Prevention (CDC) states are not to be used as the UCOD were not considered valid options for the revised UCOD.35 In addition, ICD-10 Chapter 18 R-codes were only utilized for the UCOD if no other valid UCOD was listed in Part I of the death certificate.2 As they record co-morbidities present at the time of death that were not part of the sequence of events leading to death, ICD-10 codes in Part II of the death certificate were not considered in the revision process.

The following examples demonstrate our sequential UCOD revision process. The first example is a decedent who had J96.9 (Respiratory failure, unspecified) recorded on Line 1, Position 1; J69.0 (Pneumonitis due to inhalation of food and vomit) recorded on Line 2, Position 1; and F79 (Unspecified intellectual disability) on Line 3, Position 1; with F79 identified as the UCOD. In this instance, working sequentially from the last line (Line 3), we would dismiss F79 as it is not a valid UCOD, and identify J69.0 on Line 2 as the first listed valid UCOD. A second example, involving a more complex revision process, is for a decedent who had T17.9 (Foreign body in respiratory tract, part unspecified) recorded on Line 1, Position 1; W80 (Inhalation and ingestion of other objects causing obstruction of respiratory tract – commonly termed choking) recorded on Line 1, Position 2; G80.9 (Cerebral palsy, unspecified) recorded on Line 2, Position 1; and R56.8 (Other and unspecified convulsions) listed in Line 3, Position 1; with G80.9 identified as the UCOD. In this instance, we would initially identify R56.8 as the UCOD. However, as R-codes are unspecified causes and not useful for public health,2 we would continue looking for a more valid UCOD. We would dismiss G80.9 as it is not a valid UCOD, dismiss T17.9 as CDC rules do not allow this ICD-10 code to be an UCOD,35 and identify W80 as the valid UCOD, superseding R56.8. In instances where no valid UCOD was present in Part I of the death certificate, as in the cases where the death certificate only listed an ICD-10 code for a developmental disability, we identified the UCOD as ‘Unknown.’ ”

FORMATTING AMENDMENTS (if any)
Required amendments will be listed here; please include these changes in your revised version:
1. Please embed your DATA SHARING STATEMENT in your main document file as shown in ScholarOne.

• We added a data sharing statement on page 17.

“Data sharing statement: All data is publicly available from the Centers for Disease Control and Prevention (CDC).”

2. Patient and Public Involvement:
Authors must include a statement in the METHODS section of the manuscript under the sub-heading 'Patient and Public Involvement'.

This should provide a brief response to the following questions:

How was the development of the research question and outcome measures informed by patients’ priorities, experience, and preferences?
How did you involve patients in the design of this study?
Were patients involved in the recruitment to and conduct of the study?
How will the results be disseminated to study participants?
For randomised controlled trials, was the burden of the intervention assessed by patients themselves?
Patient advisers should also be thanked in the contributorship statement/acknowledgements.

If patients and or public were not involved please state this

• We added a subheading and statements to address patient and public involvement to the end of the methods section on page 9.

• Patient and Public Involvement

“Patients and public were not involved in any aspect of this study, inclusive of development of research question and design, outcomes measures, analytic plan, and interpretation of results. As all study participants were deceased and data is anonymous, results cannot and will not be disseminated to study participants.”

OTHER EDITS
• We added subheadings to the Methods section.

VERSION 2 – REVIEW

REVIEWER	Professor Pauline Heslop University of Bristol, England
REVIEW RETURNED	04-Jan-2019

GENERAL COMMENTS	The authors have appropriately addressed the reviewers' concerns and strengthened the paper as a result. I consider that the paper is now suitable for publication and that it will make an important contribution to BMJ Open.