BMJ Open

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Journal:	*BMJ Open*
Manuscript ID	bmjopen-2018-026614
Article Type:	Research
Date Submitted by the Author:	11-Sep-2018
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Keywords:	Developmental disability, Underlying cause of death, Mortality trends

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

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Word count: 2,964

Abstract

Objective: To determine whether coding a developmental disability as the underlying cause of death obscures mortality trends of adults with developmental disability.

Design: National Vital Statistics System 2012-2016 U.S. Multiple Cause-of-Death Mortality files.

Setting: United States.

Participants: Adults with a developmental disability indicated on their death certificate aged 18 through 103 at the time of death. The study population included 33,154 adults who died between January 1, 2012 and December 31, 2016.

Primary outcome and measures: Decedents with a developmental disability coded as the underlying cause of death on the death certificate were identified utilizing an ICD-10 code for intellectual disability, cerebral palsy, Down syndrome, or other developmental disability. Death certificates that coded a developmental disability as the underlying cause of death were revised utilizing a sequential underlying cause of death revision process.

Results: There were 33,154 decedents with developmental disability: 7,901 with intellectual disability, 11,895 with cerebral palsy, 9,114 with Down syndrome, 2,479 with other developmental disabilities, and 1,765 with multiple developmental disabilities. Among all decedents, 48.5% had a developmental disability coded as the underlying cause of death, obscuring higher rates of choking deaths among all decedents, and dementia and Alzheimer disease among decedents with Down syndrome.

Conclusion: Death certificates that recorded the developmental disability in Part I of the death certificate were more likely to code disability as the underlying cause of death. While revising these death certificates provides a short-term corrective to mortality trends for this population, the severity and extent of this problem warrants a long-term change involving more precise instructions to record developmental disabilities only in Part II of the death certificate.

Strengths and limitations of this study

- This study revises death certificates that had developmental disability coded as underlying cause of death.
- This study provides evidence that coding developmental disability as underlying cause of death obscures often preventable, diseases or injuries other than disability that initiated the sequence of events leading to death.
- The high percentage of death certificates coded with a developmental disability as the underlying cause of death reveals the need to revise death certificates for adults with developmental disability prior to reporting mortality trends.
- Since the study population only includes decedents who had a developmental disability recorded on their death certificate, it is not inclusive of all decedents with a developmental disability in the U.S

- Limitations to retrospective revising of death certificates highlight the need for a long-term solution involving formal changes to policies guiding coding of developmental disability on death certificates.

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

Public health and preventive care efforts aimed at reducing premature mortality rely on population mortality data from death certificates.[1,2] Thus, it is imperative that death certificates accurately identify a valid and informative underlying cause of death (UCOD), the disease or injury that initiated the causal sequence of events leading to death. Beyond general concerns with the overall accuracy of death certificate data,[3-5] researchers express specific concern regarding the frequency in which the UCOD identifies a cause that is not valid or informative for public health and preventive care efforts aimed at reducing premature mortality.[2,6,7] One such cause of concern occurs when decedents with developmental disability have their disability coded as their UCOD.[8-14]

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 directly impact daily living via impairments in the areas of mobility, language, learning, self-care, and independent living, and extend across a person's life course.[15] As they are lifelong conditions, there is increasing consensus that developmental disabilities *should not* be considered a valid UCOD, as doing so prohibits identification of the preventable medical cause of death, and is not advantageous to public health or preventive care efforts.[2,8-16] Instead, the goal in coding the UCOD on death certificates for persons with developmental disability should be to identify the specific, and often preventable, disease or injury other than disability that initiated the sequence of events leading to death.

Cerebral palsy offers insight to the limitations of coding a developmental disability as an UCOD. Cerebral palsy describes a group of various permanent non-progressive motor disorders which stem from an injury to or malfunction in the developing brain and vary in severity, cause, and presentation.[17] Besides the motor disorders that cause activity limitations, there can be associated conditions that accompany the diagnosis of cerebral palsy such as epilepsy or problems with sensation, cognition, communication, and behavior. People with cerebral palsy also have risk factors for general health conditions shared with the general population, such as chronic heart disease.[18] Due to a higher prevalence of gastroesophageal reflux and/or dysphagia among adults with moderate to severe forms of cerebral palsy, there is increased risk among this population for aspiration on either saliva, food, or gastric contents leading to pneumonia or pneumonitis, and subsequent death.[19,20] In the event that an individual with cerebral palsy aspirates on food, develops pneumonia, and dies, it is not useful to identify the UCOD as cerebral palsy. Doing so conceals the fact that death was due to aspiration and subsequent development of pneumonia, and fails to differentiate this causal pathway from other causal pathways such as arteriosclerotic heart disease leading to acute myocardial infarction and death. As this example illustrates, coding a developmental disability as the UCOD is uninformative,[2,21,22] and, as Trollor and colleagues[12] contend, "obscures" actual mortality trends related to preventable conditions that need increased attention in public health or preventive care efforts.

Prior studies report the prevalence and obscuring effect of coding a developmental disability as the UCOD. Internationally, 16% of persons with developmental disability in general,[12] and between 34% and 56% of persons with cerebral palsy in particular,[8,23] had their disability coded as their UCOD. Among the U.S. population, 20% of decedents with an

intellectual disability,[10] and 21% of decedents with Down syndrome,[24] had their disability coded as their UCOD. Two studies, from Canada[14] and Australia,[12] that mention the obscuring effect of this practice on mortality trends report a substantial increase in deaths from respiratory disease after revising death certificates coded with a developmental disability as the UCOD. To date, researchers have not detailed the obscuring effect of coding a developmental disability as the UCOD on mortality trends for adults with developmental disability in the U.S. To determine the severity of this problem, we compare mortality trends for adult decedents with developmental disability using the originally reported and a revised UCOD from U.S. death certificate data.

Methods

Death certificate data for this study are from the National Vital Statistics System 2012-2016 U.S. Multiple Cause-of-Death Mortality files. As mortality coding in the U.S. converted to ICD-10 in 2009,[25] this study included the death certificates for adults aged 18-103 at the time of death that recorded an ICD-10 code for a developmental disability as the UCOD, or as a multiple cause of death (other co-morbidities present at time of death) - intellectual disability (F70-79), cerebral palsy (G80), Down syndrome (Q90), and other developmental disabilities (F80-89, Q91-99). This resulted in a sample of 33,154 death certificates over the 5-year period.

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.[26,27] In the instances when the UCOD was revised, a valid UCOD was located

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 CDC states are not to be used as the UCOD were not considered valid options for the revised UCOD.[28] In addition, ICD-10 Chapter 18 R-codes were only utilized if no other valid UCOD was listed in Part I of the death certificate.[2]

All analysis was conducted using STATA Version 15.0 (College Station, TX). Results are presented by developmental disability group due to disparate cause of death trends – intellectual disability, cerebral palsy, Down syndrome, other developmental disability, and multiple developmental disabilities. For each group, we report: UCOD percentages by ICD-10 chapter code for all death certificates prior to and after revisions; changes in the cause of death rank order; and percentage point differences between original and revised data. We also detail the leading specific underlying causes of death within ICD-10 chapters reflecting the most drastic changes.

Results

Full comparative results of the original and revised UCOD by ICD-10 Chapter code are presented for each disability group in Tables 1-5. Though detailed in the text, information on specific underlying causes of death per ICD-10 Chapter are not included in the tables. We compared results from aggregated data to results from each specific year prior and subsequent to revisions. The only dissimilarity was a slight increase in the percentage of decedents with Down syndrome who had Q90 coded as their UCOD beginning in 2014.

There were 7,901 decedents with intellectual disability, 11,895 decedents with cerebral palsy, 9,114 decedents with Down syndrome, 2,479 decedents with other developmental disabilities, and 1,765 decedents with multiple developmental disabilities. The UCOD was coded

as a developmental disability, and thus was revised, on the death certificates of 48.52% of all decedents with a developmental disability, but varied by disability type - 25.88% of decedents with intellectual disability, 59.53% of decedents with cerebral palsy, 58.99% of decedents with Down syndrome, 26.42% of decedents with other developmental disabilities, and 52.69% of decedents with multiple developmental disabilities.

As expected, after revising the death certificates, there was a steep decline in the percentage and rank order of decedents in each disability group with an UCOD in the ICD-10 chapter inclusive of their disability. Due to these revisions, the percentages increased for all other ICD-10 chapters for all disability groups. We detail the most remarkable changes.

Across groups, the percentage of deaths caused by diseases of the respiratory system, ICD-10 Chapter X, rose dramatically after the revision: from 15.37% to 23.67% for decedents with intellectual disability; from 8.95% to 29% for decedents with cerebral palsy; from 8.56% to 27% for decedents with Down syndrome; from 8.67% to 16.3% for decedents with other developmental disabilities; and from 12.01% to 29.24% for decedents with multiple developmental disabilities. As a result, respiratory deaths rose from the 3rd to the 1st leading cause of death for decedents with intellectual disability, cerebral palsy, Down syndrome, and multiple developmental disabilities; and from the 5th to the 2nd leading cause of death for decedents with other developmental disabilities such as autism.

For all disability groups, the most prevalent underlying causes of death identified among diseases of the respiratory system were J69 Pneumonitis due to inhalation of food/vomit and J18.9 Pneumonia, unspecified organism. The percentage of deaths from pneumonitis due to inhalation of food/vomit increased from: 5.73% to 9.57% among decedents with intellectual disability; 3.14% to 10.08% among decedents with cerebral palsy; 5.43% to 9.84% among

decedents with Down syndrome; 3.03% to 5.77% among decedents with other developmental disabilities; and 5.72% to 12.18% among decedents with multiple developmental disabilities. The percentage of deaths from pneumonia, unspecified organism rose from: 4.38% to 7.1% among decedents with intellectual disability; 2.3% to 8.53% among decedents with cerebral palsy; 0.07% to 10.19% among decedents with Down syndrome; 2.02% to 4.4% among decedents with other developmental disabilities; and 2.21% to 8.67% among decedents with multiple developmental disabilities.

To varying degrees, deaths from external causes of mortality, ICD-10 Chapter XX, increased in percent and rank order for four of the disability groups: from 2.67% to 7.53% (rank order increase from 9 to 3) for decedents with intellectual disability; from 1.65% to 5.89% (rank order increase from 9 to 5) for decedents with cerebral palsy; from 7.34% to 8.75% (rank order increase from 7 to 4) for decedents with other developmental disabilities; from 1.76% to 6.06% (rank order increase from 10 to 6) for decedents with multiple developmental disabilities. While the percentage of deaths from external causes of mortality rose from 2.79% to 3.86% for decedents with Down syndrome, the rank order dropped one position from 5 to 6.

For each disability group, the most prevalent underlying causes of death in the external causes chapter were W78-80, Accidental inhalation and ingestion of food or other objects causing obstruction of the respiratory tract. The percentage of deaths from accidental choking increased: from 0.1% to 4.80% among decedents with intellectual disability; from 0.02% to 3.76% among decedents with cerebral palsy; from 1.32% to 2.28% among decedents with Down syndrome; from 3.19% to 4.48% among decedents with other developmental disabilities; and from 0.17% to 4.09% among decedents with multiple developmental disabilities.

One other remarkable finding among all disability groups regarded the percentage of decedents who had either an unknown or an imprecise UCOD. Unknown causes of death were recorded when no other ICD-10 code was listed in Part I of the death certificate other than disability. Imprecise underlying causes occurred when no other ICD-10 code was listed in Part I of the death certificate other than the developmental disability and/or a single or multiple R code from Chapter XVIII. As a result, the UCOD was either unknown or imprecise for: 4.96% of decedents with intellectual disability; 17.17% of decedents with cerebral palsy; 8.88% of decedents with Down syndrome; 8.51% of decedents with other developmental disabilities; and 11.51% of decedents with multiple developmental disabilities. This means that even after revision, the death certificate provided little to no indication of the disease or injury that initiated the sequence of events leading to the death of these individuals.

Particular to decedents with Down syndrome, deaths from diseases of the nervous system, ICD-10 Chapter VI, rose from 5.31% to 13.18% (rank order increase from 4 to 3). The most prevalent UCOD for decedents with Down syndrome in this chapter, G30.9 Alzheimer disease, unspecified, increased from 3.27% to 9.5%. In addition, contrary to trends among the other disability groups, deaths from mental, behavioral, and neurodevelopmental disorders, ICD-10 Chapter V, rose from 1.73% to 11.82% (rank order increase from 10 to 4). The most prevalent cause of death in this chapter, F03 Unspecified dementia, increased from 1.62% to 11.4%.

Discussion

In 2012-2016 U.S. death certificate data, the coding of a developmental disability as the UCOD is widespread, but varied among specific disability groups. Results from this study expose that the practice of coding a developmental disability as the UCOD on a death certificate obscures the higher prevalence of choking related deaths (coded as either pneumonitis due to

inhalation of food/vomit, or accidental choking) among all developmental disability groups, and obscures dementia and Alzheimer disease related deaths among decedents with Down syndrome.

In instances when death certificates that code a developmental disability as the UCOD are not revised, population mortality trends for persons with various types of developmental disability are likely to be inaccurate and may, therefore, misinform public health and preventive care efforts aimed at preventing premature mortality. The primary strength of this study is that results demonstrate that the prominence of choking related deaths as a mortality risk among persons with developmental disability was severely underestimated prior to revision. As revision of the UCOD revealed, similar to the older adult population,[29-31] careful attention is needed to attend to the heightened risk of preventable choking related deaths among persons with developmental disability.[15,32,33] Thus, it is of the utmost importance that public health and preventive care efforts focus on management of the swallowing function among this population. Physicians conducting routine exams should be screening for swallowing disorders, abnormalities in the cough and gag reflex, as well as respiratory distress.[15,33] Mortality data based upon original death certificates, often coding developmental disability as UCOD, fail to reveal this important mortality trend.

Unfortunately, current guidelines for completing death certificates in the U.S. permit identifying a developmental disability as the UCOD in at least two known instances. ICD-10 codes for intellectual disability (F70-79) are permitted as UCOD in instances that the actual UCOD is not known.[34] In addition, the CDC suggests coding Down syndrome as the UCOD in the event the death certificate indicates unspecified dementia or Alzheimer disease. Continued permission of the practice of coding a developmental disability as the UCOD demands both short-term and long-term redress. As this study highlights, analysis of mortality trends for adults

with developmental disability must recognize the obscuring effect of coding a developmental disability as the UCOD and take the necessary steps to revise the UCOD on these death certificates prior to reporting findings. The sequential UCOD revision process utilized in this study reduces obfuscation of mortality trends among populations with developmental disabilities in the short-term.

While revising death certificates for adults with developmental disability is a necessary short-term solution, the primary limitations of this study indicate that retrospective revision is not a sufficient long-term solution. The first limitation regards accuracy of revised death certificates. 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. Although the only remedy present at the time, the method of retrospectively revising the UCOD should not be viewed as a sustainable long-term strategy for surveilling mortality trends among this population. Instead, focus should be on ensuring that at the time of death, the individual completing the death certificate accurately identifies the disease process or injury, other than developmental disability, that initiated the chain of events leading to death. 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 persons certifying the death certificate of decedents with a known development disability 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.

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. 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. 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] Further 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.

Acknowledgements

Author Contributions: Landes had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Concept and design: Landes, Stevens, Turk.

Acquisition of data: Landes

Analysis of data: Landes, Stevens.

Interpretation of data: Landes, Stevens, Turk.

Drafting of the manuscript: Landes, Stevens.

Critical revision of the manuscript for important intellectual content: Landes, Stevens, Turk.

Competing Interests: None.

Funding: No funding was received for this project.

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Table 1: UCOD (UCOD) trends for decedents with intellectual disability, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=7,901)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	15.37%	23.67%	8.30%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	17.18%	20.45%	3.27%
XX.	V00-Y99	External causes of mortality	9	↗	3	2.67%	7.53%	4.86%
VI.	G00-G99	Diseases of the nervous system	5	↗	4	6.81%	7.39%	0.58%
II.	C00-D49	Neoplasms	4	↘	5	7.00%	7.05%	0.05%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	6	→	6	5.94%	6.66%	0.72%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	1	↘	7	30.92%	5.72%	-25.20%
XI.	K00-K95	Diseases of the digestive system	7	↘	8	5.37%	5.71%	0.34%
I.	A00-B99	Certain infectious and parasitic diseases	8	↘	9	3.24%	3.82%	0.58%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	16	↗	10	0.00%	3.72%	3.72%
XIV.	N00-N99	Diseases of the genitourinary system	10	↘	11	2.53%	3.47%	0.94%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities	11	↘	12	1.35%	1.57%	0.22%
---	---	Unknown	16	↗	13	0.00%	1.24%	1.24%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.84%	0.92%	0.08%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	13	↘	15	0.32%	0.48%	0.16%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	16	0.32%	0.42%	0.10%
XVI.	P00-P96	Certain conditions originating in the perinatal period	14	↘	17	0.11%	0.11%	0.00%
VIII.	H60-H95	Diseases of the ear and mastoid process	15	↘	18	0.03%	0.04%	0.01%
VII.	H00-H59	Diseases of the eye and adnexa	15	↘	19	0.03%	0.03%	0.00%

*Indicates ICD-10 Chapter inclusive of intellectual disability.

Table 2: UCOD (UCOD) trends for decedents with cerebral palsy, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=11,895)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	8.95%	29.00%	20.05%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	9.42%	15.06%	5.64%
---	---	Unknown	17	↗	3	0.00%	11.71%	11.71%
VI.	G00-G99	Diseases of the nervous system*	1	↘	4	62.05%	6.53%	-55.52%
XX.	V00-Y99	External causes of mortality	9	↗	5	1.65%	5.89%	4.24%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	17	↗	6	0.00%	5.46%	5.46%
XI.	K00-K95	Diseases of the digestive system	5	↘	7	3.83%	5.15%	1.32%
II.	C00-D49	Neoplasms	4	↘	8	4.62%	4.72%	0.10%
I.	A00-B99	Certain infectious and parasitic diseases	6	↘	9	2.69%	3.99%	1.30%
XIV.	N00-N99	Diseases of the genitourinary system	11	↗	10	0.81%	3.40%	2.59%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	7	↘	11	2.02%	3.31%	1.29%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities	8	↘	12	1.65%	1.79%	0.14%
V.	F01-F99	Mental, behavioral and neurodevelopmental disorders	10	↘	13	1.08%	1.43%	0.35%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.55%	1.08%	0.53%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	15	→	15	0.14%	0.58%	0.44%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	14	↘	16	0.23%	0.43%	0.20%
XVI.	P00-P96	Certain conditions originating in the perinatal period	13	↘	16	0.31%	0.43%	0.12%
VIII.	H60-H95	Diseases of the ear and mastoid process	16	↘	17	0.01%	0.02%	0.01%
VII.	H00-H59	Diseases of the eye and adnexa	17	↘	18	0.00%	0.01%	0.01%

*Indicates ICD-10 Chapter inclusive of cerebral palsy.

Table 3: UCOD (UCOD) trends for decedents with Down syndrome, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=9,114)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	8.56%	27.00%	18.44%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	10.96%	17.38%	6.42%
VI.	G00-G99	Diseases of the nervous system	4	↗	3	5.31%	13.18%	7.87%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders	10	↗	4	1.73%	11.82%	10.09%
---	---	Unknown	16	↗	5	0.00%	5.65%	5.65%
XX.	V00-Y99	External causes of mortality	5	↘	6	2.79%	3.86%	1.07%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	7	→	7	2.15%	3.84%	1.69%
XVIII.	R00-R99	Symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	8	0.01%	3.23%	3.22%
I.	A00-B99	Certain infectious and parasitic diseases	6	↘	9	2.32%	3.12%	0.80%
XIV.	N00-N99	Diseases of the genitourinary system	11	↗	10	1.29%	2.56%	1.27%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	1	↘	11	60.27%	2.51%	-57.76%
XI.	K00-K95	Diseases of the digestive system	9	↘	12	1.78%	2.24%	0.46%
II.	C00-D49	Neoplasms	8	↘	13	2.10%	2.12%	0.02%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.35%	0.52%	0.17%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	15	0.26%	0.49%	0.23%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	14	↘	16	0.11%	0.46%	0.35%
XVI.	P00-P96	Certain conditions originating in the perinatal period	16	↘	17	0.00%	0.02%	0.02%
VIII.	H60-H95	Diseases of the ear and mastoid process	15	↘	18	0.01%	0.01%	0.00%

*Indicates ICD-10 Chapter inclusive of Down syndrome.

Table 4: UCOD (UCOD) trends for decedents with other developmental disabilities, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=2,479)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
IX.	I00-I99	Diseases of the circulatory system	2	↗	1	15.97%	19.48%	3.51%
X.	J00-J99	Diseases of the respiratory system	5	↗	2	8.67%	16.30%	7.63%
II.	C00-D49	Neoplasms	4	↗	3	11.29%	11.42%	0.13%
XX.	V00-Y99	External causes of mortality	7	↗	4	7.34%	8.75%	1.41%
VI.	G00-G99	Diseases of the nervous system	6	↗	5	7.42%	8.39%	0.97%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	8	↗	6	5.77%	6.53%	0.76%
XI.	K00-K95	Diseases of the digestive system	9	↗	7	5.16%	5.81%	0.65%
---	---	Unknown	17	↗	8	0.00%	4.88%	4.88%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	1	↘	9	17.39%	3.87%	-13.52%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	10	0.12%	3.63%	3.51%
I.	A00-B99	Certain infectious and parasitic diseases	10	↘	11	2.70%	3.03%	0.33%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	3	↘	12	14.44%	2.99%	-11.45%
XIV.	N00-N99	Diseases of the genitourinary system	11	↘	13	1.69%	2.38%	0.69%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.93%	1.17%	0.24%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	15	0.69%	0.85%	0.16%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	14	↘	16	0.24%	0.36%	0.12%
XVI.	P00-P96	Certain conditions originating in the perinatal period	16	↘	17	0.08%	0.08%	0.00%
XV.	O00-O9A	Pregnancy, childbirth and the puerperium	16	↘	17	0.08%	0.08%	0.00%

*Indicates ICD-10 Chapter inclusive of other developmental disabilities.

Table 5: UCOD (UCOD) trends for decedents with multiple developmental disabilities, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=1,765)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	12.01%	29.24%	17.23%
IX.	I00-I99	Diseases of the circulatory system	5	↗	2	9.18%	16.03%	6.85%
VI.	G00-G99	Diseases of the nervous system*	1	↘	3	32.24%	7.42%	-24.82%
XI.	K00-K95	Diseases of the digestive system	6	↗	4	5.89%	6.80%	0.91%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	5	0.00%	6.35%	6.35%
XX.	V00-Y99	External causes of mortality	10	↗	6	1.76%	6.06%	4.30%
---	---	Unknown	15	↗	7	0.00%	5.16%	5.16%
I.	A00-B99	Certain infectious and parasitic diseases	8	→	8	3.34%	4.42%	1.08%
II.	C00-D49	Neoplasms	7	↘	9	4.25%	4.31%	0.06%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	9	↘	10	1.98%	3.63%	1.65%
XIV.	N00-N99	Diseases of the genitourinary system	11	→	11	1.19%	3.06%	1.87%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	4	↘	12	11.22%	2.89%	-8.33%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	2	↘	13	15.81%	2.49%	-13.32%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.74%	1.25%	0.51%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	15	→	15	0.00%	0.45%	0.45%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	16	0.28%	0.34%	0.06%
XVI.	P00-P96	Certain conditions originating in the perinatal period	14	↘	17	0.11%	0.11%	0.00%

*Indicates ICD-10 Chapter inclusive of multiple developmental disabilities.

STROBE 2007 (v4) Statement—Checklist of items that should be included in reports of *cross-sectional studies*

Section/Topic	Item #	Recommendation	Reported on page #
Title and abstract	1	(*a*) Indicate the study's design with a commonly used term in the title or the abstract	1
		(*b*) Provide in the abstract an informative and balanced summary of what was done and what was found	2
Introduction			
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported	4-6
Objectives	3	State specific objectives, including any prespecified hypotheses	6
Methods			
Study design	4	Present key elements of study design early in the paper	6-7
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection	6
Participants	6	(*a*) Give the eligibility criteria, and the sources and methods of selection of participants	6
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable	6-7
Data sources/ measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group	6
Bias	9	Describe any efforts to address potential sources of bias	*6*
Study size	10	Explain how the study size was arrived at	6
Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why	6
Statistical methods	12	(*a*) Describe all statistical methods, including those used to control for confounding	6-7
		(*b*) Describe any methods used to examine subgroups and interactions	6-7
		(*c*) Explain how missing data were addressed	NA
		(*d*) If applicable, describe analytical methods taking account of sampling strategy	NA
		(*e*) Describe any sensitivity analyses	NA
Results			

Participants	13*	(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed	7-8
		(b) Give reasons for non-participation at each stage	NA
		(c) Consider use of a flow diagram	NA
Descriptive data	14*	(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders	7-8
		(b) Indicate number of participants with missing data for each variable of interest	NA
Outcome data	15*	Report numbers of outcome events or summary measures	7-8
Main results	16	(*a*) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included	7-10
		(*b*) Report category boundaries when continuous variables were categorized	NA
		(*c*) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period	NA
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses	NA
Discussion			
Key results	18	Summarise key results with reference to study objectives	10
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias	11-12
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence	11-12
Generalisability	21	Discuss the generalisability (external validity) of the study results	13
Other information			
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based	14

*Give information separately for cases and controls in case-control studies and, if applicable, for exposed and unexposed groups in cohort and cross-sectional studies.

Note: An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at http://www.plosmedicine.org/, Annals of Internal Medicine at http://www.annals.org/, and Epidemiology at http://www.epidem.com/). Information on the STROBE Initiative is available at www.strobe-statement.org.

BMJ Open

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

Journal:	*BMJ Open*
Manuscript ID	bmjopen-2018-026614.R1
Article Type:	Research
Date Submitted by the Author:	10-Dec-2018
Complete List of Authors:	Landes, Scott; Syracuse University, Department of Sociology and Aging Studies Institute Stevens, James; Syracuse University, Department of Sociology and Aging Studies Institute Turk, Margaret; SUNY Upstate Medical University, Department of Physical Medicine and Rehabilitation
Primary Subject Heading:	Public health
Secondary Subject Heading:	Epidemiology
Keywords:	Developmental disability, Underlying cause of death, Mortality trends

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

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Word count: 3,624

Abstract

Objective: To determine whether coding a developmental disability as the underlying cause of death obscures mortality trends of adults with developmental disability.

Design: National Vital Statistics System 2012-2016 U.S. Multiple Cause-of-Death Mortality files.

Setting: United States.

Participants: Adults with a developmental disability indicated on their death certificate aged 18 through 103 at the time of death. The study population included 33,154 adults who died between January 1, 2012 and December 31, 2016.

Primary outcome and measures: Decedents with a developmental disability coded as the underlying cause of death on the death certificate were identified utilizing an ICD-10 code for intellectual disability, cerebral palsy, Down syndrome, or other developmental disability. Death certificates that coded a developmental disability as the underlying cause of death were revised utilizing a sequential underlying cause of death revision process.

Results: There were 33,154 decedents with developmental disability: 7,901 with intellectual disability, 11,895 with cerebral palsy, 9,114 with Down syndrome, 2,479 with other developmental disabilities, and 1,765 with multiple developmental disabilities. Among all decedents, 48.5% had a developmental disability coded as the underlying cause of death, obscuring higher rates of choking deaths among all decedents, and dementia and Alzheimer disease among decedents with Down syndrome.

Conclusion: Death certificates that recorded the developmental disability in Part I of the death certificate were more likely to code disability as the underlying cause of death. While revising these death certificates provides a short-term corrective to mortality trends for this population, the severity and extent of this problem warrants a long-term change involving more precise instructions to record developmental disabilities only in Part II of the death certificate.

Strengths and limitations of this study

- This study revises death certificates that had developmental disability coded as underlying cause of death.
- This study provides evidence that coding developmental disability as underlying cause of death obscures often preventable, diseases or injuries other than disability that initiated the sequence of events leading to death.
- The high percentage of death certificates coded with a developmental disability as the underlying cause of death reveals the need to revise death certificates for adults with developmental disability prior to reporting mortality trends.
- Since the study population only includes decedents who had a developmental disability recorded on their death certificate, it is not inclusive of all decedents with a developmental disability in the U.S

- Limitations to retrospective revising of death certificates highlight the need for a long-term solution involving formal changes to policies guiding coding of developmental disability on death certificates.

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

Public health and preventive care efforts aimed at reducing premature mortality rely on population mortality data from death certificates.[1,2] Thus, it is imperative that death certificates accurately identify a valid and informative underlying cause of death (UCOD), the disease or injury that initiated the causal sequence of events leading to death. Beyond general concerns with the overall accuracy of death certificate data,[3-5] researchers express specific concern regarding the frequency in which the UCOD identifies a cause that is not valid or informative for public health and preventive care efforts aimed at reducing premature mortality.[2,6,7] One such cause of concern occurs when decedents with developmental disability have their disability coded as their UCOD.[8-14]

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]

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

this improvement, on average, research informs that adults with developmental disability die 20-25 years earlier than those in the general population.[18,19,21] There is increasing consensus that developmental disabilities *should not* be considered a valid UCOD, as doing so prohibits identification of the preventable medical cause of death, and is not advantageous to public health or preventive care efforts.[2,8-15,23] Instead, the goal in coding the UCOD on death certificates for persons with developmental disability should be to identify the specific, and often preventable, disease or injury other than disability that initiated the sequence of events leading to death.

Cerebral palsy offers insight to the limitations of coding a developmental disability as an UCOD. Cerebral palsy describes a group of various permanent non-progressive motor disorders which stem from an injury to or malfunction in the developing brain and vary in severity, cause, and presentation.[24] Besides the motor disorders that cause activity limitations, there can be associated conditions that accompany the diagnosis of cerebral palsy such as epilepsy or problems with sensation, cognition, communication, and behavior. People with cerebral palsy also have risk factors for general health conditions shared with the general population, such as chronic heart disease.[25] Due to a higher prevalence of gastroesophageal reflux and/or dysphagia among adults with moderate to severe forms of cerebral palsy, there is increased risk among this population for aspiration on either saliva, food, or gastric contents leading to pneumonia or pneumonitis, and subsequent death.[26,27] In the event that an individual with cerebral palsy aspirates on food, develops pneumonia, and dies, it is not useful to identify the UCOD as cerebral palsy. Doing so conceals the fact that death was due to aspiration and subsequent development of pneumonia, and fails to differentiate this causal pathway from other causal pathways such as arteriosclerotic heart disease leading to acute myocardial infarction and death. As this example illustrates, coding a developmental disability as the UCOD is

uninformative,[2,28,29] and, as Trollor and colleagues[12] contend, "obscures" actual mortality trends related to preventable conditions that need increased attention in public health or preventive care efforts.

Prior studies report the prevalence and obscuring effect of coding a developmental disability as the UCOD. Internationally, 16% of persons with developmental disability in general,[12] and between 34% and 56% of persons with cerebral palsy in particular,[8,30] had their disability coded as their UCOD. Among the U.S. population, 20% of decedents with an intellectual disability,[10] and 21% of decedents with Down syndrome,[31] had their disability coded as their UCOD. Two studies, from Canada[14] and Australia,[12] that mention the obscuring effect of this practice on mortality trends report a substantial increase in deaths from respiratory disease after revising death certificates coded with a developmental disability as the UCOD. To date, researchers have not detailed the obscuring effect of coding a developmental disability as the UCOD on mortality trends for adults with developmental disability in the U.S. To determine the severity of this problem, we compare mortality trends for adult decedents with developmental disability using the originally reported and a revised UCOD from U.S. death certificate data.

Methods

Data

Death certificate data for this study are from the National Vital Statistics System 2012-2016 U.S. Multiple Cause-of-Death Mortality files. As mortality coding in the U.S. converted to ICD-10 in 2009,[32] this study included the death certificates for adults aged 18-103 at the time of death that recorded an ICD-10 code for a developmental disability as the UCOD, or as a multiple cause of death (other co-morbidities present at time of death) - intellectual disability (F70-79),

cerebral palsy (G80), Down syndrome (Q90), and other developmental disabilities (F80-89, Q91-99). This resulted in a sample of 33,154 death certificates over the 5-year period.

Sequential UCOD Revision Process

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.'*Analytic Plan*

All analysis was conducted using STATA Version 15.0 (College Station, TX). Results are presented by developmental disability group due to disparate cause of death trends – intellectual disability, cerebral palsy, Down syndrome, other developmental disability, and multiple developmental disabilities. For each group, we report: UCOD percentages by ICD-10 chapter code for all death certificates prior to and after revisions; changes in the cause of death rank order; and percentage point differences between original and revised data. We also detail the leading specific underlying causes of death within ICD-10 chapters reflecting the most drastic changes.

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.

Results

Full comparative results of the original and revised UCOD by ICD-10 Chapter code are presented for each disability group in Tables 1-5. Though detailed in the text, information on specific underlying causes of death per ICD-10 Chapter are not included in the tables. We compared results from aggregated data to results from each specific year prior and subsequent to revisions. The only dissimilarity was a slight increase in the percentage of decedents with Down syndrome who had Q90 coded as their UCOD beginning in 2014.

There were 7,901 decedents with intellectual disability, 11,895 decedents with cerebral palsy, 9,114 decedents with Down syndrome, 2,479 decedents with other developmental disabilities, and 1,765 decedents with multiple developmental disabilities. The UCOD was coded as a developmental disability, and thus was revised, on the death certificates of 48.52% of all decedents with a developmental disability, but varied by disability type - 25.88% of decedents with intellectual disability, 59.53% of decedents with cerebral palsy, 58.99% of decedents with Down syndrome, 26.42% of decedents with other developmental disabilities, and 52.69% of decedents with multiple developmental disabilities.

As expected, after revising the death certificates, there was a steep decline in the percentage and rank order of decedents in each disability group with an UCOD in the ICD-10 chapter inclusive of their disability. Due to these revisions, the percentages increased for all other ICD-10 chapters for all disability groups. We detail the most remarkable changes.

Across groups, the percentage of deaths caused by diseases of the respiratory system, ICD-10 Chapter X, rose dramatically after the revision: from 15.37% to 23.67% for decedents with intellectual disability; from 8.95% to 29% for decedents with cerebral palsy; from 8.56% to 27% for decedents with Down syndrome; from 8.67% to 16.3% for decedents with other developmental disabilities; and from 12.01% to 29.24% for decedents with multiple developmental disabilities. As a result, respiratory deaths rose from the 3rd to the 1st leading cause of death for decedents with intellectual disability, cerebral palsy, Down syndrome, and multiple developmental disabilities; and from the 5th to the 2nd leading cause of death for decedents with other developmental disabilities such as autism.

For all disability groups, the most prevalent underlying causes of death identified among diseases of the respiratory system were J69 Pneumonitis due to inhalation of food/vomit and J18.9 Pneumonia, unspecified organism. The percentage of deaths from pneumonitis due to inhalation of food/vomit increased from: 5.73% to 9.57% among decedents with intellectual disability; 3.14% to 10.08% among decedents with cerebral palsy; 5.43% to 9.84% among decedents with Down syndrome; 3.03% to 5.77% among decedents with other developmental disabilities; and 5.72% to 12.18% among decedents with multiple developmental disabilities. The percentage of deaths from pneumonia, unspecified organism rose from: 4.38% to 7.1% among decedents with intellectual disability; 2.3% to 8.53% among decedents with cerebral palsy; 0.07% to 10.19% among decedents with Down syndrome; 2.02% to 4.4% among decedents with other developmental disabilities; and 2.21% to 8.67% among decedents with multiple developmental disabilities.

To varying degrees, deaths from external causes of mortality, ICD-10 Chapter XX, increased in percent and rank order for four of the disability groups: from 2.67% to 7.53% (rank

order increase from 9 to 3) for decedents with intellectual disability; from 1.65% to 5.89% (rank order increase from 9 to 5) for decedents with cerebral palsy; from 7.34% to 8.75% (rank order increase from 7 to 4) for decedents with other developmental disabilities; from 1.76% to 6.06% (rank order increase from 10 to 6) for decedents with multiple developmental disabilities. While the percentage of deaths from external causes of mortality rose from 2.79% to 3.86% for decedents with Down syndrome, the rank order dropped one position from 5 to 6.

For each disability group, the most prevalent underlying causes of death in the external causes chapter were W78-80, Accidental inhalation and ingestion of food or other objects causing obstruction of the respiratory tract. The percentage of deaths from accidental choking increased: from 0.1% to 4.80% among decedents with intellectual disability; from 0.02% to 3.76% among decedents with cerebral palsy; from 1.32% to 2.28% among decedents with Down syndrome; from 3.19% to 4.48% among decedents with other developmental disabilities; and from 0.17% to 4.09% among decedents with multiple developmental disabilities.

One other remarkable finding among all disability groups regarded the percentage of decedents who had either an unknown or an imprecise UCOD. Unknown causes of death were recorded when no other ICD-10 code was listed in Part I of the death certificate other than disability. Imprecise underlying causes occurred when no other ICD-10 code was listed in Part I of the death certificate other than the developmental disability and/or a single or multiple R code from Chapter XVIII. As a result, the UCOD was either unknown or imprecise for: 4.96% of decedents with intellectual disability; 17.17% of decedents with cerebral palsy; 8.88% of decedents with Down syndrome; 8.51% of decedents with other developmental disabilities; and 11.51% of decedents with multiple developmental disabilities. This means that even after

revision, the death certificate provided little to no indication of the disease or injury that initiated the sequence of events leading to the death of these individuals.

Particular to decedents with Down syndrome, deaths from diseases of the nervous system, ICD-10 Chapter VI, rose from 5.31% to 13.18% (rank order increase from 4 to 3). The most prevalent UCOD for decedents with Down syndrome in this chapter, G30.9 Alzheimer disease, unspecified, increased from 3.27% to 9.5%. In addition, contrary to trends among the other disability groups, deaths from mental, behavioral, and neurodevelopmental disorders, ICD-10 Chapter V, rose from 1.73% to 11.82% (rank order increase from 10 to 4). The most prevalent cause of death in this chapter, F03 Unspecified dementia, increased from 1.62% to 11.4%.

Discussion

In 2012-2016 U.S. death certificate data, the coding of a developmental disability as the UCOD is widespread, but varied among specific disability groups. Results from this study expose that the practice of coding a developmental disability as the UCOD on a death certificate obscures the higher prevalence of choking related deaths (coded as either pneumonitis due to inhalation of food/vomit, or accidental choking) among all developmental disability groups, and obscures dementia and Alzheimer disease related deaths among decedents with Down syndrome.

In instances when death certificates that code a developmental disability as the UCOD are not revised, population mortality trends for persons with various types of developmental disability are likely to be inaccurate and may, therefore, misinform public health and preventive care efforts aimed at preventing premature mortality. The primary strength of this study is that results demonstrate that the prominence of choking related deaths as a mortality risk among persons with developmental disability was severely underestimated prior to revision. As revision of the UCOD revealed, similar to the older adult population,[36-38] careful attention is needed to

attend to the heightened risk of preventable choking related deaths among persons with developmental disability.[15,39,40] Thus, it is of the utmost importance that public health and preventive care efforts focus on management of the swallowing function among this population. Physicians conducting routine exams should be screening for swallowing disorders, abnormalities in the cough and gag reflex, as well as respiratory distress.[15,40] Mortality data based upon original death certificates, often coding developmental disability as UCOD, fail to reveal this important mortality trend.

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. Continued permission of the practice of coding a developmental disability as the UCOD demands both short-term and long-term redress. As this study highlights, analysis of mortality trends for adults with developmental disability must recognize the obscuring effect of coding a developmental disability as the UCOD and take the necessary steps to revise the UCOD on these death certificates prior to reporting findings. The sequential UCOD revision process utilized in this study reduces obfuscation of mortality trends among populations with developmental disabilities in the short-term.

While revising death certificates for adults with developmental disability is a necessary short-term solution, the primary limitations of this study indicate that retrospective revision is not a sufficient long-term solution. The first limitation regards accuracy of revised death certificates. Although sequentially revising the UCOD for U.S. death certificates that identify a

developmental disability as the UCOD increases 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. Although the only remedy present at the time, the method of retrospectively revising the UCOD should not be viewed as a sustainable long-term strategy for surveilling mortality trends among this population. 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 either 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.

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.

Acknowledgements

Author Contributions: Landes had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Concept and design: Landes, Stevens, Turk.

Acquisition of data: Landes

Analysis of data: Landes, Stevens.

Interpretation of data: Landes, Stevens, Turk.

Drafting of the manuscript: Landes, Stevens.

Critical revision of the manuscript for important intellectual content: Landes, Stevens, Turk.

Competing Interests: None.

Funding sources: No funding was received for this project.

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

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Table 1: UCOD (UCOD) trends for decedents with intellectual disability, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=7,901)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	15.37%	23.67%	8.30%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	17.18%	20.45%	3.27%
XX.	V00-Y99	External causes of mortality	9	↗	3	2.67%	7.53%	4.86%
VI.	G00-G99	Diseases of the nervous system	5	↗	4	6.81%	7.39%	0.58%
II.	C00-D49	Neoplasms	4	↘	5	7.00%	7.05%	0.05%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	6	→	6	5.94%	6.66%	0.72%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	1	↘	7	30.92%	5.72%	-25.20%
XI.	K00-K95	Diseases of the digestive system	7	↘	8	5.37%	5.71%	0.34%
I.	A00-B99	Certain infectious and parasitic diseases	8	↘	9	3.24%	3.82%	0.58%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	16	↗	10	0.00%	3.72%	3.72%
XIV.	N00-N99	Diseases of the genitourinary system	10	↘	11	2.53%	3.47%	0.94%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities	11	↘	12	1.35%	1.57%	0.22%
---	---	Unknown	16	↗	13	0.00%	1.24%	1.24%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.84%	0.92%	0.08%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	13	↘	15	0.32%	0.48%	0.16%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	16	0.32%	0.42%	0.10%
XVI.	P00-P96	Certain conditions originating in the perinatal period	14	↘	17	0.11%	0.11%	0.00%
VIII.	H60-H95	Diseases of the ear and mastoid process	15	↘	18	0.03%	0.04%	0.01%
VII.	H00-H59	Diseases of the eye and adnexa	15	↘	19	0.03%	0.03%	0.00%

*Indicates ICD-10 Chapter inclusive of intellectual disability.

Table 2: UCOD (UCOD) trends for decedents with cerebral palsy, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=11,895)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	8.95%	29.00%	20.05%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	9.42%	15.06%	5.64%
---	---	Unknown	17	↗	3	0.00%	11.71%	11.71%
VI.	G00-G99	Diseases of the nervous system*	1	↘	4	62.05%	6.53%	-55.52%
XX.	V00-Y99	External causes of mortality	9	↗	5	1.65%	5.89%	4.24%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	17	↗	6	0.00%	5.46%	5.46%
XI.	K00-K95	Diseases of the digestive system	5	↘	7	3.83%	5.15%	1.32%
II.	C00-D49	Neoplasms	4	↘	8	4.62%	4.72%	0.10%
I.	A00-B99	Certain infectious and parasitic diseases	6	↘	9	2.69%	3.99%	1.30%
XIV.	N00-N99	Diseases of the genitourinary system	11	↗	10	0.81%	3.40%	2.59%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	7	↘	11	2.02%	3.31%	1.29%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities	8	↘	12	1.65%	1.79%	0.14%
V.	F01-F99	Mental, behavioral and neurodevelopmental disorders	10	↘	13	1.08%	1.43%	0.35%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.55%	1.08%	0.53%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	15	→	15	0.14%	0.58%	0.44%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	14	↘	16	0.23%	0.43%	0.20%
XVI.	P00-P96	Certain conditions originating in the perinatal period	13	↘	16	0.31%	0.43%	0.12%
VIII.	H60-H95	Diseases of the ear and mastoid process	16	↘	17	0.01%	0.02%	0.01%
VII.	H00-H59	Diseases of the eye and adnexa	17	↘	18	0.00%	0.01%	0.01%

*Indicates ICD-10 Chapter inclusive of cerebral palsy.

Table 3: UCOD (UCOD) trends for decedents with Down syndrome, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=9,114)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	8.56%	27.00%	18.44%
IX.	I00-I99	Diseases of the circulatory system	2	→	2	10.96%	17.38%	6.42%
VI.	G00-G99	Diseases of the nervous system	4	↗	3	5.31%	13.18%	7.87%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders	10	↗	4	1.73%	11.82%	10.09%
---	---	Unknown	16	↗	5	0.00%	5.65%	5.65%
XX.	V00-Y99	External causes of mortality	5	↘	6	2.79%	3.86%	1.07%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	7	→	7	2.15%	3.84%	1.69%
XVIII.	R00-R99	Symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	8	0.01%	3.23%	3.22%
I.	A00-B99	Certain infectious and parasitic diseases	6	↘	9	2.32%	3.12%	0.80%
XIV.	N00-N99	Diseases of the genitourinary system	11	↗	10	1.29%	2.56%	1.27%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	1	↘	11	60.27%	2.51%	-57.76%
XI.	K00-K95	Diseases of the digestive system	9	↘	12	1.78%	2.24%	0.46%
II.	C00-D49	Neoplasms	8	↘	13	2.10%	2.12%	0.02%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.35%	0.52%	0.17%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	15	0.26%	0.49%	0.23%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	14	↘	16	0.11%	0.46%	0.35%
XVI.	P00-P96	Certain conditions originating in the perinatal period	16	↘	17	0.00%	0.02%	0.02%
VIII.	H60-H95	Diseases of the ear and mastoid process	15	↘	18	0.01%	0.01%	0.00%

*Indicates ICD-10 Chapter inclusive of Down syndrome.

Table 4: UCOD (UCOD) trends for decedents with other developmental disabilities, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=2,479)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
IX.	I00-I99	Diseases of the circulatory system	2	↗	1	15.97%	19.48%	3.51%
X.	J00-J99	Diseases of the respiratory system	5	↗	2	8.67%	16.30%	7.63%
II.	C00-D49	Neoplasms	4	↗	3	11.29%	11.42%	0.13%
XX.	V00-Y99	External causes of mortality	7	↗	4	7.34%	8.75%	1.41%
VI.	G00-G99	Diseases of the nervous system	6	↗	5	7.42%	8.39%	0.97%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	8	↗	6	5.77%	6.53%	0.76%
XI.	K00-K95	Diseases of the digestive system	9	↗	7	5.16%	5.81%	0.65%
---	---	Unknown	17	↗	8	0.00%	4.88%	4.88%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	1	↘	9	17.39%	3.87%	-13.52%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	10	0.12%	3.63%	3.51%
I.	A00-B99	Certain infectious and parasitic diseases	10	↘	11	2.70%	3.03%	0.33%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	3	↘	12	14.44%	2.99%	-11.45%
XIV.	N00-N99	Diseases of the genitourinary system	11	↘	13	1.69%	2.38%	0.69%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.93%	1.17%	0.24%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	15	0.69%	0.85%	0.16%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	14	↘	16	0.24%	0.36%	0.12%
XVI.	P00-P96	Certain conditions originating in the perinatal period	16	↘	17	0.08%	0.08%	0.00%
XV.	O00-O9A	Pregnancy, childbirth and the puerperium	16	↘	17	0.08%	0.08%	0.00%

*Indicates ICD-10 Chapter inclusive of other developmental disabilities.

Table 5: UCOD (UCOD) trends for decedents with multiple developmental disabilities, 2012-2016 U.S. Multiple Cause-of-Death Mortality files (*N*=1,765)

ICD-10 Chapter			Rank order change			Original UCOD	Revised UCOD	Absolute Difference
X.	J00-J99	Diseases of the respiratory system	3	↗	1	12.01%	29.24%	17.23%
IX.	I00-I99	Diseases of the circulatory system	5	↗	2	9.18%	16.03%	6.85%
VI.	G00-G99	Diseases of the nervous system*	1	↘	3	32.24%	7.42%	-24.82%
XI.	K00-K95	Diseases of the digestive system	6	↗	4	5.89%	6.80%	0.91%
XVIII.	R00-R99	Symptoms, signs, and abnormal clinical and laboratory findings, not elsewhere classified	15	↗	5	0.00%	6.35%	6.35%
XX.	V00-Y99	External causes of mortality	10	↗	6	1.76%	6.06%	4.30%
---	---	Unknown	15	↗	7	0.00%	5.16%	5.16%
I.	A00-B99	Certain infectious and parasitic diseases	8	→	8	3.34%	4.42%	1.08%
II.	C00-D49	Neoplasms	7	↘	9	4.25%	4.31%	0.06%
IV.	E00-E89	Endocrine, nutritional and metabolic diseases	9	↘	10	1.98%	3.63%	1.65%
XIV.	N00-N99	Diseases of the genitourinary system	11	→	11	1.19%	3.06%	1.87%
V.	F01-F99	Mental, behavioral, and neurodevelopmental disorders*	4	↘	12	11.22%	2.89%	-8.33%
XVII.	Q00-Q99	Congenital malformations, deformations, and chromosomal abnormalities*	2	↘	13	15.81%	2.49%	-13.32%
XIII.	M00-M99	Diseases of the musculoskeletal system and connective tissue	12	↘	14	0.74%	1.25%	0.51%
XII.	L00-L99	Diseases of the skin and subcutaneous tissue	15	→	15	0.00%	0.45%	0.45%
III.	D50-D89	Diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	13	↘	16	0.28%	0.34%	0.06%
XVI.	P00-P96	Certain conditions originating in the perinatal period	14	↘	17	0.11%	0.11%	0.00%

*Indicates ICD-10 Chapter inclusive of multiple developmental disabilities.

STROBE 2007 (v4) Statement—Checklist of items that should be included in reports of *cross-sectional studies*

Section/Topic	Item #	Recommendation	Reported on page #
Title and abstract	1	(*a*) Indicate the study's design with a commonly used term in the title or the abstract	1
		(*b*) Provide in the abstract an informative and balanced summary of what was done and what was found	2
Introduction			
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported	4-6
Objectives	3	State specific objectives, including any prespecified hypotheses	6
Methods			
Study design	4	Present key elements of study design early in the paper	6-7
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection	6
Participants	6	(*a*) Give the eligibility criteria, and the sources and methods of selection of participants	6
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable	6-7
Data sources/ measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group	6
Bias	9	Describe any efforts to address potential sources of bias	*6*
Study size	10	Explain how the study size was arrived at	6
Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why	6
Statistical methods	12	(*a*) Describe all statistical methods, including those used to control for confounding	6-7
		(*b*) Describe any methods used to examine subgroups and interactions	6-7
		(*c*) Explain how missing data were addressed	NA
		(*d*) If applicable, describe analytical methods taking account of sampling strategy	NA
		(*e*) Describe any sensitivity analyses	NA
Results			

Participants	13*	(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed	7-8
		(b) Give reasons for non-participation at each stage	NA
		(c) Consider use of a flow diagram	NA
Descriptive data	14*	(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders	7-8
		(b) Indicate number of participants with missing data for each variable of interest	NA
Outcome data	15*	Report numbers of outcome events or summary measures	7-8
Main results	16	(*a*) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included	7-10
		(*b*) Report category boundaries when continuous variables were categorized	NA
		(*c*) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period	NA
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses	NA
Discussion			
Key results	18	Summarise key results with reference to study objectives	10
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias	11-12
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence	11-12
Generalisability	21	Discuss the generalisability (external validity) of the study results	13
Other information			
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based	14

*Give information separately for cases and controls in case-control studies and, if applicable, for exposed and unexposed groups in cohort and cross-sectional studies.

Note: An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at http://www.plosmedicine.org/, Annals of Internal Medicine at http://www.annals.org/, and Epidemiology at http://www.epidem.com/). Information on the STROBE Initiative is available at www.strobe-statement.org.