



RESEARCH BRIEF #87

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CDC Guidelines Hide the Alzheimer Disease Mortality Burden among Adults with Down Syndrome

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KEY FINDINGS

- Current CDC guidelines on completing death certificates hide the actual Alzheimer disease mortality burden among adults with Down Syndrome.
- While death certificates show that adults with versus without Down Syndrome are equally likely to die to Alzheimer disease, this is an administrative mirage.
- The actual mortality burden from Alzheimer disease and unspecified dementia is 2.7x higher for adults with than without Down syndrome between 2005 and 2019.

Alzheimer disease is much more common among adults with than without Down syndrome. While an estimated 11.3% of adults in the general population over the age of 65 have Alzheimer disease,¹ nearly all adults with Down syndrome have Alzheimer neuropathology (e.g., lesions, neurofibrillary tangles, beta-amyloid plaques) by age 40,^{2,3} and half develop Alzheimer disease by age 60.⁴ Yet, this disparity appears to disappear when looking at Alzheimer disease mortality burden – the percentage of decedents who have Alzheimer disease reported as their cause of death. Is this a medical miracle? No, it is an administrative mirage. The reporting of Alzheimer disease mortality for people with Down syndrome is not accurate in the National Vital Statistics System. In 2014 the Centers for Disease Control and Prevention (CDC), in tandem with the WHO, changed the death certification guidelines for people with Down syndrome to require that Down syndrome be reported as the underlying cause of death in instances when the individual actually died from either Alzheimer disease or unspecified dementia. As a result, the percentage of death certificates for people with Down syndrome that report Alzheimer disease or unspecified dementia as the underlying cause of death dropped precipitously from 12.1% in 2013 to 1.6% in 2014, and to 0% in 2015-2019.

This brief summarizes the findings from our recently [published study](#), wherein we sought to address this administrative mirage by producing an accurate accounting of the disparity in Alzheimer disease mortality burden between adults with versus without Down syndrome for years 2005-2019. To do so, we first identified all death certificates that reported Down syndrome as the underlying cause of death, with Alzheimer disease or unspecified dementia as a contributing cause. We then amended the underlying cause of death for these cases using a sequential revision process, which in many cases (73.8%), was Alzheimer disease or unspecified dementia.

The Administrative Mirage

Figure 1 shows a) the percentage of death certificates that report Alzheimer disease or unspecified dementia as the underlying (main) cause of death for adults without Down syndrome; b) the percentage of death certificates that report Alzheimer disease or unspecified dementia as the underlying cause of death for adults with Down syndrome (actual death certificates); and c) the percentage of death certificates for adults with Down syndrome that should have reported Alzheimer disease or unspecified dementia instead of Down syndrome as the underlying cause of death (i.e., our corrected death certificates).

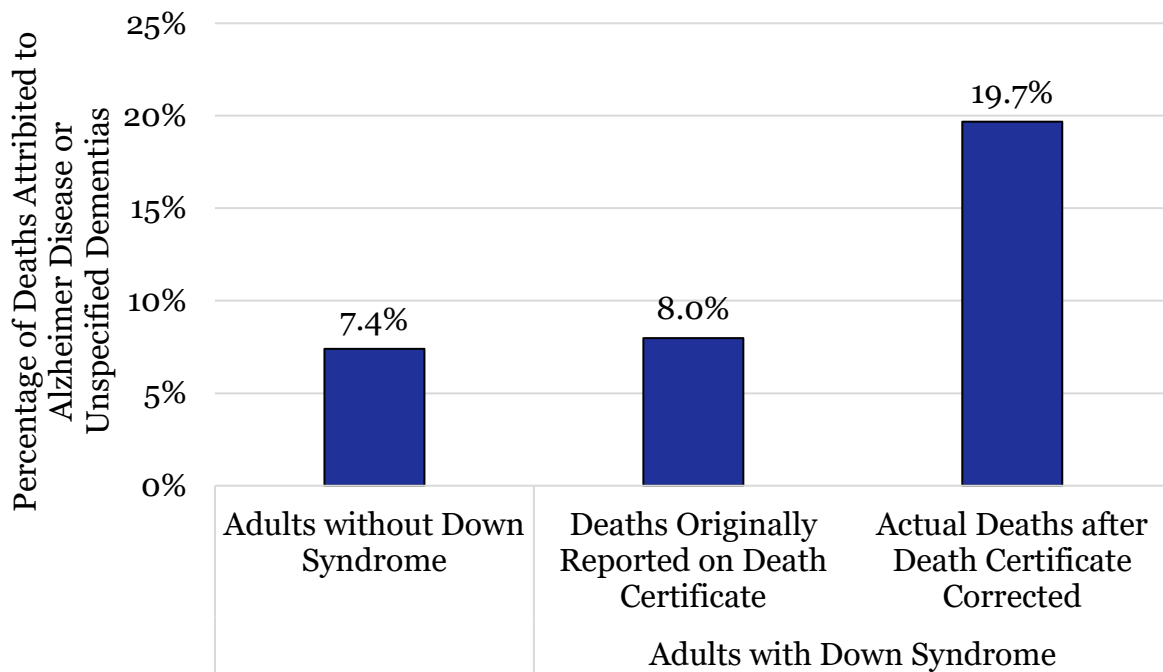


Figure 1. Percentage of all Deaths due to Alzheimer Disease or Unspecified Dementia among Adult Decedents ages 18+ with versus without Down Syndrome

Data Source: 2005-2019 U.S. Death Certificates, National Vital Statistics System

As shown in Figure 1, it appears that the mortality burden from Alzheimer disease and unspecified dementia in 2005-19 was similar for adults with (8.0%) and without (7.4%) Down syndrome. But this does not reflect the true burden of these diseases among adults with Down syndrome. After amending the death certificates that reported Down syndrome as the underlying cause of death, we see that in actuality, nearly 20% of adults with Down syndrome

who died 2005-19, died from Alzheimer disease or unspecified dementia

The CDC Should Instruct Death Certifiers Not to Report Down Syndrome as the Underlying Cause of Death on Death Certificates

This difference in the originally reported and actual mortality burden from Alzheimer disease or unspecified dementia is both startling and troubling. It is true that adults with Down syndrome are much more likely to develop Alzheimer disease neuropathology and be diagnosed with this disease at younger ages than the general population. But this fact does not justify hiding the actual mortality burden of this disease among people with Down syndrome. Doing so obscures the true burden of this disease in this population, and thereby increases the likelihood that this population will continue to be marginalized in Alzheimer disease health care and research efforts. To address this inaccuracy, the CDC should reverse the coding rule change regarding Alzheimer disease and unspecified dementia deaths among people with Down syndrome enacted in 2014 and further specify that Down syndrome should not be reported as the underlying cause of death.

Data and Methods

We used 2005-2019 U.S. Death Certificates for the study. The study included deaths among adults ages 18+. Down syndrome was identified if ICD-10 code Q90.9 was reported as the underlying or contributing cause of death. The 'originally reported' category for Down syndrome is based on death certificates prior to amendment. The 'actual' category for Down syndrome is based on death certificates after amending those that inaccurately report Down syndrome as the underlying cause of death.

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