

Comparative Mortality of Persons with Intellectual Disability in California: An Update (2000–2010)

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This paper updates our 2003 study¹ on the effect of intellectual disability (ID) on mortality in persons with no significant physical disability. As previously, we used the California Department of Developmental Services database to compute mortality rates by age, sex, and severity of ID. There were 64,207 subjects age 5 and older, who contributed 386,000 person-years of follow-up and 1514 deaths during the 2000 to 2010 study period. The excess death rates increased with age, ranging from 0.1 to 6.8 per 1000 in mild/moderate ID, and 3.4 to 6.7 in severe/profound.

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Approximately 1% of the US population² meets the American Association on Intellectual and Developmental Disabilities (AAIDD) definition of intellectual disability (ID), which characterizes disability based on limitations in intellectual functioning and adaptive behavior before age 18.³ Mild, moderate, severe, and profound ID compose roughly 85%, 10%, 4%, and 2% of the population with ID, respectively.²

In many cases, the etiology of ID is a progressive condition (eg, Down syndrome⁴) that in itself is associated with reduced survival. In addition, persons with ID are much more likely than those in the general population to have significant motor dysfunction and other comorbidities (eg, epilep-

sy) that may result in shortened life expectancy. Our aim was to estimate the increase in mortality due to ID in persons without significant physical impairment, comorbidity, or underlying degenerative condition.

SUBJECTS STUDIED

The database of the California Department of Developmental Services is compiled from annual Client Development Evaluation Reports⁵ (CDERs) on over 350,000 persons with developmental disabilities, who have been served since 1980, including over 280,000 persons followed between 2000 and 2010. The reliability of CDER items has been assessed previously and judged satisfactory.^{6–9}

Table 1. Demographic Characteristics of 64,207 Persons with ID and No Physical Disability or Other Serious Condition in the 2000–2010 California Department of Developmental Services Registry

Category	Distribution
Sex	
Male	57%
Female	43%
Ethnicity	
White	43%
Hispanic	34%
Black	16%
Other	7%
Age at first evaluation (within study period)	
5–14	28%
15–29	36%
30–44	23%
45–59	1%
60+	2%
Intellectual disability level	
Mild	75%
Moderate	20%
Severe	4%
Profound	1%

We identified 174,191 persons over the age of 5 during the 2000–2010 study period with a specified level of ID. Mild ID corresponds to an IQ of 50 to 70, moderate to 35 to 50, severe to 20 to 35, and profound to less than 20.

We then excluded persons whose ID was caused by degenerative or genetic conditions such as Down syndrome, or major medical conditions such as cancer or cardiac disease. For this we used CDER items for etiology based on the International Classification of Disease, 9th Revision Clinical Modification (ICD-9).¹⁰ In the same way, we also excluded persons with autism or epilepsy, and those whose ID was related to motor vehicle accidents or near drowning.

Finally, we excluded persons with any significant physical disability, as it is well known that reduced mobility is strongly associated with increased mortality.^{11–18} All study participants met the following 6 criteria: (1) assumes and maintains sitting

position independently, (2) uses fingers of hand independently of each other, (3) fully extends arms, (4) stands well alone and balances well for at least 5 minutes, (5) walks well alone for at least 20 feet and balances well, and (6) moves up and down stairs without need for handrail. The resulting subset was 64,207 persons.

Demographic characteristics are summarized in Table 1. The majority of the subjects was under age 30 and had mild ID. Compared with the general population of persons with ID, our sample has a lower percentage of mild ID (75% cf. 85%). This may be because the California database includes only persons receiving services from the state (eg, board and care, physical therapy, medical care), and is, therefore, more heavily weighted to the more severely impaired. On the other hand, many of those excluded from our subset due to having impaired motor function or comorbidities were more likely to have a more severe form of ID. Because our analyses are stratified by the level of severity of ID, the fact that our subset may differ from national averages does not engender a bias in the severity-specific risks.

METHODS

The CDER database was matched to mortality data from the California Department of Health Services, Bureau of Vital Statistics.¹⁹ The exposure period for each person started with the first CDER evaluation during the study period that met the study inclusion criteria. It ended with the earliest of (a) the date when these criteria were no longer met, (b) the date of death, (c) the end of the study period, December 31, 2010, and (d) 3 years from the last CDER. This last condition was included to avoid the possible bias introduced by persons who could have moved from the state, or for some other reason left the system, and thus would have a hiatus in their evaluations.

Deaths were counted only if they occurred within the exposure period. The total expo-

Table 2. Comparative Mortality Rates for Persons with Mild or Moderate Mental Retardation (2000–2010)

Attained Age (years)	Exposure Patient-years E	Number of Deaths		Mortality Ratio 100d/d'	Mean Annual Mortality Rate per 1000		
		Observed d	Expected* d'		Observed q	Expected q'	Excess q-q'
5–19	113,586	59	46.8	126%	0.5	0.4	0.1
20–39	153,593	341	175.2	195%	2.2	1.1	1.1
40–59	86,666	620	352.3	176%	7.2	4.1	3.1
60+	12,170	322	239.1	135%	26.5	19.6	6.8
All	366,015	1342	813.5	165%	3.7	2.2	1.4

* Basis of expected deaths: 2005 abridged US Life Table rates for males and females.

sure time was allocated to the appropriate sex and age intervals. There were a total of 386,000 person-years of follow-up, and 1514 observed deaths.

As previously stated,¹ we combined the mild and moderate groups, and also the severe and profound groups. The 2005 US Abridged Life Table²⁰ provided the expected mortality rates (q') shown in Tables 2 and 3. The 2005 table was used because it represented the approximate midpoint of the exposure time in the 2000–2010 study period. The expected number of deaths was computed separately by sex as the product of the quinquennial age-specific mortality rate for the general population and the appropriate exposure time. Age- and sex-specific excess death rates (EDRs) and mortality ratios (MRs) were computed. The EDRs and MRs were fairly comparable for males and fe-

males; the results are, therefore, presented as combined.

To determine whether there was a secular trend toward improved survival over the larger 1980–2010 period, we used a multiplicative hazard model (logistic regression) applied to the person-years of exposure.^{21,22} The model contained terms for sex, age, level of ID, and calendar year.

RESULTS

As shown in Tables 2 and 3, the overall mortality in each group was higher than for the US general population. The overall MR for mild/moderate ID was 165%, and for severe/profound ID, it was 185%. The overall EDRs were 1.4 per 1000 and 3.9 per 1000, respectively. As has been found previously for other chronic disabilities,^{23,24} the MR tends to

Table 3. Comparative Mortality Rates for Persons with Severe or Profound Mental Retardation (2000–2010)

Attained Age (years)	Exposure Patient-years E	Number of Deaths		Mortality Ratio 100d/d'	Mean Annual Mortality Rate per 1000		
		Observed d	Expected* d'		Observed q	Expected q'	Excess q-q'
5–19	1818	7	0.8	830%	3.8	0.5	3.4
20–39	6960	33	8.6	383%	4.7	1.2	3.5
40–59	9447	79	42.5	186%	8.4	4.5	3.9
60+	1759	53	41.2	129%	30.1	23.4	6.7
All	19,984	172	93.1	185%	8.6	4.7	3.9

* Basis of expected deaths: 2005 abridged US Life Table rates for males and females.

decrease with age and the EDR to increase. A single MR or EDR should, therefore, not be used at all ages.

In mild/moderate ID, the EDRs ranged from 0.1 per 1000 at ages 5–19 to 6.8 per 1000 at ages 60+. In severe/profound, they ranged from 3.4 to 6.7, respectively. With one exception (age 60+), the MRs and EDRs were higher in severe/profound ID than in mild/moderate.

There was a 1.0% annual improvement in survival over the larger 1980–2010 period (95% confidence interval 0.6% to 1.5%). This is very similar to that in the underlying US general population. As a result, the EDRs would be expected to increase only slightly over the narrow 2000–2010 time period, and the MRs to remain roughly constant. Rather than compute the EDRs or MRs separately by calendar year, however, or use a model to estimate these figures for 2010, we present the average values over the 2000–2010 period, based on comparison with US mortality rates at the midpoint of the period, 2005.

A detailed study of the causes of excess mortality is beyond the scope of the present paper. We did find, however, that as in the general population, externally caused deaths (eg, accidents, homicide, drowning) were more common in males than in females. We also found that the proportion of externally caused deaths was larger in the mild/moderate group than in the severe/profound. This is consistent with previous findings.¹

DISCUSSION

The population considered here appears to be the largest group with ID to be studied with respect to long-term age- and severity-specific mortality. As shown in Tables 2 and 3, the overall mortality ratios are 165% for mild/moderate ID and 185% for severe/profound. The overall ratios may be misleading, however, as the demographics of the present study population do not necessarily

reflect that of the general population with ID. In addition, the MRs are known to vary with age, while the overall figure are (by construction) weighted according to the age distribution.

As noted, mortality rates in the population improved at 1% per year, in line with that of the general population. Similarly, Strauss et al²⁹ reported improvement of 0.9% per year for adults with mild or moderate cerebral palsy. A limitation of our finding is that it could in part reflect the change in criteria for diagnosing ID over the years. Prior to 1992, significant deficiencies in adaptive behavior were not evaluated in conjunction with intelligence testing, so a person diagnosed as having moderate ID in 1990, for example, might in fact have been functioning at what today would be considered to be the mild level. Also, not until 2002 was it clarified that the disability had to be established before the age of 18.³ Persons with acquired conditions, who were healthy at birth, may thus have been included in the prior ID population. Such criteria may have resulted in increased actual severity of disability within each nominal category of ID over these years, which could mask any secular improvement.

The mortality ratios found here are considerably lower than those published elsewhere.^{25–27} For example, Brackenridge & Elder²⁵ give rating guidelines equivalent to mortality ratios of 200% to 250% for persons with mild or moderate ID. More recently, Tyrer et al²⁶ report mortality ratios of 286% for males and 363% for females aged 20 and over with moderate to profound ID in the United Kingdom between 1993 and 2005. The reason for the disparity may be that those studies did not exclude persons with physical disability, epilepsy, or Down syndrome. The latter often have congenital heart defects and develop heart disease at a young age. They may represent a large portion of the overall population with ID.²⁸

The results of Tables 2 and 3 can be used to construct life tables, and thus to obtain life expectancies or median survival times. The

application of the excess death rates and mortality ratios reported here are, if anything, optimistic, as they preclude the possibility of loss of function. In addition, it should be emphasized that these reflect only the excess mortality due to ID, and not that due to frequently concomitant medical conditions and/or physical disabilities. If other conditions pertain, they should be rated separately.

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