

# Behavioral Capabilities and Mortality Risk in Adults With and Without Down Syndrome

**David Strauss**

University of California, Riverside

**Warren B. Zigman**

New York State Institute for Basic Research in Developmental Disabilities

---

Among adults with mental retardation, mortality rates for those with Down syndrome are higher than for those without Down syndrome. We studied age-related changes in functioning and their relation to subsequent mortality in adults with mental retardation. Among people without Down syndrome, recent loss of basic skills was associated with substantially elevated mortality rates. This was not so in the Down syndrome group, however. Adults with Down syndrome tended to experience regression in adaptive behavior earlier than did those without Down syndrome. Incidence rates in the two groups diverged subsequent to age 40. Adults without Down syndrome, however, did also tend to regress when older.

---

Over the past half century, the average lifespan of people with mental retardation has risen dramatically (Carter & Jancar, 1983; Durkin, Schupf, Stein, & Susser, 1994). Nevertheless, age-specific mortality risk is still increased in adults with Down syndrome compared to the overall population of people with mental retardation (Strauss & Eyman, 1996). In addition, virtually all individuals with Down syndrome 35 years of age and older will develop neuropathology consistent with a neuropathological diagnosis of Alzheimer disease (Ball & Nuttall, 1980; Jervis, 1948; Malamud, 1972; Solitaire & Lamarche, 1966; Wisniewski, Wisniewski, & Wen, 1985).

Increased mortality later in life may be due to a generalized manifestation of premature aging (Brown, 1985; Martin, 1978) or specifically associated with the development of Alzheimer disease, that is,

manifested clinically as significant regression in adaptive functioning (Burt, Loveland, & Lewis, 1992; Evenhuis, 1990; Jervis, 1948; Lai & Williams, 1989; Malamud, 1972; Silverstein et al., 1988; Silverstein, Herbs, Nasuta, & White, 1986; Thase, Liss, Smeltzer, & Maloon, 1982; Thase, Tigner, Smeltzer, & Liss, 1984; Wisniewski et al., 1985; Zigman, Schupf, Lubin, & Silverman, 1987; Zigman, Schupf, Silverman, & Sterling, 1989; Zigman, Schupf, Sersen, & Silverman, 1995).

A number of cross-sectional studies (e.g., Silverstein et al., 1988; Silverstein et al., 1986; Zigman et al., 1987; Zigman et al., 1989) have provided data regarding age-specific levels of functioning for individuals with mental retardation, with and without Down syndrome, on a number of adaptive behavior domains. These studies generally support the proposition that

---

adults with Down syndrome over the age of 50 exhibit poorer performance on these domains than do either younger individuals with Down syndrome or younger and older individuals without Down syndrome (Collacott, 1992, 1993; Silverstein et al., 1988; Silverstein et al., 1986; Zigman et al., 1987; Zigman et al., 1989). The cross-sectional nature of these studies, however, prohibits the direct test of the hypothesis that individuals with Down syndrome are exhibiting actual behavioral regression.

In two longitudinal studies Schupf, Silverman, Sterling, and Zigman (1989) and Zigman et al. (1995) have attempted to describe the prevalence rates of dementia in cohorts of both live and deceased adults with Down syndrome using a series of more or less restrictive diagnostic criteria. Although prevalence rates varied as a function of cohort, study design, and criterion utilized, two major findings were observed: (a) adults with Down syndrome over the age of 50 were significantly more likely to exhibit regression, regardless of measurement criteria employed, and (b) the regression in functioning exhibited by adults with Down syndrome over the age of 50, presumably due to Alzheimer-type neuropathology, cannot be totally accounted for by their increased risk for terminal drop due to premature mortality.

There are, however, virtually no studies in which investigators have attempted to describe the incidence of behavioral regression, or the relation of behavioral change and mortality in adults with mental retardation, with and without Down syndrome. Our focus in the current investigation was on these issues. Specifically, we examined (a) the cross-sectional age-specific prevalence rates of optimal- and minimal-functioning in toileting, eating, and ambulation skills as a function of Down syndrome etiology (to replicate and extend previous research findings); (b) the longitudinal relation of clinically significant regression in these skills to subse-

quent mortality risk; and (c) the longitudinal age-specific incidence rates for clinically significant regression in these skills as a function of Down syndrome etiology. Our specific interest in these three dependent variables follows from previous findings that declines in these areas are powerful predictors of mortality (e.g., Eyman, Olmstead, & Grossman, 1993; Strauss & Kastner, 1996) and from description of the clinical signs of Alzheimer-type dementia in adults with Down syndrome (Lai & Williams, 1989).

In order to determine age-specific incidence rates for clinically significant regression, we included in our diagnostic criteria a requirement that negative changes not be reversed upon subsequent testing. The requirement of two consecutive measures of negative change was imposed to exclude changes in skills that may result from transient illnesses, occasional behavior problems, or questions relating to data reliability and validity.

## Method

### *Instrument*

Our data source was the Client Development Evaluation Report (Department of Developmental Services, 1978), a statewide information system providing diagnostic, health, placement, and behavioral information. It is completed annually (additionally on moving to a new residence) for anyone receiving services from the California Department of Developmental Services ( $N > 100,000$ ). Further details on this instrument, including data regarding its more than satisfactory reliability, are available in research conducted by Eyman and his colleagues (e.g., Arias, Ito, & Takagi, 1983; Eyman, Call, & White, 1991; Eyman, Grossman, Chaney, & Call, 1990; Eyman, Grossman, Chaney, & Call, 1993; Eyman, Olmstead et al., 1993; Harris, Eyman, & Mayeda, 1982; Widaman, 1984; Widaman, Stacy, & Borthwick, 1985).

## Subjects

Subjects were all adults with developmental disabilities over the age of 40 who received services from the California Department of Developmental Services between January 1980 and December 1992 ( $N = 22,740$ ). We included all people age 40 or over in the study, even though some authors have used thresholds such as 50 or 55 to define *elderly* (e.g., Haveman & Maaskant, 1989; Krauss & Seltzer, 1986). We chose this age because our data indicated that prevalence rates for toileting and other skills, and the rates of loss of these skills, vary substantially between ages 40 and 50 to 55 (especially for people with Down syndrome).

The population was stratified according to data on the Client Development Evaluation Report regarding (a) the presence or absence of an etiological diagnosis of Down syndrome, according to the *International Classification of Diseases-9th edition* (U.S. Department of Health and Human Services, 1980) and (b) level of mental retardation (mild/moderate/severe [higher functioning] and profound/other [lower functioning]). The latter group included individuals with "suspected mental retardation," a California category consisting of people whose level of mental retardation has never been formally tested. Despite the label "suspected," these people in many respects appear to be comparable to the group of individuals with profound mental retardation. (For further information regarding this group see Eyman, Grossman et al., 1993; Eyman, Olmstead et al., 1993; Strauss, Eyman, & Grossman, in press). The severe group was combined with the mild/moderate group rather than the profound group because in most of the analyses, results for individuals with severe mental retardation more closely resembled those for the mild/moderate group. Initially, a third stratification variable, gender, was included in the analyses, but it was dropped as the results reflected no major differences. Table 1 displays demographics of the sample.

**Table 1**  
Study Population by Down Syndrome Status, Age, and Level of Mental Retardation

| Demographic information     | With Down syndrome |      | Without Down syndrome |      | Total<br><i>n</i> |
|-----------------------------|--------------------|------|-----------------------|------|-------------------|
|                             | <i>n</i>           | %    | <i>n</i>              | %    |                   |
| Total sample                | 2,013              | 8.9  | 20,727                | 91.1 | 22,740            |
| Age group                   |                    |      |                       |      |                   |
| 40 ≤ age < 50               | 1,619              | 9.4  | 15,537                | 90.6 | 17,156            |
| 50 ≤ age < 60               | 834                | 10.3 | 7,242                 | 89.7 | 8,076             |
| 60 ≤ age < 70               | 267                | 6.6  | 3,753                 | 93.4 | 4,020             |
| Age ≥ 70                    | 29                 | 2.0  | 1,431                 | 98.0 | 1,460             |
| Level of mental retardation |                    |      |                       |      |                   |
| Mild, moderate, and severe  | 1,534              | 9.0  | 15,483                | 91.0 | 17,017            |
| Profound and other          | 479                | 8.4  | 5,244                 | 91.6 | 5,723             |

Note. Some overlapping existed among the groups because some persons were counted in two consecutive age groups.

## Adaptive Skills

We focused on three adaptive skills: toileting, eating, and ambulation. On the Client Development Evaluation Report, these variables are scored on scales with four or more levels, but for purpose of analyses we collapsed each variable to a 3-point scale: high, medium, or low. Operational definitions were as follows. *Toileting*: high—goes to toilet by self, completes by self; low—not toilet trained or habit trained. *Eating*: high—uses eating utensils with no spillage; low—does not feed self, must be fed completely. *Ambulation*: high—walks well alone at least 6.16 m, balances well; low—does not walk. The intermediate levels were combined (into the medium group) because preliminary analysis indicated them to be fairly homogeneous with respect to mortality rates.

## Results

### Cross-Sectional Analyses (Prevalence Rates)

For each of the three skills, we stratified the Client Development Evaluation Report evaluations on the basis of etiology (Down syndrome vs. not Down syndrome), level

of mental retardation (higher functioning vs. lower functioning), and age group (i.e., 40 to 44, 45 to 49, 50 to 54, 55 to 59, 60 to 64, 65 to 69, and 70+), resulting in a  $2 \times 2 \times 7$  design. To describe the prevalence of a particular skill in each subgroup, we computed the number of people in the group who displayed the highest level of skill together with the number displaying the lowest level. Inspection of the data suggested that tracking only the highest and lowest levels of the skill provided a satisfactory picture of the pattern of skill change with age. The average number of subjects per subgroup was about 7,000, large enough that it appeared preferable to report the raw percentages rather than those derived from a fitted statistical model. The exceptions were the subgroups of people with Down syndrome over 65 years of age, where the reduced sample sizes would lead to unstable estimates; these groups were excluded from the analysis.

To compare the subgroups of people with and without Down syndrome with respect to age-related prevalence of full skills in the three adaptive behaviors, we fitted logistic regression models as a function of age group, level of mental retardation, and etiology. For all three variables, and both levels of mental retardation, decrease in scores across age groups was significantly steeper in the group with Down syndrome than in the group without Down syndrome (linear logistic regression model [Hosmer & Lemeshow, 1989]; likelihood ratio  $\chi^2$  test,  $p < .01$ ). A corresponding comparison between adults with and without Down syndrome was made regarding the age-related prevalence of the proportions with no skill. Once again, all six analyses showed significantly higher prevalence rates in the subgroups with Down syndrome, chi-square tests,  $p < .01$ .

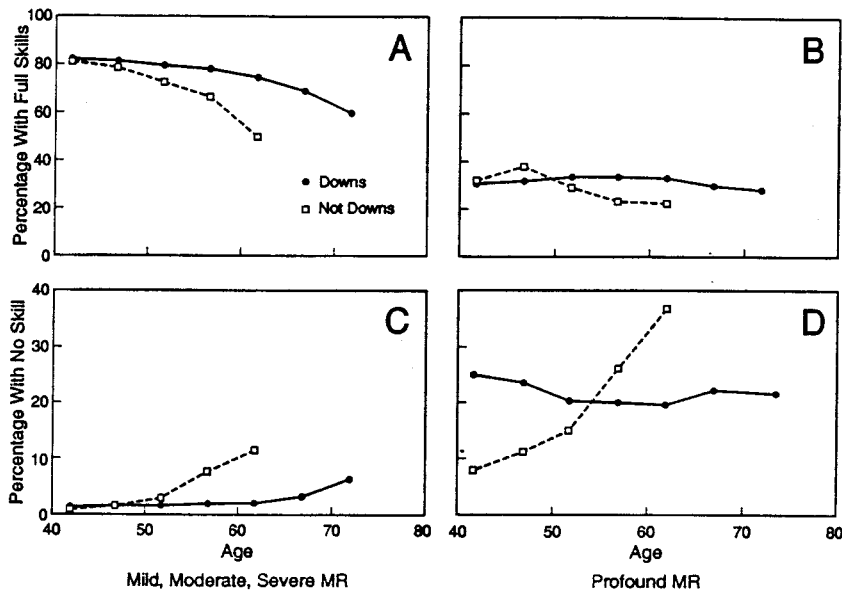
We compared prevalence rates for adults without Down syndrome who were over 70 years of age with those age 60 to 69 years. In all six cases the subgroups of adults over age 70 contained proportion-

ally fewer people with full skills and more people with no skills than did the younger groups (two-sample  $z$  test,  $p < .01$  for all comparisons.) Figures 1, 2, and 3 display the age-related prevalence rates as a function of adaptive skill and level of mental retardation.

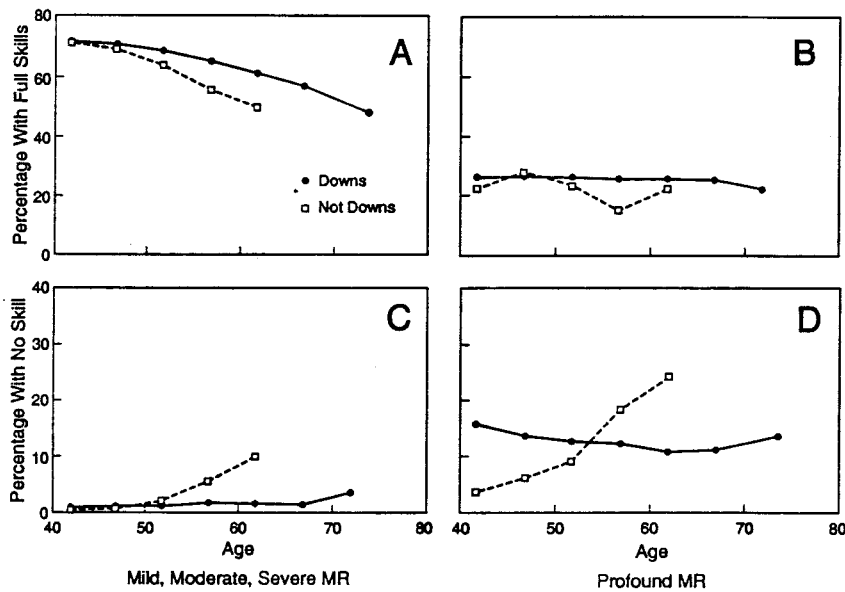
### ***Mortality Analyses***

In this component of the study, we focused on people whose current Client Development Evaluation Report rating was at the low level. Within this group we compared people at the lowest level on the previous Client Development Evaluation Report (the "did not regress" group) with people whose previous Client Development Evaluation Report indicated medium skill ("partial regression") or high skill ("full regression"). The dependent variable was 24-month mortality. Probabilities were computed using a modification of the product-limit survival method, with standard errors derived from the Greenwood formula (Cox & Oakes, 1984). The results for all three adaptive skills for subjects without and with Down syndrome are displayed in Tables 2 and 3, respectively. Regarding the toileting variable, 24-month mortality for adults without Down syndrome is nearly doubled for the "partial regression" group and more than tripled for the "full regression" group. As can be seen in Table 1, for both eating and ambulation skills, the "partial regression" group again has a 24-month risk of death that is about double that of the baseline group, and the risks in the "full regression" group are more than tripled. The consistency of the results across the three skills is noteworthy.

The pattern for people with Down syndrome (see Table 3) is markedly different. The 24-month mortality probabilities for those who had not recently regressed ranged from 27% to 36%, depending on the skill considered. These rates were dramatically higher than those for the group without Down syndrome. Within the group with Down syndrome, more-



**Figure 1.** Proportion of people with high or with low toileting skills by age and by Down syndrome status. Number of Client Development Evaluation Report evaluations: mild/moderate/severe, not Down syndrome—from 33,266 (age group 40 to 44 years) to 10,617 (age 65+); profound/suspected, not Down syndrome—from 12,354 (age group 40 to 44 years) to 3,112 (age 65+); mild/moderate/severe, Down syndrome—3,578 (age 40 to 44 years) to 632 (age 60+); profound/suspected, Down syndrome—1,036 (ages 40 to 44) to 295 (age 60+).



**Figure 2.** Proportion of people with full or no eating skills, by age and by Down syndrome status. The sample sizes are the same as in Figure 1.

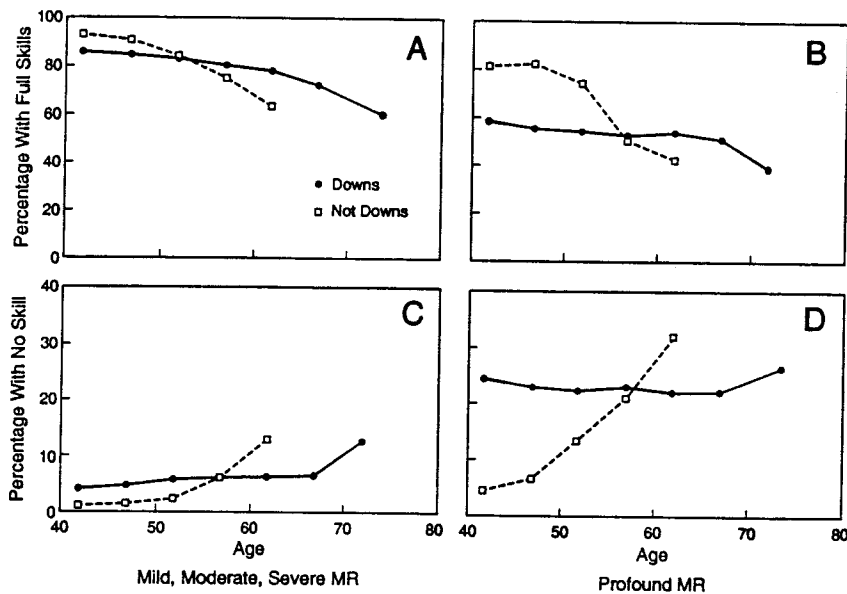


Figure 3. Proportion of people with full or no ambulatory skills, by age and by Down status. The sample sizes are the same as in Figures 1 and 2.

**Table 2**  
Mortality Probabilities (24-Month) for People Without Down Syndrome Whose Current Client Developmental Evaluation Report (CDER) Indicates Lowest Level of Adaptive Skills

| Adaptive skill | Levels from previous CDER |       |                 |        |     |     |      |    |     |
|----------------|---------------------------|-------|-----------------|--------|-----|-----|------|----|-----|
|                | Low                       |       |                 | Medium |     |     | High |    |     |
|                | %                         | n     | SE <sup>a</sup> | %      | n   | SE  | %    | n  | SE  |
| Toileting      | 6.8                       | 5,607 | 0.4             | 11.1   | 469 | 1.5 | 23.8 | 97 | 4.5 |
| Eating         | 7.0                       | 3,435 | 0.4             | 15.8   | 293 | 2.2 | 29.9 | 55 | 6.3 |
| Ambulation     | 6.6                       | 7,882 | 0.4             | 12.1   | 349 | 2.0 | 24.4 | 88 | 3.9 |

<sup>a</sup>Standard error.

**Table 3**  
Mortality Probabilities (24-Month) for People With Down Syndrome Whose Current Client Developmental Evaluation Report (CDER) Indicates Lowest Level of Adaptive Skills

| Adaptive skill | Levels from previous CDER |     |                 |                          |    |     |
|----------------|---------------------------|-----|-----------------|--------------------------|----|-----|
|                | Low                       |     |                 | Medium/high <sup>b</sup> |    |     |
|                | %                         | n   | SE <sup>a</sup> | %                        | n  | SE  |
| Toileting      | 29.3                      | 323 | 2.6             | 24.5                     | 97 | 4.4 |
| Eating         | 35.7                      | 187 | 3.7             | 37.8                     | 81 | 5.6 |
| Ambulation     | 27.7                      | 304 | 2.7             | 28.6                     | 79 | 5.3 |

<sup>a</sup>Standard error. <sup>b</sup>Categories combined because of small sample sizes.

tive skills is a major mortality predictor regardless of whether the skill loss was recent.

### Age-Specific Incidence Analyses

By *incidence rate* for a loss of skill, we mean the proportion of people having that skill who would be expected to lose it in a given time interval. We used a time interval of one year. The term *incidence rate* is used in the epidemiological sense, as an occurrence/exposure ratio (Rothman, 1986). Suppose that A, B, C, and D are four consecutive Client Development Evaluation Report assessments. A regres-

over, the data show no effect of having previously regressed. In summary, among people with Down syndrome who are over 40 years old, the lack of basic adap-

sion in skill is said to have occurred between B and C if both A and B are at the high levels, and both C and D are at the medium or low levels. The requirement of two consecutive high followed by two medium or low Client Development Evaluation Report assessments was imposed so as to exclude temporary gains or losses in skill (e.g., those that may result from a brief illness).

A diagram illustrating the procedure may be helpful. Figure 4 shows the time axis, with a stream of 10 Client Development Evaluation Report assessments for a hypothetical individual. The large dots correspond to Client Development Evaluation Report assessments where the individual was rated at the high level of the skill; the small dots indicate the medium or low levels. An individual is "exposed" to a regression in skill whenever the two previous Client Development Evaluation Report assessments are high and there are at least two subsequent Client Development Evaluation Report assessments. The exposed time in Figure 4 is represented by the three hatched inter-Client Development Evaluation Report intervals. Only one occurrence of regression takes place during the three exposed intervals illustrated in the figure, in interval  $t_5$ . The individual in this example thus contributes one occurrence of regression in a total "exposure" time of  $(t_3 + t_4 + t_5)$ . Other definitions of regression would have been

possible, but preliminary analyses revealed that such alternatives did not add appreciably to the picture.

The procedure resulted in between 50,000 and 70,000 exposed inter-Client Development Evaluation Report intervals, depending on which of the three skills was considered. If B and C are a consecutive pair of Client Development Evaluation Reports and the interval between them is an exposed one, then the dependent variable DECLINE is taken to be 1 if a regression in skill occurred and 0 if it did not. This was related to the following set of explanatory variables: age (actual age at time of the Client Development Evaluation Report B); etiology (1 if Down syndrome, 0 if not); level of mental retardation (1 if higher functioning, 0 if lower functioning); and logtime (the logarithm of the time interval between B and C).

For each skill variable in turn, a logistic regression model of DECLINE on the covariates was developed, using standard modeling techniques (Hosmer & Lemeshow, 1989, chapters 4 and 5). These models express the logarithm of the odds on a regression in skill (i.e.,  $\log\{\text{Prob}(\text{DECLINE} = 1)/\text{Prob}(\text{DECLINE} = 0)\}$ ), as a function of age, etiology, and the other variables. The log-odds on a loss of skill increased linearly with age for the no Down syndrome group but at a higher than linear rate for the Down syndrome group. For the Down syndrome group an

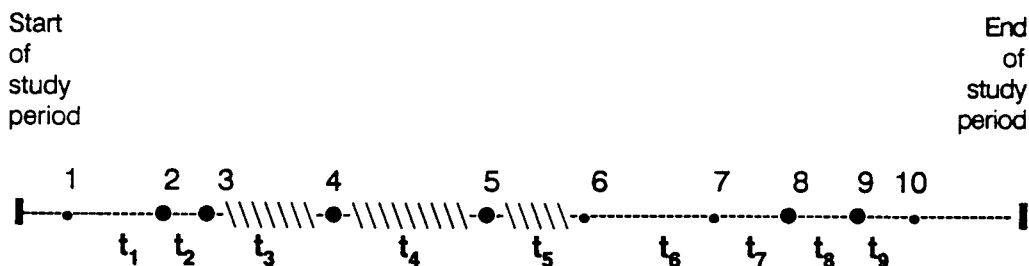


Figure 4. Illustration of definition of *regression in skill* as used in incidence rate computations. The example shows a hypothetical individual with 10 Client Development Evaluation Reports during the study period. Large dots indicate high skill (in toileting, eating, or ambulation); small dots denote medium or low skills. The interval  $t_3$  is shaded to indicate that the individual was at risk, or exposed, to a regression, because the two preceding reports showed high skill and two future reports (4 and 5) were available. Similarly, Intervals  $t_4$ ,  $t_5$  were exposed to regression. Interval  $t_6$  was not because it was followed by only one Client Development Evaluation Report. There was one occurrence of regression, in Interval  $t_5$ .

additional quadratic term in age sufficed in the case of toileting and eating, whereas the 1.5th power of age proved to give a better fit in the case of ambulation. The differences in rates of increase of individuals with and without Down syndrome were highly significant for all three skills. The results of the model-fitting procedure for the three adaptive skills are shown in Figures 5, 6, and 7. For each mental retardation level, the curves plot the probability of a regression in toileting, eating, or ambulation in a given year against AGE.

Figure 5 shows incidence rates for loss of toileting skills for adults at the two levels of mental retardation. The regres-

sion rates in the group with lower functioning adults were systematically higher than in the group with higher functioning individuals. Further, all subgroups showed an increasing rate of regression with age. The most striking feature in Figure 5 is the contrast of the groups with and without Down syndrome. Thus, as can be seen in the first panel of Figure 5, the rate for adults with Down syndrome was slightly higher than that for adults without Down syndrome at age 40, but the difference between the two widened rapidly with age. By age 65, the rate for the Down syndrome group was more than double that of the other group. Among people

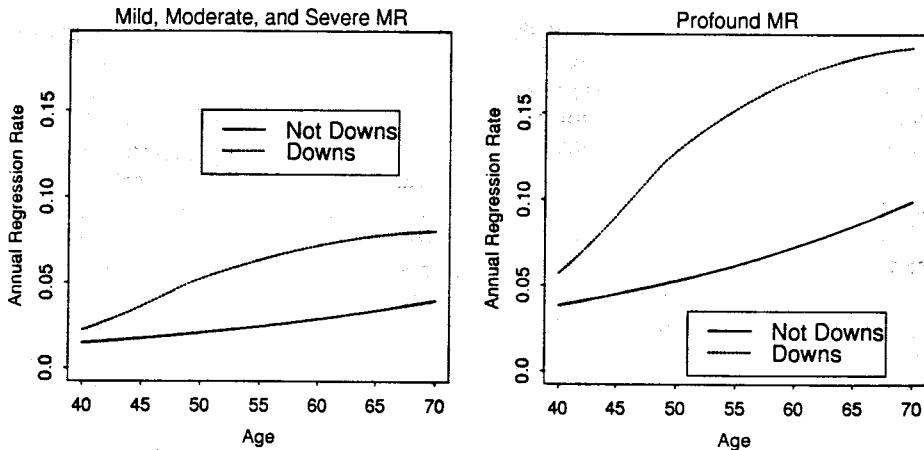


Figure 5. Rates of regression in toileting skill by age and by Down syndrome status. The curves derive from logistic models described in the text.

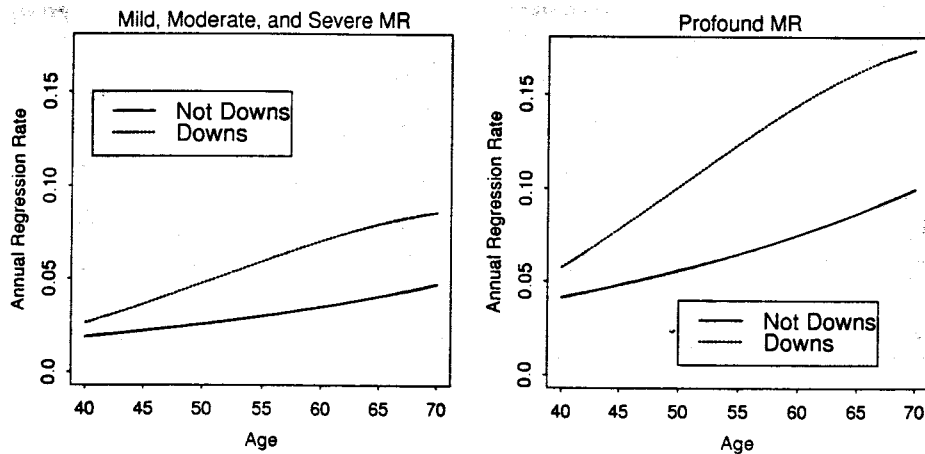


Figure 6. Rates of regression in eating skill by age and Down syndrome status.



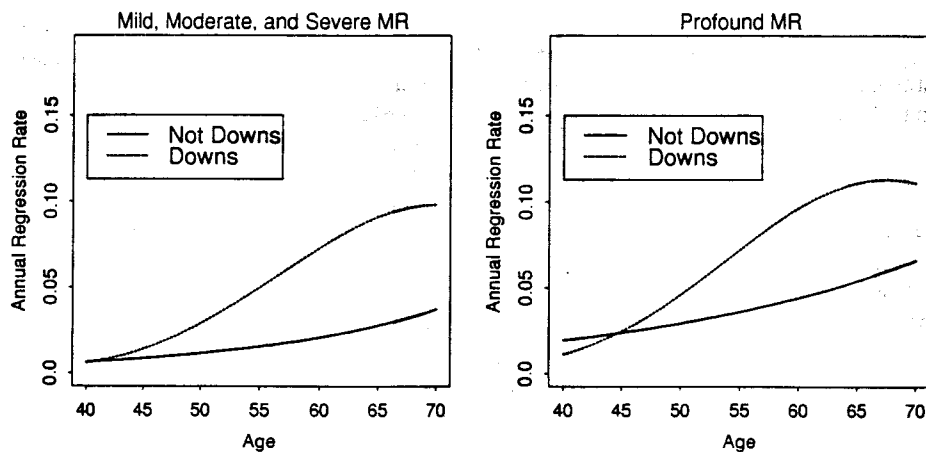


Figure 7. Rates of regression in ambulatory skill.

with mental retardation who were lower functioning, the rate for adults with Down syndrome was again only slightly higher at age 40, but the surviving people with Down syndrome who still had toileting skills at age 65 had almost a 20% chance of losing their skill in a given year. This rate was about triple that of the group without Down syndrome.

Figures 6 and 7, for eating and ambulation skills, show similar accelerating regression rates with age for people with Down syndrome. Figure 7 indicates that at age 40 in the group of people with mental retardation who are lower functioning, the rate of adults with Down syndrome is actually the lower of the two (this is confirmed by inspection of the raw data), but with advancing age the pattern sharply reverses itself.

The incidence rates shown in Figures 5, 6, and 7 paint a picture that is broadly compatible with the prevalence graphs in Figures 2, 3, and 4: a general decline with age that is much more marked among adults with Down syndrome. There is a distinction to be made, however; the prevalence rates are cross-sectional, whereas the incidence rates refer to longitudinal changes for individuals. For many purposes, including that of using regression in skills as a predictor of mortality, incidence rather than overall prevalence is the relevant statistic.

## Discussion

We had three general goals for the current investigation: to determine (a) age-specific levels of adaptive functioning in adults with mental retardation with and without Down syndrome, (b) the relation between regression in adaptive functioning and subsequent mortality, and (c) the incidence rate of significant regression in functioning of adults with mental retardation with and without Down syndrome. These three goals have a similar inclusive purpose, namely, to determine whether there are age-related changes in functioning or mortality in adults with mental retardation that may be related to the development of Alzheimer-type neuropathological changes. Collectively, the three sets of results support the hypothesis that individuals with Down syndrome are at earlier and higher risk for adaptive regression, whether prevalence, incidence, or mortality rates serve as the dependent variable. Each set of findings is described separately.

First, cross-sectional findings regarding age-specific levels of functioning in toileting, eating, and ambulation skills replicated previous research on similar groups of subjects (Silverstein et al., 1988; Silverstein et al., 1986; Zigman et al., 1987; Zigman et al., 1989). Cross-sectional comparisons, however, must be interpreted

with care (Breslow & Day, 1980). Specifically, the figures should not be interpreted as age-plots of an individual's probability of having the skill in question. In particular, the proportions for the older age groups were based on the pool of surviving subjects, and these survivors were people whose average health at age 40, for example, was better than that of those who did not survive. This healthy survivor effect is likely to be most marked in the group of adults with Down syndrome, where the attrition rate was especially high due to premature mortality.

It is worth observing that adults over age 70 with mental retardation without Down syndrome had lower levels of skills than did younger adults without Down syndrome. This age-specific decline appeared after the onset of age-specific declines for adults with mental retardation who had Down syndrome. This clinical finding is consistent with the recent neuropathological findings suggesting that the age-specific prevalence rates of Alzheimer-type neuropathology may be higher in adults with mental retardation without Down syndrome compared to adults without mental retardation (Barcikowska et al., 1989; Popovitch et al., 1990), and the recent finding reported by Zigman, Schupf, Zigman, and Silverman (1993) that adults with mental retardation due to causes other than Down syndrome may be prone to increased rates of age-related regression in adaptive functioning. Given the current size of the population of adults with mental retardation due to other causes and its projected growth in the future, it is important to determine the prevalence and incidence of mortality and dementia in these cohorts. Note that the healthy survivor effect inherent in this cross-sectional analysis has probably reduced the magnitude of the age-related declines.

Second, among the adults without Down syndrome there was notable consistency among the longitudinal relations between mortality and regression in adaptive behavior across the three skills examined. Individuals who exhibited a partial

change in functioning (i.e., -1 level change) in any of the three skills assessed displayed mortality rates about double those displayed by individuals with no changes in adaptive abilities. Individuals with full changes in functioning (i.e., -2 level changes) in any of the three skills assessed displayed mortality rates that were roughly tripled. These analyses provide further support for the data presented by Eyman, Call, and White (1989), but overcome a possible confound in the earlier analysis. Specifically, our analysis differs from their study in that Eyman et al. compared people who had not recently changed their level with those who had recently regressed. People in the latter group were, thus, currently all at the lower level, whereas the former group contained a mixture of the two (with the majority at the higher level). As a result, it was not possible to separate the effects of regression from the effect of the current level of skill.

Third, as shown in Figures 5, 6, and 7, the incidence rates of regression in skills rose more steeply for adults with than for those without Down syndrome. This was so even before age 50; for example, the regression rate for toileting among people age 40 to 49 was 3.8% ( $\pm 0.4\%$ ) in the group with Down syndrome and 2.1% ( $\pm 0.1\%$ ) in the group without Down syndrome, two-sample  $z$  test,  $p < .01$ . This finding is consistent with Strauss and Eyman's (1996) finding of a significantly higher relative risk (i.e., mortality in adults with Down syndrome relative to adults with mental retardation due to other causes) in the age range 45 to 49 than in younger age groups. These data vary from those presented by Zigman et al. (1995), who found significant differences due to etiology only after age 50. However, the current data do indicate a much sharper contrast between the two groups after age 50 (Figures 5, 6, and 7). Differences between these two studies also may be due to differences in the analytic strategies (Zigman et al., 1995, presented prevalence data) or possibly to

state-specific differences in the definition of consumer characteristics within the service system.

As can be seen in Figures 5, 6, and 7, there was a steady increase in incidence rate for adults with mental retardation and no Down syndrome. This analysis is not confounded by healthy survivor effects, as was the case in the cross-sectional analysis reported earlier, and, thus, provides confirmation of the finding of adaptive regression in elderly adults with mental retardation due to other causes. Our conclusions in this study are clear and reliable across the range of adaptive domains assessed. Adults with Down syndrome are clearly subject to a substantially increased risk of adaptive regression at younger ages than are individuals with mental retardation due to other causes. Adults with mental retardation and no Down syndrome, however, do seem to display regression in adaptive behavior, some years after the onset of such regression in adults with Down syndrome. Whether these changes in either group are related to the development of Alzheimer-type neuropathology or are related to patterns of benign senescence or comorbid medical conditions will have to await further neuropathological research on the results of autopsies.

A limitation of the present data is that there are no recognized standardized criteria for the diagnosis of dementia in adults with mental retardation with and without Down syndrome. Recent criteria specified by Aylward, Burt, Thorpe, Lai, and Dalton (1995) represent an important beginning to the establishment of a "gold standard." Their work on the reliability and validity of these criteria is still in progress. For the present time, the use of incidence data must rest on an operationally defined condition (in this instance a specified degree of regression in functioning). Without neuropathological confirmation of the Alzheimer-type pathology, and clinical diagnostic confirmation of Alzheimer-type dementia, these data must be considered to be tentative. However,

they do add to a growing body of research displaying significant longitudinal changes in adaptive functioning in adults with mental retardation with and without Down syndrome.

## References

- Aylward, E. H., Burt, D. B., Thorpe, L. U., Lai, F., & Dalton, A. J. (1995). *Diagnosis of dementia in individuals with intellectual disability*. Washington, DC: American Association on Mental Retardation.
- Arias, M., Ito, E., & Takagi, N. (1983). Concurrent validity of the Client Development and Evaluation Report. In A. B. Silverstein & A. L. Fluharty (Eds.), *Pacific State archives VIII* (pp. 28-33). Pomona: University of California at Los Angeles, Developmental Disabilities Immersion Program.
- Ball, M. J., & Nuttall, K. (1980). Neurofibrillary tangles, granulovacuolar degeneration, and neuron loss in Down syndrome: Quantitative comparison with Alzheimer dementia. *Annals of Neurology*, 7, 462-465.
- Barcikowska, M., Silverman, W. P., Zigman, W. B., Kozlowski, P. B., Kujawa, M., Rudelli, R., & Wisniewski, H. M. (1989). Alzheimer-type neuropathology and clinical symptoms of dementia in mentally retarded people without Down syndrome. *American Journal on Mental Retardation*, 93, 551-557.
- Breslow, N. E., & Day, N. E. (1980). *Statistical methods in cancer research: Vol. 1. The analysis of case-control studies*. Lyon, France: International Agency on Cancer Research.
- Brown, W. T. (1985). Genetics of aging. In M. P. Janicki & H. M. Wisniewski (Eds.), *Aging and developmental disabilities: Issues and approaches* (pp. 185-194). Baltimore: Brookes.
- Burt, D. B., Loveland, K. A., & Lewis, K. R. (1992). Depression and the onset of dementia in adults with mental retardation. *American Journal on Mental Retardation*, 96, 502-511.
- Carter, G., & Jancar, J. (1983). Mortality in the mentally handicapped: A 50 year survey at the Stokes Park group of hospitals. *Journal of Mental Deficiency Research*, 27, 143-156.
- Collacott, R. A. (1992). The effect of age and residential placement on adaptive behaviour of adults with Down's syndrome. *British*

- Journal of Psychiatry*, 161, 675-679.
- Collacott, R. A. (1993).** Epilepsy, dementia and adaptive behaviour in Down's syndrome. *Journal of Intellectual Disability Research*, 37, 153-160.
- Cox, D., & Oakes, D. (1984).** *Analysis of survival data*. London: Chapman and Hall.
- Department of Developmental Services. (1978).** *Client Development Evaluation Report*. Sacramento, CA: Author.
- Durkin, M. S., Schupf, N., Stein, Z. A., & Susser, M. W. (1994).** Epidemiology of mental retardation. In M. Levene & R. Lifford (Eds.), *Fetal and neonatal neurology and neurosurgery* (2nd ed.). London: Churchill, Livingstone.
- Evenhuis, H. M. (1990).** The natural history of dementia in Down's syndrome. *Archives of Neurology*, 47, 263-267.
- Eyman, R. K., Call, T. L., & White, J. F. (1989).** Mortality of elderly mentally retarded persons in California. *The Journal of Applied Gerontology*, 8, 203-215.
- Eyman, R. K., Call, T. L., & White, J. F. (1991).** Life expectancy of persons with Down syndrome. *American Journal on Mental Retardation*, 95, 603-612.
- Eyman, R. K., Grossman, H. J., Chaney, R. H., & Call, T. L. (1990).** The life expectancy of profoundly handicapped people with mental retardation. *New England Journal of Medicine*, 323, 584-589.
- Eyman, R. K., Grossman, H. J., Chaney, R. H., & Call, T. L. (1993).** Survival of profoundly disabled people with severe mental retardation. *American Journal of Diseases of Children*, 147, 329-336.
- Eyman, R. K., Olmstead, C. E., Grossman, H. J., & Call, T. L. (1993).** Mortality and the acquisition of basic skills by children and adults with severe disabilities. *American Journal of Diseases of Children*, 147, 216-222.
- Harris, C. W., Eyman, R. K., & Mayeda, T. (1982).** *An interrater reliability study of the Client Development Evaluation Report: Final report to the California Department of Developmental Disabilities*. Pomona: University of California at Los Angeles, Mental Retardation Research Center, Lanterman State Hospital.
- Haveman, M. J., & Maaskant, M. A. (1989).** Defining fragility of the elderly severely mentally handicapped according to mortality risk, morbidity, motor handicaps and social functioning. *Journal of Mental Deficiency Research*, 33, 389-397.
- Hosmer, D. W., & Lemeshow, S. (1989).** *Applied logistic regression*. New York: Wiley.
- Jervis, G. A. (1948).** Early senile dementia in mongoloid idiocy. *American Journal of Psychiatry*, 105, 102-106.
- Krauss, M. W., & Seltzer, M. M. (1986).** Comparison of elderly and adult mentally retarded persons in community and institutional settings. *American Journal of Mental Deficiency*, 91, 237-243.
- Lai, F., & Williams, R. S. (1989).** A prospective study of Alzheimer disease in Down syndrome. *Archives of Neurology*, 46, 849-853.
- Malamud, N. (1972).** Neuropathology of organic brain syndromes associated with aging. In C. M. Gaitz (Ed.), *Aging and the brain* (pp. 63-77). New York: Plenum Press.
- Martin, G. M. (1978).** Genetic syndromes in man with potential relevance to pathobiology of aging. In D. Bergsma & D. E. Harrison (Eds.), *Genetic effects on aging, birth defects: Original articles* (Vol. XIV, pp. 5-39). New York: Liss.
- Popovitch, E. R., Wisniewski, H. M., Barcikowska, M., Silverman, W., Bancher, C., Sersen, E., & Wen, G. Y. (1990).** Alzheimer neuropathology in non-Down's syndrome mentally retarded adults. *Acta Neuropathologica* (Berlin), 80, 362-367.
- Rothman, K. J. (1986).** *Modern epidemiology*. Boston: Little, Brown.
- Schupf, N., Silverman, W. P., Sterling, R. C., & Zigman, W. B. (1989).** Down syndrome, terminal illness and risk for dementia of the Alzheimer type. *Brain Dysfunction*, 2, 181-188.
- Silverstein, A. B., Herbs, D., Miller, T. J., Nasuta, R., Williams, D. L., & White, J. F. (1988).** Effects of age on the adaptive behavior of institutionalized and noninstitutionalized individuals with Down syndrome. *American Journal on Mental Retardation*, 92, 455-460.
- Silverstein, A. B., Herbs, D., Nasuta, R., & White, J. F. (1986).** Effects of age on the adaptive behavior of institutionalized individuals with Down syndrome. *American Journal of Mental Retardation*, 90, 659-662.
- Solitaire, G. C., & Lamarche, J. B. (1966).** Alzheimer's disease and senile dementia as seen in mongoloids: Neuropathological observations. *American Journal of Mental Deficiency*, 70, 840-848.
- Strauss, D., & Eyman, R. K. (1996).** Mortal-

- ity of people with mental retardation in California, 1986–1991. *American Journal of Mental Retardation*, 100, 643–653.
- Strauss, D., Eyman, R. K., & Grossman, H. J. (in press).** The prediction of mortality in children with severe mental retardation: The effect of placement. *American Journal of Public Health*.
- Strauss, D., & Kastner, T. (1996).** Comparative mortality of people with developmental disabilities in institutions and the community. *American Journal of Mental Retardation*, 101, 26–40.
- Thase, M. E., Liss, L., Smeltzer, D., & Maloon, J. (1982).** Clinical evaluation of dementia in Down's syndrome: A preliminary report. *Journal of Mental Deficiency Research*, 26, 239–244.
- Thase, M. E., Tigner, R., Smeltzer, D. J., & Liss, L. (1984).** Age-related neuropsychological deficits in Down syndrome. *Biological Psychiatry*, 19, 571–585.
- U.S. Department of Health and Human Services. (1980).** *The international classification of diseases, 9th revision, Clinical modification* (Vol. 1). Washington, DC: U.S. Government Printing Office.
- Widaman, K. F. (1984, August).** *Interrater reliability of adaptive behavior assessments: Item and factor levels.* Paper presented at the annual meeting of the American Psychological Association, Toronto, Ontario.
- Widaman, K. F., Stacy, A. W., & Borthwick, S. A. (1985, May).** *Multitrait-multimethod of evaluating adaptive and maladaptive behavior of mentally retarded people.* Paper presented at the annual meeting of the American Association on Mental Deficiency, Philadelphia.
- Wisniewski, K., Wisniewski, H. M., & Wen, G. Y. (1985).** Occurrence of Alzheimer's neuropathology and dementia in Down syndrome. *Annals of Neurology*, 17, 278–282.
- Zigman, W. B., Schupf, N., Lubin, R. A., & Silverman, W. P. (1987).** Premature regression of adults with Down syndrome. *American Journal of Mental Deficiency*, 92, 161–168.
- Zigman, W. B., Schupf, N., Sersen, E., & Silverman, W. (1995).** Prevalence of dementia in adults with and without Down syndrome. *American Journal on Mental Retardation*, 100, 403–412.
- Zigman, W. B., Schupf, N., Silverman, W. P., & Sterling R. C. (1989).** Changes in adaptive functioning of adults with developmental disabilities. *Australian and New Zealand Journal of Developmental Disabilities*, 15, 277–287.
- Zigman, W. B., Schupf, N., Zigman, A., & Silverman, W. (1993).** Aging and Alzheimer disease in people with mental retardation. In N. W. Bray (Ed.), *International review of research in mental retardation* (Vol. 19, pp. 41–70). New York: Academic Press.

---

Received 8/15/95, first decision 12/19/95, accepted 4/8/96.

---

Preparation of this paper was supported by funds provided by the University of California, Riverside, National Institute of Child Health and Human Development Grant No. HD21056, by New York State through its Office of Mental Retardation and Developmental Disabilities, and by National Institutes of Health Grants No. R29 HD24170, and a subproject of PO1 AG11531 (Henry Wisniewski, principal investigator) to the second author. The authors acknowledge the helpful comments of April Zigman on an earlier version of this paper. They are also grateful to Thomas Call, Deborah Gingras, and Kenneth Lin for substantial assistance. Requests for reprints should be sent to David Strauss, Department of Statistics, University of California, Riverside, Riverside, CA 92521.