Survival of Profoundly Disabled People With Severe Mental Retardation

Richard K. Eyman, PhD; Herbert J. Grossman, MD; Robert H. Chaney, MD; Thomas L. Call, MA

• *Objective.*—**To define further the association between survival and clinical disabilities in profoundly disabled people with mental retardation in an 11-year period.**

Research Design.—**An 11-year follow-up study of the survival of six mutually exclusive subgroups. The presence of severe, profound, or suspected mental retardation and incontinence were considered in all individuals when forming the subgroups. Varying combinations of abilities in mobility, rolling, feeding, and arm-hand use were also considered.**

Participants.—**Six subgroups of severely disabled subjects. Included were 128 248 of 155 851 persons who received services from the California Department of Developmental Services between January 1980 and March 1991.**

Measurements/Main Results.—**Survival estimates for individuals who were immobile and could not roll over were short regardless of arm-hand use or feeding status, as were estimates for people who were tube fed. For individuals who could roll over, but were otherwise immobile, survival was relatively improved.**

Conclusion.—**Individuals who are unable to move their extremities or bodies voluntarily or who require tube feeding have very shortened life expectancies.**

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A number of studies[1-16] have documented relatively high mortality among specific groups of people with mental retardation and severe disabilities. Examples of variables found to be related to life expectancy include basic skills such as ambulation or mobility, method of feeding, bowel and bladder control,[5,7,15,17] level of retardation,[1,16] and, to a lesser extent, cause of retardation and disabilities.[3,10,18,19] However, Roboz,[15] Evans et al,[17] Eyman et al,[5,7,8] and Kudrjavcev et al[11] have argued that immobility, in conjunction with either feeding and toileting difficulties *or* severe cerebral palsy, is especially predictive of poor survival.

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From the School of Education, University of California, Riverside (Dr Eyman and Mr Call); the Departments of Pediatrics, Neurology, and Psychiatry, University of Michigan, Ann Arbor, and the University of Michigan Medical School (Dr Grossman); and UCLA Neuropsychiatric Institute, Mental Retardation Research Center at Lanterman Developmental Center, Pomona (Dr Chaney).

Reprint requests to University of California, School of Education, Riverside, CA 92521-0128 (Dr Eyman).

The purpose of this study was to define further the association between survival and clinical disabilities in profoundly disabled people with mental retardation in an 11-year 3-month period (January 1980 through March 1991). The most important new information is that voluntary arm-hand movement and the ability to roll over were associated with improved survival.

SUBJECTS AND METHODS

Subjects included 128 248 of 155 851 persons who received services from the California Department of Developmental Services between January 1980 and March 1991. All persons had severe disabilities, usually associated with mental retardation, and were referred to one of the 21 regional centers that are contracted by the state to provide services to all individuals who live in their geographic catchment areas. In California, final responsibility for residential placement also is given to the regional centers, which are community-based-private corporations.

During diagnostic evaluations coordinated by the regional centers, levels of retardation were determined. The categories used were consistent with the classification systems in the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders, Revised Third Edition*[20] and the *Manual of Classification in Mental Retardation* of the American Association on Mental Retardation[21] and included profound (IQ of less than 20 to 25), severe (IQ of between 20 to 25 and 35 to 40), moderate (IQ of between 35 to 40 and 50 to 55), and mild (IQ of between 50 to 55 and 70 to 75) retardation. The use of IQ ranges indicates that clinical judgment should be used in addition to estimated IQ in determining degrees of mental retardation.

An additional category is used in California to identify individuals who are suspected to have mental retardation but for whom a level of retardation has not yet been determined because they have not been tested. Most of these people are young and live with their families. In many instances, the diagnostic evaluation teams are uncertain about the degree of retardation in the presence of other profound disabilities or health conditions.

A diagnosis of cerebral palsy is also made at this time based on impaired movement and posture attributable to prenatal or perinatal brain damage. Epilepsy is classified according to the type and frequency of seizures as well as the need for medication for treatment. For the purpose of this study, subjects were subdivided into four groups with generalized seizures: those known to have had less than one seizure per day, those having one or more seizures per day, those who had never had seizures, and those with an undetermined seizure status.

The following characteristics were also assessed relative to survival based on completion of a Client Development Evaluation Report (CDER).[22]

1. *Mobility.*—Subjects were defined as immobile if they lacked

the ability to walk, crawl, creep, or scoot. Some subjects classified as immobile could be seated in a wheelchair with support.

2. *Rolling.*—All subjects were evaluated on their ability to roll from side to side, from back to front only, and from front to back and back to front. A person who could accomplish any voluntary rolling was designated as able to roll, in contrast with others who were unable to perform any of the rolling alternatives.

3. *Hand Use.*—All subjects were evaluated as to whether or not they had any functional use of the hands, eg, the ability to make raking motion with their hands or to grasp with their hands.

4. *Arm Use.*—Persons who could move their arms from their shoulders or extend their arms were distinguished from individuals who had no functional use of their arms.

5. *Toileting Skills.*—Persons who were not toilet trained or habit trained were defined as not toilet trained. Others who had some ability to take care of their toileting needs, ie, were habit trained or could indicate a need to toilet, were considered to be in a toilet-trained category.

6. *Eating Skills.*—Persons who had to be fed completely by others were classified as unable to feed themselves. Finger feeding was categorized as self-feeding.

7. *Tube Feeding.*—Feeding by nasogastric or gastrostomy tubes was recorded for patients who were unable to swallow or who aspirated food frequently. All of these skills were evaluated every year during the study period to ensure that subjects remained correctly defined during follow-up.

The reliabilities of the data on individual characteristics, deaths, and other variables were investigated separately. The reliability and validity of the CDER have been previously found to be satisfactory.[23-26] Items pertaining to skills used for this study had interrater agreements of 87% or greater. Moreover, the California Department of Developmental Services periodically verifies the accuracy of this information because it is used to reimburse service providers.

While descriptive information on cerebral palsy and epilepsy is given in the Table, neither of these variables has been found to be a strong predictor of early mortality in the presence of severe or profound retardation, immobility, being fed by others, or lack of toilet training.[7] The cause of mental retardation is often indeterminate.[27] Some causes can be established, such as certain chromosomal or neurodegenerative diseases, but these causes pertained to a very small percentage of subjects in our sample. In many cases, cause of retardation remains unknown because of lack of accurate gestational or birth history or because appropriate diagnostic tests were not performed.

Instrumentation

The source of information on client characteristics was the CDER.[22] A CDER must be completed once a year and on movement to a new facility for anyone receiving services from the California Department of Developmental Services. This report is the mechanism for local agencies to report residential placement and diagnostic and health information to the state. The report contains a 66-item measure of adaptive behavior. From the CDER we obtained information on age, placement, sex, ethnic background, level of retardation, cause of mental retardation, mobility, and presence of major medical problems, seizures, cerebral palsy, and toileting and eating skills. Because the CDER is administered annually by case workers (when subjects receive community care) or psychiatric technicians (when subjects receive care in institutions), we were able to determine the skill level and whether a subject had died for each year for each individual receiving services from the regional centers between 1980 and 1991. All of these data collectors have been trained in the use of this instrument. Their evaluations are also based on personal observations and, when appropriate, interviews with parents.

Deaths are reported to the California Department of Developmental Services as they occur by the 21 regional centers in California. All deaths occurring between January 1980 and March 1991 were included in the analysis of the data. The at-risk population was approximately 72 000 each year; roughly 980 deaths were reported per year.

Sample

The sample, which was drawn from among 155 851 persons with severe disabilities, included individuals with documented or suspected mental retardation who received services from the State Department of Developmental Services between January 1980 and March 1991. This group included nearly all children (defined by Public Law 94-142 as persons younger than 22 years) with profound and severe mental retardation known to school districts throughout the state. This population also included approximately 50% of children with moderate mental retardation and 10% of the children with mild mental retardation known to California schools. Adults with mental retardation were identified either because they continued to receive services first provided when they were children or because their aging parents were concerned about the care of their children in future years and sought services for them.[28] These individuals lived in a variety of places including their own homes, state institutions, group homes, health care facilities, and skilled nursing facilities.

The Table summarizes the sample available (N=128 248) relative to the total population present between January 1980 and March 1991. Eighteen percent (N=27 603) of the 155 851 persons receiving services were unavailable for follow-up because they moved out of the state or no longer requested services from the state within 2 years of the start of the study. Most people served by the regional centers for 2 years or less were younger children living at home and in the category of suspected mental retardation whose parents no longer requested services after a diagnostic evaluation by the regional center (Table). We wished to standardize the at-risk population to a minimum participation of 2 years.

The survival model used (product limit)[29] accommodated varying times of enrollment as well as censoring, ie, death not observed in the follow-up period. Approximately 70% of the population served by the California system was available for follow-up of more than 5 years and about 40% was available for follow-up throughout the 11-year period. There was no evidence of differential censoring among persons who remained in the regional center system for more than 2 years. More than 60% of the individuals in the subgroups whose survival we studied (Table) either continued to receive services or died in the 11-year period. New admissions were allowed during follow-up on the condition that people newly admitted could be followed up for a minimum of 2 years.

The 128 248 persons who received services from the state were divided into six mutually exclusive subgroups based on different combinations of basic movement and feeding skills. Deficits in all skills were evaluated each year during the 11-year follow-up period. Subjects whose skills improved in the period of study were excluded from analysis because they would then belong in a less vulnerable group with a better life expectancy. The members of all six subgroups had profound, severe, or suspected mental retardation and were incontinent. Subgroup 1 consisted of those who were immobile, could not roll, required tube feeding, and had no arm-hand use. Subgroup 2 consisted of persons who were immobile, could not roll, required tube feeding, but had some arm-hand use. Subgroup 3 consisted of persons who were immobile, could not roll, had no arm-hand use, but could take food if it was fed to them by others. Subgroup 4 consisted of persons who were immobile, could not roll, could take food if fed by others, and had some arm-hand use. Subgroup 5 comprised persons who were immobile, but who could roll over, could take food if fed by others, and had some arm-hand use.

Very few immobile subjects who required continuous tube feeding during the period of this study were able to roll over. As we have shown previously,[30] few people who met the above criteria for inclusion in this study later acquired mobility, feeding, or toileting skills. Hence, it was not surprising to find that very few people showed improvement in skills in the study period.

Characteristics of Severely Disabled People Followed Up for 11 Years*

	Subjects Available for Follow-up (n=128 248)†	Subjects Unavailable for Follow-up (n=27 603)†	Subgroups§					
			1 (n=985)	2 (n=382)	3 (n=1376)	4 (n=1699)	5 (n=2324)	6 (n=460)
Sex								
Male	56.1	56.6	53.5	50.1	52.0	52.2	53.6	52.8
Female	43.9	43.1	46.5	49.9	48.0	47.8	46.4	47.2
Unknown	None	0.3	None	None	None	None	None	None
Mental retardation								
Mild	34.0	31.1	None	None	None	None	None	None
Moderate	23.3	14.9	None	None	None	None	None	None
Severe	12.8	7.3	6.4	9.2	10.0	11.6	16.5	15.4
Profound	11.9	5.7	63.6	35.9	46.4	31.1	42.4	52.8
Suspected	18.0	41.0	30.0	54.9	43.6	57.3	41.1	31.8
Ethnicity								
Mixed	3.3	4.0	4.7	7.1	4.8	5.5	3.5	4.8
Asian	3.6	3.6	5.3	5.8	4.6	4.0	3.7	3.3
Black	10.4	12.0	8.9	8.9	8.3	8.6	8.6	9.6
Other	3.4	4.9	3.8	2.9	3.5	3.4	4.0	3.0
Hispanic	21.0	24.1	27.8	30.5	27.8	32.4	30.9	24.1
White	57.2	51.3	48.4	43.8	49.2	44.6	47.9	53.7
Unknown	1.1	0.1	1.1	1.0	1.8	1.5	1.4	1.5
Age, y								
0–4	26.6	35.5	64.6	80.1	61.0	71.7	63.6	62.8
5–9	10.0	13.2	10.7	6.3	9.4	6.5	8.7	9.3
10–14	9.7	9.4	10.6	3.9	6.7	5.0	6.4	8.7
15-19	13.7	12.2	8.1	5.0	8.1	5.4	6.6	7.4
20-24	13.2	10.4	2.6	1.3	6.2	3.6	4.7	5.7
25–29	8.7	6.3	1.8	1.0	4.1	3.7	4.0	3.3
30–34	6.2	4.4	0.8	1.6	2.5	1.6	3.1	1.3
35–39	3.8	2.7	0.3	0.8	1.0	1.5	1.3	0.7
40–44	2.4	1.7	0.4	None	0.7	0.6	1.0	0.7
45–49	1.8	1.2	0.1	None	0.3	0.4	0.6	0.1
50–54	1.4	1.0	None	None	None	None	None	None
55–59	1.1	0.7	None	None	None	None	None	None
60–64	0.7	0.5	None	None	None	None	None	None
65–69	0.4	0.3	None	None	None	None	None	None
≥70	0.3	0.4	None	None	None	None	None	None
Unknown	None	0.1	None	None	None	None	None	None
Seizure frequency								
No seizures	58.6	51.6	27.6	22.6	30.6	27.8	33.7	35.5
<1 per day	9.1	7.5	28.6	17.8	20.5	16.8	22.1	27.7
≥1 per day	2.8	2.3	13.1	17.6	14.8	11.5	10.5	12.8
Undetermined	29.5	38.6	30.7	42.0	34.1	43.9	33.7	24.0
Level of cerebral palsy								
None	56.1	49.4	18.2	16.5	19.8	16.2	15.1	16.2
Mild	4.4	4.1	0.9	1.1	1.2	1.9	2.7	0.7
Moderate	6.9	6.4	6.5	9.7	7.1	8.7	13.6	10.0
Severe	7.2	5.6	52.4	37.8	42.8	35.5	44.0	54.3
Suspected	2.4	1.9	6.6	9.7	6.9	4.5	4.4	5.6
Unknown	23.0	32.6	15.4	25.2	22.2	33.2	20.2	13.2
Type of residence								
Own home	68.5	78.3	40.4	62.5	62.8	74.8	71.2	60.2
Small group-home	9.1	6.1	3.9	6.3	5.6	4.2	5.8	5.4
Large group-home	6.3	4.4	1.5	1.6	2.0	1.2	1.2	1.5
Health facility	2.3	1.3	2.0	2.1	6.8	6.4	6.1	4.6
Skilled nursing facility	1.2	0.9	7.3	5.2	3.8	2.4	2.0	4.6
State hospital	8.2	3.3	34.4	13.9	16.0	8.9	13.0	18.8
Private hospital	0.5	0.5	9.9	7.3	2.3	0.9	0.4	4.1
Other	3.9	5.2	0.6	1.1	0.7	1.2	0.3	0.8
Serious major medical condition								
No	90.0	87.9	49.6	64.3	81.7	87.4	87.1	65.4
Yes	9.6	4.7	45.6	29.9	15.0	10.1	12.0	33.9
Unknown	0.4	7.4	4.8	5.8	3.3	2.5	0.9	0.7

(Continued)

Characteristics of Severely Disabled People Followed Up for 11 Years* (cont)								
	Subjects Available for Follow-up (n=128 248)†	Subjects Unavailable for Follow-up (n=27 603)†	Subgroups§					
			1 (n=985)	2 (n=382)	3 (n=1376)	4 (n=1699)	5 (n=2324)	6 (n=460)
Ambulation								
Cannot walk	22.7	19.4	100.0	100.0	100.0	100.0	100.0	100.0
Partial ability	10.3	10.6	None	None	None	None	None	None
Walks alone	66.9	69.9	None	None	None	None	None	None
Unknown	0.1	0.1	None	None	None	None	None	None
Crawling/standing								
Cannot crawl	16.0	12.5	100.0	100.0	100.0	100.0	100.0	100.0
Crawls	4.9	4.7	None	None	None	None	None	None
Stands	79.1	82.8	None	None	None	None	None	None
Rolling/sitting								
Cannot roll over	10.6	8.1	100.0	100.0	100.0	100.0	None	None
Rolls over	5.8	4.8	None	None	None	None	75.2	82.4
Sits	83.6	87.0	None	None	None	None	24.8	17.6
Unknown	None	0.1	None	None	None	None	None	None
Toileting								
Not trained	29.3	30.1	100.0	100.0	100.0	100.0	100.0	100.0
Partially trained	18.2	16.4	None	None	None	None	None	None
Trained	52.4	53.4	None	None	None	None	None	None
Unknown	0.1	0.1	None	None	None	None	None	None
Arm use								
No functional use	5.2	4.1	100.0	7.4	100.0	3.9	2.2	20.8
Partial use	11.3	9.1	None	66.1	None	66.3	51.9	48.0
Full extension	83.4	86.7	None	26.5	None	29.8	45.9	31.5
Unknown	0.1	0.1	None	None	None	None	None	None
Hand use								
No functional use	7.4	5.7	~~100.0~~	41.0	100.0	34.3	14.6	33.7
Partial use	21.7	18.9	None	58.0	None	63.7	79.8	63.0
Full use	70.8	75.3	None	1.0	None	2.0	5.6	3.3
Unknown	0.1	0.1	None	None	None	None	None	None
Receptive nonverbal communication								
None	20.5	16.5	96.8	88.5	91.1	85.4	75.3	77.0
Some	79.4	83.5	3.2	11.5	8.9	14.6	24.7	23.0
Unknown	0.1	None	None	None	None	None	None	None
Use of feeding tube								
Yes	2.0	1.4	100.0	100.0	None	None	None	100.0
No	98.0	98.6	None	None	100.0	100.0	100.0	None
No. of deaths	7974	None	543	174	581	457	302	124

*Values are percentages unless otherwise indicated. All subjects were served by the California Department of Developmental Services between 1980 and 1991. N=155 851.

†All people evaluated using the Client Development Evaluation Report from January 1980 through March 1991.

‡People who were unavailable for follow-up within 2 years of the start of the study.

§Subjects in all subgroups had severe, profound, or suspected mental retardation and were immobile and incontinent. Subgroup 1 was characterized by tube feeding, inability to roll, and no arm-hand use; subgroup 2, tube feeding, inability to roll, and some arm-hand use; subgroup 3, feeding by others, inability to roll, and no arm-hand use; subgroup 4, feeding by others, inability to roll, some arm-hand use; subgroup 5, feeding by others, ability to roll, and some arm-hand use; and subgroup 6, tube feeding and the ability to roll.

However, of 1055 people who were immobile and intermittently tube fed, 542 individuals were otherwise identical to subgroup 1, and 270 subjects were otherwise identical to subgroup 2. Survival functions (cumulative proportion surviving) for these groups are not presented because the estimates given were similar to those for subgroups 1 and 2. A sixth subgroup of 460 people received continuous (n=217) or intermittent (n=243) tube feeding and demonstrated immobility, some arm-hand use, and the ability to roll over. Because we were unable to select a sizable group of subjects who were continuously tube fed, had some arm-hand use, and could roll over, we added subgroup 6 to our study sample.

An exhaustive analysis of subjects using feeding status, ability to roll, and arm-hand use identified eight subgroups. Two subgroups were not studied because very few individuals met the criteria for these subgroups. In one subgroup, subjects were tube fed, were able to roll, and had no arm-hand use. In the other, subjects could take food if fed by others and could roll but had no arm-hand use. Most individuals who could roll over had some arm-hand use.

The Table also characterizes the six subgroups studied by variables other than those used to define these cohorts. Most subjects in all subgroups were very young children, and, except for subjects in subgroup 1, most lived in their own homes. Subgroup 1, which comprised the most medically fragile subjects, included subjects with more severe medical problems such as chronic upper respiratory tract infections and profound mental retardation. It was not unusual to find most of these people in out-of-home placements. More subjects in all subgroups had additional medical problems than did subjects in the unselected population being served. The cause of mental retardation was not studied variable because accurate diagnoses were often suspect[27] many individuals were listed as having retardation of unknown causes.

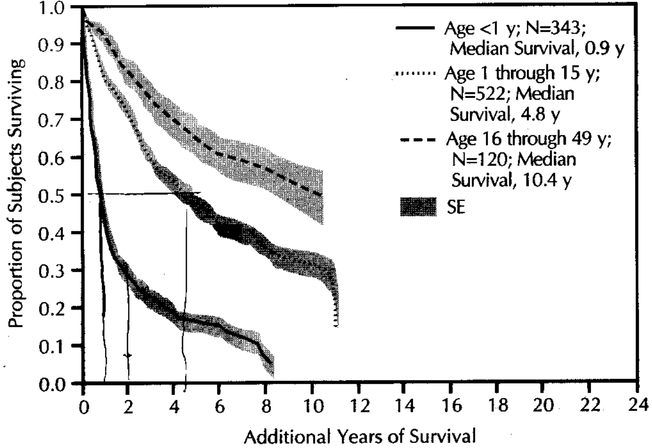

Fig 1.—*Survival of subjects in subgroup 1. Subjects were immobile, were tube fed, could not roll, were incontinent, had no arm-hand use, and had severe, profound, or suspected mental retardation.*

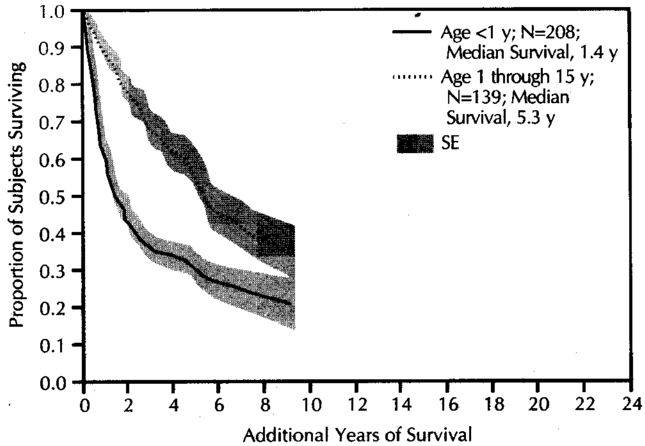

Fig 2.—*Survival of subjects in subgroup 2. Subjects were immobile, were tube fed, could not roll, were incontinent, had some arm-hand use, and had severe, profound, or suspected mental retardation.*

Statistical Analysis

The Kaplan-Meier product limit[31] method was used with the BMDP-1L statistical software package[29] to estimate the length of survival in an 11-year period for the five specific high-risk cohorts described earlier. The product limit survival functions estimate the probability that a subject will survive to a specified time. Hence, for each of the six cohorts described, a separate cumulative survival rate was computed by age groups younger than 1 year, 1 through 15 years, and 16 years and older in an 11-year period. These age groups were chosen based on homogeneity of the survival rates in the categories listed. For example, survival functions for 1- through 5-year-olds were almost identical to and not significantly different from ($P>.01$ by a Mantel-Cox statistic) those for 6- through 15-year-olds in any of the subgroups. Subjects older than 16 years were insufficient in number for comparison with other age groups. It should also be noted that some cumulative survival curves and hazard functions did not extend through the 11-year follow-up period. When the SEs of the estimated survival points or hazard rates became large because of subject attrition, we terminated further estimation.

In addition to providing cumulative survival rates, the BMDP-1L product limit estimated a hazard function that, in a discrete form, was a plot of death rates in the 11-year follow-up period. As Singer and Willett[32] noted, mathematical definitions of hazard differ depending on whether time is measured discretely or continuously. If time is measured discretely, as was necessary for this study, hazard is the conditional probability that an individual in a specific subgroup will die in a particular time interval (eg, in a 1-year interval) given that the person did not die by the beginning of the interval. The hazard function allows the reader to identify where mortality, as opposed to survival, was especially high or low during the follow-up period. In the interest of brevity, the hazard plots are not presented but are noted in instances where mortality was particularly high during a given year in the follow-up period. As Singer and Willett[32] stated, the hazard function effectively captures the distribution of risk across time.

Although the method of survival analysis used allowed for censoring (ie, discrimination between individuals unavailable to follow-up and individuals still alive at the end of the follow-up period), it was assumed that censoring occurred independent of subject characteristics in each survival or hazard analysis. For example, one type of subject should not be followed up more carefully than another, as differential censoring can occur in that instance.

RESULTS

It should again be noted that all individuals studied were considered to be profoundly or severely mentally retarded or were suspected to be mentally retarded. All cohorts lacked any toileting skills. Too few people with severe disabilities but only moderate or mild retardation were available for separate estimation of survival. Figure 1 illustrates survival rates for the most disabled subset of subjects, who were immobile, used a feeding tube, could not roll over, and had no arm-hand use. Children in this subgroup who were younger than 1 year had very poor survival rates. Their median survival was less than 1 additional year. About 5% of these children were still alive after 8 years. Children who lived to age 1 year or older had much higher survival rates, but survival was still limited (less than 11 more years). Children aged 1 through 15 years had a median survival of 4.8 more years, and about 20% of these children were alive after 11 years. After age 16 years, longevity was even better, inasmuch as the median survival was estimated to be 10.4 more years. For children younger than 1 year, very high mortality was observed shortly after birth, then declined rapidly until the third year of follow-up and began a sharp increase after the fifth year. The hazard rates for subjects in the other age groups revealed what seemed to be a constant overall trend accompanied by some random variation between individual years of follow-up.

Figure 2 traces the survival functions of a smaller group of individuals who used a feeding tube, were immobile, and could not roll but who had some arm-hand use. There is no survival curve for subjects aged 16 years or older because only 26 such people were included in our sample; SEs of the survival estimates for these 26 subjects were too high for meaningful interpretation. Children younger than 1 year had a median survival of 1.4 years. About 20% of these children were still alive after 11 years. For the subjects aged 1 through 15 years, the median survival was 5.3 more years, and about 35% of subjects in this group were still alive after 11 years. Mortality of the very young children was very high shortly after birth, steadily decreasing until the third year of follow-up, increased again at 9 years. For 1- through 15-year-olds, the hazard rates displayed some peaks and valleys in mortality during follow-up, but the general trend appeared relatively constant.

A number of severely disabled individuals who are immobile, cannot roll, and have no arm-hand use *can* be fed by mouth and do not require tube feeding. Figure 3 shows survival functions for this cohort. In general, survival was poor for children younger than 1 year at the beginning of

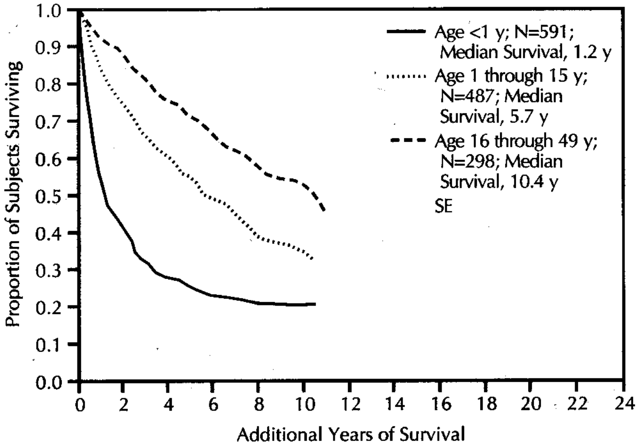

Fig 3.—*Survival of subjects in subgroup 3. Subjects were immobile, were fed by others, could not roll, were incontinent, had no arm-hand use, and had severe, profound, or suspected mental retardation.*

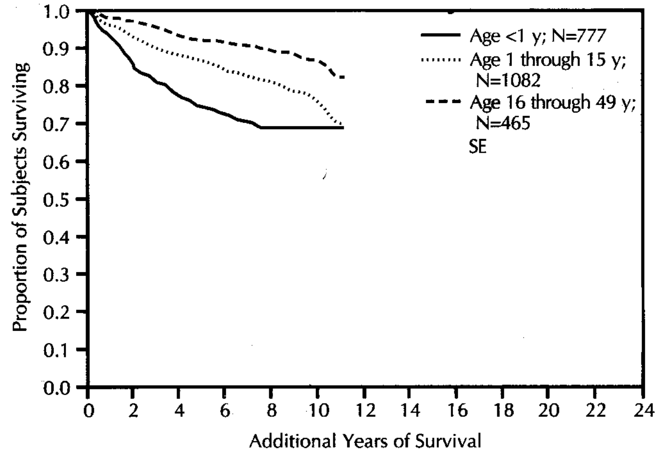

Fig 5.—*Survival of subjects in subgroup 5. Subjects were immobile, were fed by others, could roll, were incontinent, had some arm-hand use, and had severe, profound, or suspected mental retardation.*

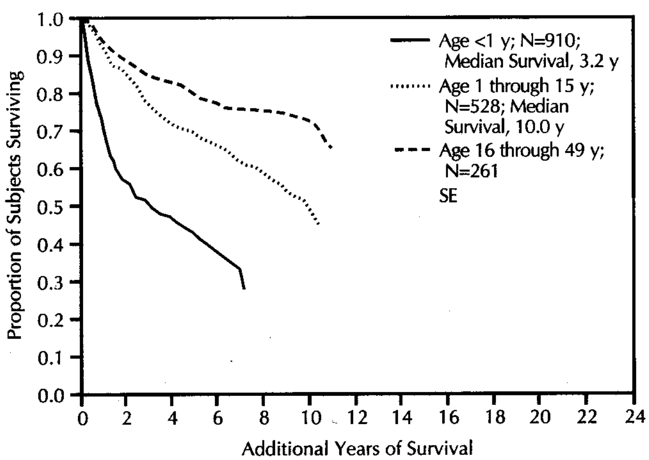

Fig 4.—*Survival of subjects in subgroup 4. Subjects were immobile, were fed by others, could not roll, were incontinent, had some arm-hand use, and had severe, profound, or suspected mental retardation.*

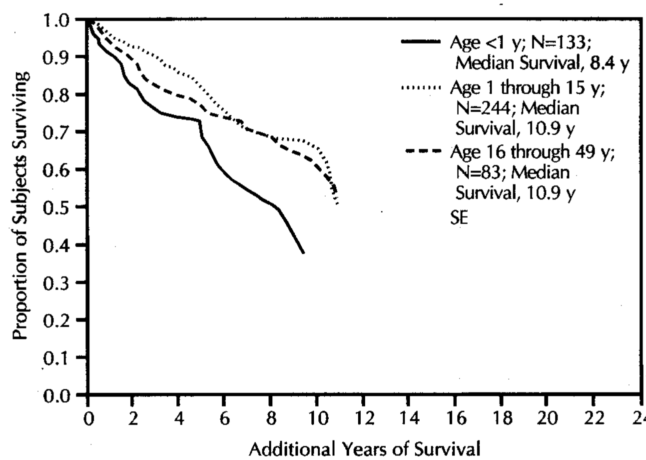

Fig 6.—*Survival of subjects in subgroup 6. Subjects were immobile, were tube fed, could roll, were incontinent, had some arm-hand use, and had severe, profound, or suspected mental retardation.*

he follow-up period. Approximately 20% were still alive fter 11 years. Survival of the 1- through 15-year-olds was so generally poor, with approximately 30% of subjects alive after 11 years. Subjects aged 16 to 49 years had a median survival of 10.4 years, and about 40% of these subjects lived longer than 11 years. Mortality in the follow-up period was relatively constant except for children younger than 1 year, whose mortality was initially high and decreased in the follow-up period. One reason for this decrease in mortality was the declining number of children at risk as a result of earlier high mortality.

In contrast with the last cohort of individuals who were completely immobile, were fed by others, and had no arm-hand use, there was a larger subgroup who, in the presence of immobility, had some voluntary use of their arms and hands, ie, who could extend an arm and/or use a thumb and fingers to grasp and move objects. Figure 4 displays survival functions by age groups for these people. While survival was still generally poor, it was discernibly better than survival for the subgroup who had no arm-hand use. For example, the median survival for subjects younger than 1 year was more than 3 years compared with 10 years for 1- through 15-year-olds. More than 60% of subjects in the oldest age group were still alive after 11 years. Mortality for subjects in the youngest age group was high for the first, second, and seventh years of follow-up. The mortality trends for the two older age groups were generally constant.

In addition to the other previously described subgroups of immobile subjects, there was a substantial number of disabled people who could voluntarily roll over, although they could not crawl, creep, or scoot from one place to another. All of these individuals were also found to have some arm-hand use. Figure 5 indicates that survival in this subgroup was comparatively good. None of the age groups evidenced a 50% cumulative death rate in the 11-year follow-up period. Rather, 70% of subjects in the highest risk group, children younger than 1 year, were still alive after 11 years. About 70% of the 1- through 15-year-olds were also alive after 11 years, and 80% of subjects in the oldest age group were alive after 11 years. Hence, the ability to roll over seemed to be associated with greatly improved survival. Mortality was relatively low for all follow-up years.

Figure 6 presents survival functions by age group for 460 individuals who intermittently or continuously r quired tube feeding but who could roll over. All o these people had some arm-hand use, but could not crawl. Survival was generally better for these subjects than for other subjects who could not roll, but was still much more limited than for subjects in subgroup 5, who were not tube fed but could roll over. Differences in survival by age group also were not as great for this subgroup as for the others, with median survival ranging between 8.4 and 10.9 years. The hazard rates for subgroup 6 appeared constant during the 11-year follow-up period except for peak mortality rates at 5 years' and 9 years' follow-up for the youngest age group.

COMMENT

We have estimated the survival of severely retarded people based on several combinations of profound disabilities. These estimates suggest a very shortened life expectancy for individuals unable to move their extremities or bodies voluntarily who need tube feeding.

Several studies[4-6,8,33] have shown the importance of mobility, feeding, and toileting skills to survival. Other studies[11,17] have focused on cerebral palsy. A recent study by Eyman et al,[7] found that immobile subjects had a much shorter life expectancy than those who could move. Those who also required tube feeding had a very short expectancy, ie, 4 to 5 additional years. Those who could eat if fed by others had a mean life expectancy of about 9 additional years. Finally, those who were mobile, but not ambulatory, had a life expectancy of about 23 more years.

The present study has refined the above results by focusing on the importance of rolling and arm-hand use. For individuals who could roll over, but were otherwise immobile (ie, cannot crawl), survival was relatively good (about 70% to 80% of such subjects were alive after 11 years). Subjects who could not roll over but had some arm-hand use still had a limited life expectancy (11 more years), although it was much longer than life expectancy for people who could not roll over and had no arm-hand use (6 more years). Survival in the latter group was very short, as reported in our previous study.[7] Immobile children who required tube feeding had the shortest survival (1 to 5 more years).

While it has been demonstrated previously that gross body mobility (crawling and walking) relates to life expectancy,[4,5,7,8,15,33-35] to our knowledge this is the first study to investigate the effects of the ability to roll and arm-hand use on survival. In association with other risk factors, these abilities provide a satisfactory means of calculating life expectancy for any person with mental retardation. Life expectancy was shortest for subgroup 1, which included those with the most profound level of mental retardation and serious medical conditions (Table), and was increasingly better for subgroups 2 through 5. Subgroup 6 resembled subgroup 4, except members of subgroup 6 were tube fed but could roll. Others have shown that prognosis is especially limited by lack of mobility in addition to severe central nervous system damage, which causes choking and shallow respiration[8]; seizure disorder[15]; feeding problems[7]; inability for toileting[33]; and cerebral palsy.[11,17]

With restriction of mobility, early death is mainly due to respiratory complications. Pulmonary ventilation is lessened, causing the high incidence of respiratory infection.[15] Inadequate cough and swallowing reflexes and shallow respiration add further to risk in nonambulatory patients.[8] Death is usually due to respiratory infection in individuals who are severely retarded and have associated disabilities,[3,4,13,36,37] who are immobile,[15] and who are in a persistent vegetative state.[35] Most patients with Down syndrome are reported to have died of respiratory infection.[4,19,38-44] In one institution, Blisard et al[2] reported that 72% of deaths of profoundly disabled residents were due to respiratory complications, especially pneumonia and aspiration with or without pneumonia.

Aspiration, indeed, produces a risk for pneumonia in these severely disabled persons. Gastrostomy feeding, even with fundoplication, does not totally prevent aspiration and pneumonia[45] and is a mortality risk,[46] although it reduces the likelihood of gastroesophageal reflux, esophagitis, and bleeding. Another entity predisposing to pneumonia is pulmonary edema, which promotes alveolar bacterial infection. Finally, premature aging in severely mentally disabled persons[47-49] relates to a depressed immune system evidenced by decreased humoral and especially T-cell function in antibody production.

Improved medical care has not altered significantly the poor prognosis for those with the most severe impairments.[3,9,12] A more constructive approach at this time is prevention: prevention of development of the underlying brain damage and prevention of complications, such as worsening of restricted mobility, by physical therapy.

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