Survival Rates Among Children With Severe Neurologic Disabilities

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ABSTRACT

Background. This study was done to determine survival rates in subpopulations of severely neurologically disabled children who reside in pediatric skilled nursing facilities and to compare these survival rates with those in previously published studies.

Methods. Data were collected at three pediatric skilled nursing facilities over the 1986 to 1996 decade. The total study population numbered 447. We studied in detail six groups of the most severely disabled children and correlated their survival rates with clinical parameters and the presence of other significant diseases.

Results. The survival rates in our six groups of severely disabled children were significantly better than those previously reported. In group 1, our 8-year survival rate was 66%, as compared with 5% in the previous study. In group 2, our 8-year survival rate was 89%, versus 22% in the previous study. We obtained better survival rates in all six groups studied, irrespective of the analysis including children less than 1 year old, between 1 year and 15 years old, or more than 15 years old. The most significant determinant for reduced survival was the presence of other significant diseases. Those with other significant diseases had a 10-year survival rate of 45%, whereas those who were relatively healthy had a survival rate of 90%. Patients who received gastrostomy tube feedings had a better 10-year survival rate than those fed by nasogastric tube (78% vs 41%). This difference was independent of the presence of other significant medical diseases.

Conclusions. Our results show substantially better survival rates than those previously reported. These improved results are most likely related to much more intense medical management of severely disabled children in skilled nursing facilities than at home or in other residential settings. Our study also showed a significantly better survival rate for those fed by gastrostomy tube as compared with nasogastric tube.

REDUCED SURVIVAL RATES among neurologically disabled children have been noted in a number of studies.[1-4] The most comprehensive and detailed analysis of physically disabled and mentally retarded individuals was done in California, where a total of 7,226 severely disabled children were studied.[1,2] This investigation revealed extremely short life expectancy results. The shortest survival occurred in those who were immobile, could not roll, had no arm or hand use, and were tube fed.

Allocation of resources to the care of disabled individuals is an important issue, particularly when private and public funds for long-term medical care are being restricted and reduced. Accurate longevity data are important in developing appropriate public policy and funding availability.

The data presented in the California study are extremely pessimistic and are not in keeping with our own clinical observations. We therefore decided to investigate our own experience in the care of children with severe neurologic handicaps at three pediatric skilled nursing facilities in the Chicago area. The focus of our study was to identify subgroups of severely disabled children who would be identical to the previously described groups[1,2] and to determine their survival rates. Since the California results are based on the largest population numbers, have the most clearly defined clinical groupings, and are cited with the most frequency,[1,2] we collected our data to match these studies exactly.

An additional focus was to identify factors that would affect survival rates. We paid particular attention to clinical parameters, the pres-

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TABLE 1. Clinical Grouping* of Patients With Cerebral Palsy, Profound Mental Retardation, Immobility, and Incontinence, but Without Progressive Neurologic Disease

Group 1	Group 4
Tube fed	Fed by others
Not rolling	Not rolling
No hand or arm use	Hand or arm use present
Group 2	Group 5
Tube fed	Fed by others
Not rolling	Able to roll
Hand or arm use present	Hand or arm use present
Group 3	Group 6
Fed by others	Tube fed
Not rolling	Able to roll
No hand or arm use	Hand or arm use present

*In accordance with Eyman et al.[1]

ence of other significant diseases, and the use of gastrostomy and nasogastric feeding tubes.

MATERIALS AND METHODS

Clinical information was gathered from three pediatric skilled nursing facilities for children with severe neurologic disabilities—Marklund Nursing Home, Little Angels Nursing Home, and Alden Village. The current number of residents in each facility is 100, 48, and 99, respectively. All three facilities are located in the Chicago metropolitan area. These facilities are not affiliated with each other, and each has its own independent complement of nursing staff. Two facilities are run by not-for-profit organizations and one by a for-profit organization. One of us (A.V.P.) examined all of the current residents and abstracted the clinical information. All of the medical records of discharged individuals over the previous 10 years (including all deaths) were thoroughly reviewed and the clinical information was abstracted (I.K., S.L., and D.M.). This 10-year period was chosen because the medical approach to the care of the children was consistent, since the same medical directors provided care during this time (A.V.P. has been the medical director of these facilities since 1990). Previously, much less intensive medical intervention had been used. For those individuals who had been discharged to another facility or home, the age and clinical status at the time of discharge were determined. The collection of the data was started in June 1995 and completed in June 1996. Thus, this study encompasses clinical outcomes of severely disabled children for the 1986 to 1996 decade.

In collecting all of the clinical data, particular attention was used to detail the clinical findings of each individual in accordance with previously published subgrouping definitions (Table 1).[1] We defined cerebral palsy as being a severe impairment of motor movements, the cause of which occurred before 28 days of age, in keeping with previous studies.[1,2] The vast majority of the cerebral palsy cases were due to prenatal or perinatal events. In addition, we abstracted clinical information concerning the underlying neurologic disease, presence of tracheostomy, toilet training abilities, and type of cerebral palsy (spastic quadriplegia, spastic diplegia, athetoid, ataxic, and hypotonic). The method of feeding each individual was determined: self-feeder, orally fed by others, nasogastric tube, and gastrostomy tube. The degree of mental retardation was determined from available psychologic examinations (profound mental retardation was defined as an IQ <20; severe mental retardation as an IQ between 20 and 35; moderate mental retardation as an IQ between 36 and 50). Illinois law mandates that all residents of a pediatric skilled nursing facility have a recent psychologic evaluation documenting the degree of mental impairment. The IQ determinations were done by many different psychologists using a wide battery of available testing instruments. Mobility parameters were assessed as being able to walk, crawl, creep, scoot, or unable to move at all (immobile). The ability to roll over was assessed as side-to-side, front-to-back, back-to-front, and unable to roll at all (an individual was considered to be able to roll if any one of the three rolling functions could be done). The presence of any hand or arm use was also assessed. Seizure type and frequency were determined and categorized as: <1 per month, between 1 per week and 1 per month, between 1 per day and 1 per week, and >1 per day. The presence of other significant medical diseases was determined. We defined other significant diseases as two or more bouts of pneumonia per year, recurrent bowel obstruction, refractory seizures (more than one seizure per day), severe asthma, cardiac arrythmia, and progressive cardiac failure. We separately identified individuals who had progressive neurologic diseases. We also ascertained the cause of death.

To compare survival rates in our study with those in the previously published study, we abstracted the data from published graphs.[1] In these graphs, 8-year survival rates can be determined. Thus, for comparison's sake, we determined our own 8-year survival rates. Also, the previously published survival rates were further subgrouped by age: (1) individuals less

TABLE 2. Neurologic Diagnoses by Percentage for the Entire Study Population (N = 447)

Prenatal encephalopathy, cause unknown	32.4%
Hypoxic-ischemic encephalopathy at birth	14.3%
Encephalopathy with hydrocephalus	8.7%
Encephalopathy with intraventricular hemorrhage after birth	6.5%
Congenital infection	5.7%
Head injury	5.4%
Central nervous system malformation	4.9%
Meningitis	4.3%
Encephalitis	2.5%
Down's syndrome	1.8%
Other chromosomal abnormalities	1.6%
Other congenital syndromes	2.5%
Progressive central nervous system degenerative diseases	3.6%
Other conditions	5.8%
Total	100%

than 1 year old, (2) individuals between 1 year and 15 years old, and (3) individuals more than 15 years old. We likewise subdivided our groups into these age categories to obtain comparable survival rate data.

For statistical analysis chi-square testing was used. Data are presented as the mean ± 1 standard deviation of the mean.

RESULTS

Epidemiology

The total study population consisted of 447 patients—215 from Marklund Nursing Home, 109 from Little Angels Nursing Home, and 123 from Alden Village. There were 224 female and 223 male patients, whose ages ranged from 0 to 42 years with a mean age of 13.6 ± 9.8 years. All three of the facilities are pediatric skilled nursing homes, but because of difficulties in finding placement for adults, a significant proportion are more than 21 years of age (113 of the total study group). All of these individuals had been admitted as children and grew up in these settings. By racial background, 59% were white, 25% were black, 13% were Hispanic, and 3% were Asian. The neurologic diagnoses are listed in Table 2.

Of the 447 patients, 371 (83%) had cerebral palsy (ie, neurologic event occurring before 28 days of age); of these 371, 308 (83%) had spastic quadriplegia, 45 (12%) had spastic diplegia, and 18 (5%) had hypotonia. Of the total population, 49 (11%) had a tracheostomy; 438 (98%) were incontinent; 416 (93%) had profound mental retardation, 22 (5%) had severe mental retardation, and 9 (2%) had moderate mental retardation; 233 (52%) were fed by gastrostomy tube, 40 (9%) by nasogastric tube, 152 (34%) were orally fed by others, and 22 (5%) were self-feeders; 201 (45%) displayed no hand or arm movement, 94 (21%) had nonpurposeful hand or arm movements, and 152 (34%) had purposeful hand or arm movements; 353 (79%) were

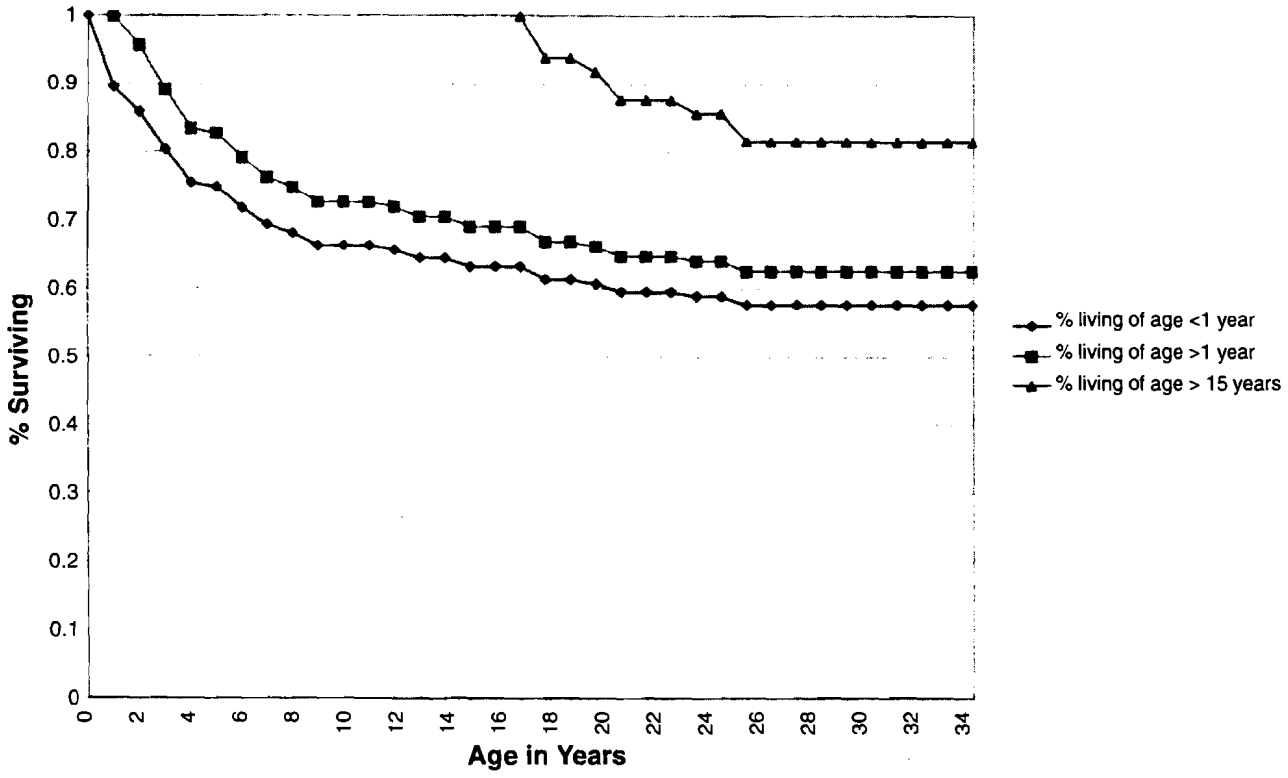

FIGURE 1. Survival in group 1.

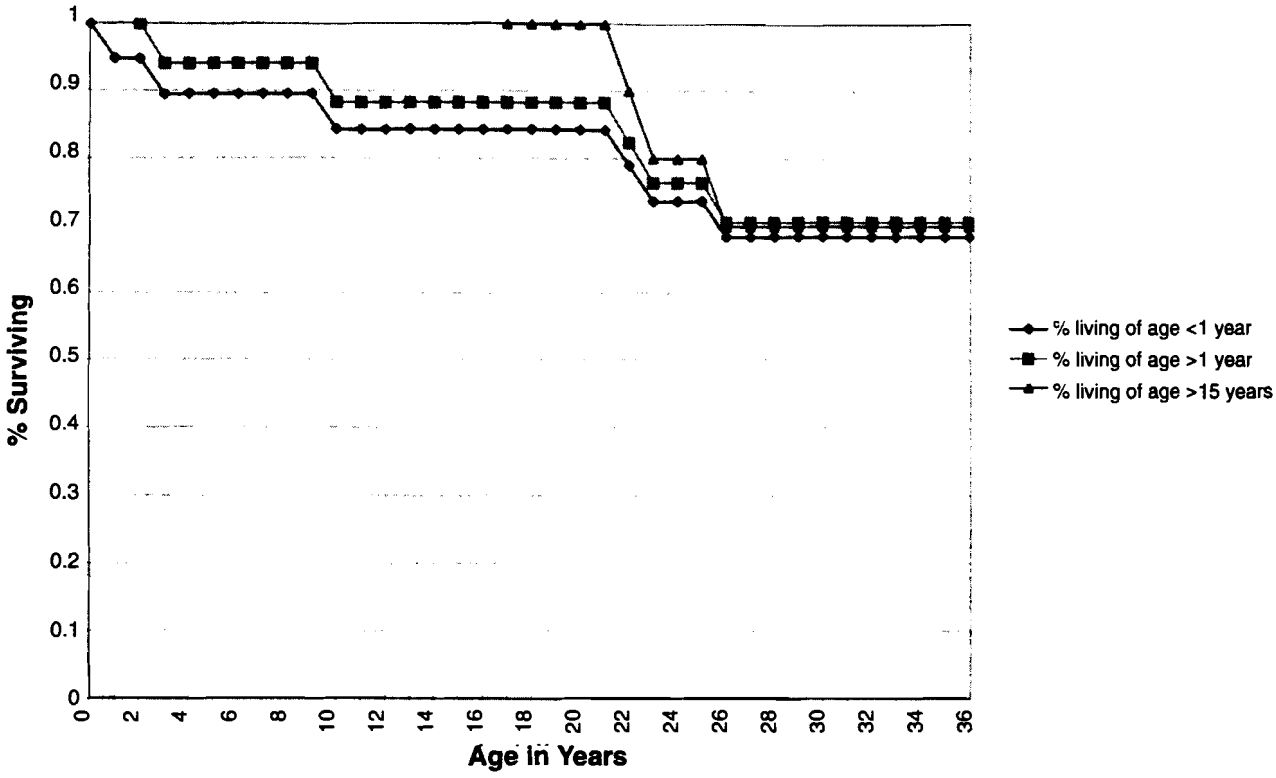

FIGURE 2. Survival in group 2.

immobile, 18 (4%) were able to walk, 31 (7%) were able to crawl, 54 (12%) could creep, and 94 (21%) could scoot on the stomach (individuals who were mobile were entered into more than one category); 308 (69%) were unable to roll, 130 (29%) could roll from side to side, 103 (23%) from front to back, and 107 (24%) from back to front (individuals who could roll were entered into more than one category of rolling); 161 (36%) did not have seizures, 179 (40%) had <1 seizure per month, 76 (17%) had between 1 seizure per week and 1 per month, 18 (4%) had between 1 seizure per day and 1 seizure per week, and 13 (3%) had >1 seizure per day. Of those with seizures, 277 (97%) had generalized seizures, and 9 (3%) had focal seizures; 283 (99%) of patients with seizures were treated with anticonvulsants.

Sixteen individuals (3.6%) had progressive neurologic diseases: 3 had adrenoleukodystrophy, 3 had Rett syndrome, 1 had an unknown degenerative disease, and 1 each had neuronal ceroid lipofuscinosis, Hurler's syndrome, Tay-Sachs disease, Canavan's disease, medium-chain acyl-coA dehydrogenase deficiency, Krabbe's disease, mucolipidosis, cytochrome oxidase deficiency, and Werdnig-Hoffmann syndrome. Half of these patients died during the study.

Of the study population, 121 (27%) died. Causes of death were pneumonia (93), cardiac arrest (15), progressive central nervous system degeneration (7), progressive heart failure (5), and bowel obstruction (1). Death rates during each year from 1985 through 1996 did not differ significantly. The percentages of deaths that occurred from 1985 through 1996 were, in sequence by year (starting with 1985): 0.9%, 2.2%, 2.9%, 1.8%, 2.7%, 2.5%, 3.1%, 2.0%, 2.5%, 2.2%, 3.6%, and 0.7% (for 1996 only deaths through June 1996 are included). Among the patients who died, there was no significant difference in sex or racial distribution. The mean age of those who died was 7.68 ± 8.81 years (age range, 0 to 35 years).

Survival Rates

When the study population was limited to those individuals who had cerebral palsy, profound mental retardation, immobility, incontinence, and no progressive central nervous system degenerative disease, the number of study cases was reduced to 251. This population of 251 was subdivided into clinical groups 1 through 6 (Table 1). These groups correspond to groups 1 through 6 of Eyman et al:[1] group 1, 163 patients (mean age, 10.6 years); group 2, 19 patients (mean age, 15.7 years); group 3, 47 patients (mean age, 15.2 years);

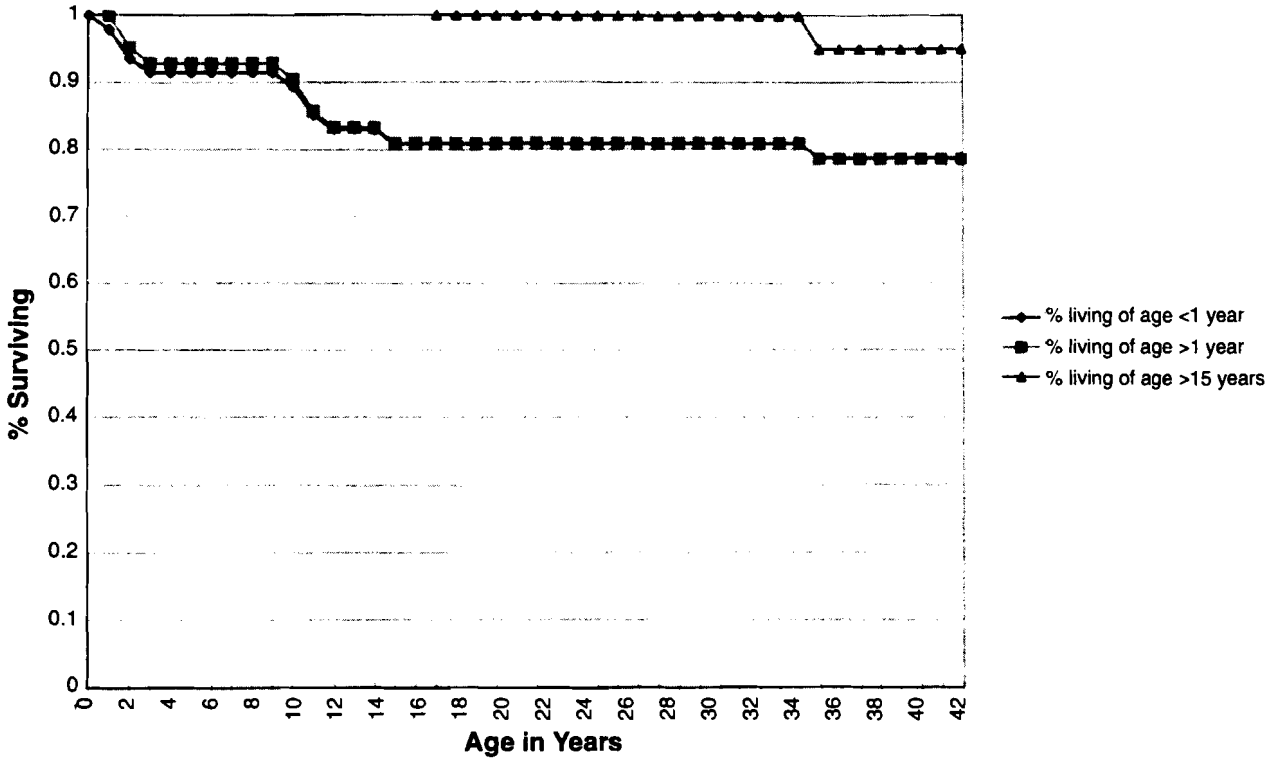

FIGURE 3. Survival in group 3.

group 4, 11 patients (mean age, 18.9 years); group 5, 9 patients (mean age, 18.8 years); and group 6, 2 patients (mean age, 21 years).

Survival rates in the first four groups are illustrated in Figures 1 through 4. There were no deaths in groups 5 and 6 during the study period. Figures 1 through 4 illustrate the study group starting from birth, along with separate graphs for those who had survived until the first and 15th birthdays. This subgrouping is identical to that used by Eyman et al,[1] so that these results could be directly compared with the previously published data. The 10-year survival rates for groups 1 through 6, respectively, were 66%, 84%, 87%, 91%, 100%, and 100%. It is clear that the most disabled group (group 1) had the lowest survival rate. The difference in 10-year survival between group 1 and group 3 was statistically significant ($P < .01$).

Tables 3, 4, and 5 compare 8-year survival rates for each group in our study and the previously published study.[1] The 8-year survival rate of the entire study group starting with patients less than 1 year of age is analyzed and compared in Table 3. In groups 1 through 4, the survival rate in our study is significantly higher than that in the previous study.[1] The most dramatic difference was in the most disabled group (group 1), in which the previous report showed a 5% survival rate compared with our rate of 66%. Likewise, for the next most severely disabled groups (groups 2 and 3), the previous report indicates survival rates of 22% and 21%, whereas we obtained rates of 89% and 92%. Even for group 4, we obtained a survival rate of 91%, versus 30% in the comparison study.[1]

In comparing the 8-year survival rates of the groups that are limited to those who have survived between 1 year and 15 years of age (Table 4) and those who have survived beyond 15 years of age (Table 5), we again found that

TABLE 3. Eight-Year Survival Rate Among Patients Less Than 1 Year of Age

Group	*Previous Study*[1]	*Current Study*	P *Value*
1	5%	66%	< .001
2	22%	89%	< .001
3	21%	92%	< .001
4	30%	91%	< .001
5	70%	100%	NS
6	50%	100%	NS

TABLE 4. Eight-Year Survival Rate for Patients Between 1 Year and 15 Years of Age

Group	*Previous Study*[1]	*Current Study*	P *Value*
1	35%	73%	< .001
2	38%	95%	< .001
3	40%	94%	< .001
4	60%	91%	NS
5	80%	100%	NS
6	70%	100%	NS

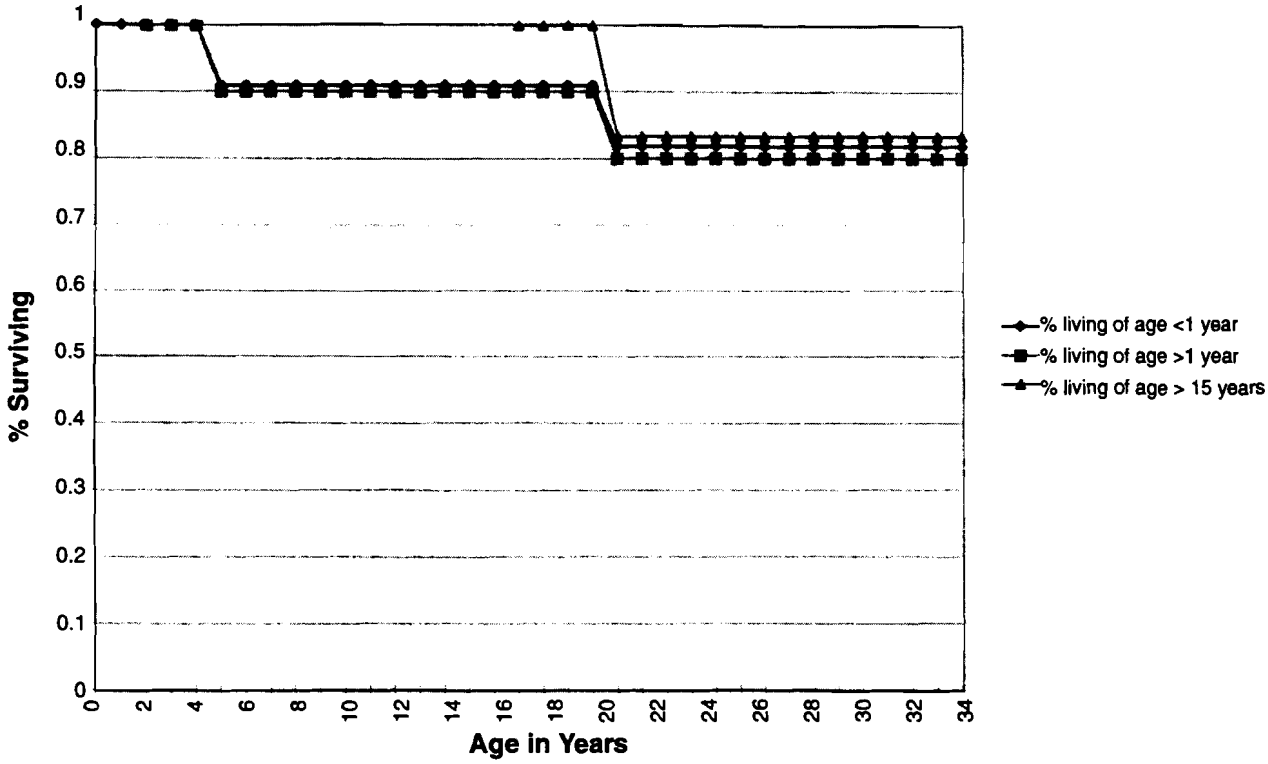

FIGURE 4. Survival in group 4.

our survival rates in groups 1 through 3 for these age subgroups were significantly better than those reported by Eyman et al.[1]

Factors Affecting Survival

To analyze factors that may affect survival rates, a different subset of the entire study group was investigated. When the total group was limited to individuals who had cerebral palsy and did not have a progressive neurologic disease, the study number was 367. This was done to increase statistical power.

Of this study population, 66 (18%) had other significant diseases. Of these, 44 (66%) had recurrent pneumonia, 12 (18%) had refractory seizures, 5 (7%) had cardiac failure, 3 (5%) had recurrent bowel obstruction, and 2 (4%) had severe asthma. Figure 5 shows the survival rates among patients who did not have other significant diseases and those who did. There is a significant difference in survival rates, the healthier group of individuals having a 10-year survival rate of 90% as compared with 45% in the group of ill individuals ($P < .0001$). There was no significant difference between those with and those without other significant diseases regarding sex, race, type of cerebral palsy, toileting abilities, and degree of mental retardation. There was a significant association between the incidence of other medical conditions and the presence of a tracheostomy ($P < .005$), presence of seizures and seizure frequency ($P < .002$), absence of hand or arm movement ($P < .0001$), inability to roll ($P < .0001$), and immobility ($P < .0001$).

There was a significant association between the presence and severity of epilepsy and survival rates (Fig 6). Those who did not have seizures had a 10-year survival rate of 87%, those with <1 seizure per month and between 1 seizure per week and 1 per month had rates of 83%, those with between 1 seizure per day and 1 per week had a survival rate of 68%, and those with >1 seizure per day had a 10-year survival rate of 33% ($P < .001$).

We found no correlation between survival rate and presence of a tracheostomy tube (Fig 7). The 10-year survival rate was 83% for those with no tracheostomy and 75% for those with a tracheostomy (P = NS).

TABLE 5. Eight-Year Survival Rate for Patients More than 15 Years of Age

Group	*Previous Study*[1]	*Current Study*	P *Value*
1	58%	86%	< .001
2	No data	79%	No data
3	60%	100%	< .001
4	76%	82%	NS
5	92%	100%	NS
6	70%	100%	NS

Survival rates according to feeding methods are presented in Figure 8. The 10-year survival rate was 95% for self-feeders, 95% for those orally fed by others, 78% for those fed by gastrostomy tube, and 41% for those fed by nasogastric tube. When the orally fed and self-fed group was compared with the group fed by gastrostomy tube, the survival rate difference was statistically significant ($P < .001$). Likewise, when the orally fed and self-fed group was compared with the children fed by nasogastric tube, the survival rate difference was statistically significant ($P < .0001$). When the children fed by gastrostomy tube were compared with those who had nasogastric tube feedings, the difference was also statistically significant ($P < .001$). There was a significant association between the use of tube feeding and the presence of other significant diseases ($P < .0001$). Of those who were fed by gastrostomy tube, 25% had other significant diseases, whereas 44% of those fed by nasogastric tube and 4% of those fed by others had other significant diseases. None of the self-feeders had other significant diseases. There was no significant difference between patients fed by gastrostomy tube and those fed by nasogastric tube regarding the presence of a tracheostomy (19% vs 13%, respectively), immobility (92% vs 97%), and inability to roll (86% vs 90%).

Survival rates for gastrostomy and nasogastric tube fed individuals were separated according to the presence or absence of other significant diseases (Fig 9). The 10-year survival rate for those with no significant disease was 86% for those fed by gastrostomy tube (n = 140) and 56% for those fed by nasogastric tube (n = 18) ($P < .01$). Among patients who had other significant diseases, the 10-year survival rate was 49% for those fed by gastrostomy tube (n = 47) and 18% for those fed by nasogastric tube (n = 11) (P = NS).

In the total study group, survival rates for male and female patients were not significantly different.

There was an association between the degree of mental retardation and survival rate. Ten-year survival rates were 100% for those with moderate mental retardation, 95% for those with severe mental retardation, and 81% for those with profound mental retardation. However, this association was not statistically significant.

There were significant associations between degrees of mobility and survival rates. Those who were totally immobile had a 10-year survival rate of 76%, whereas any degree of mobility (walking, crawling, creeping, or scooting) gave a survival rate of 96%

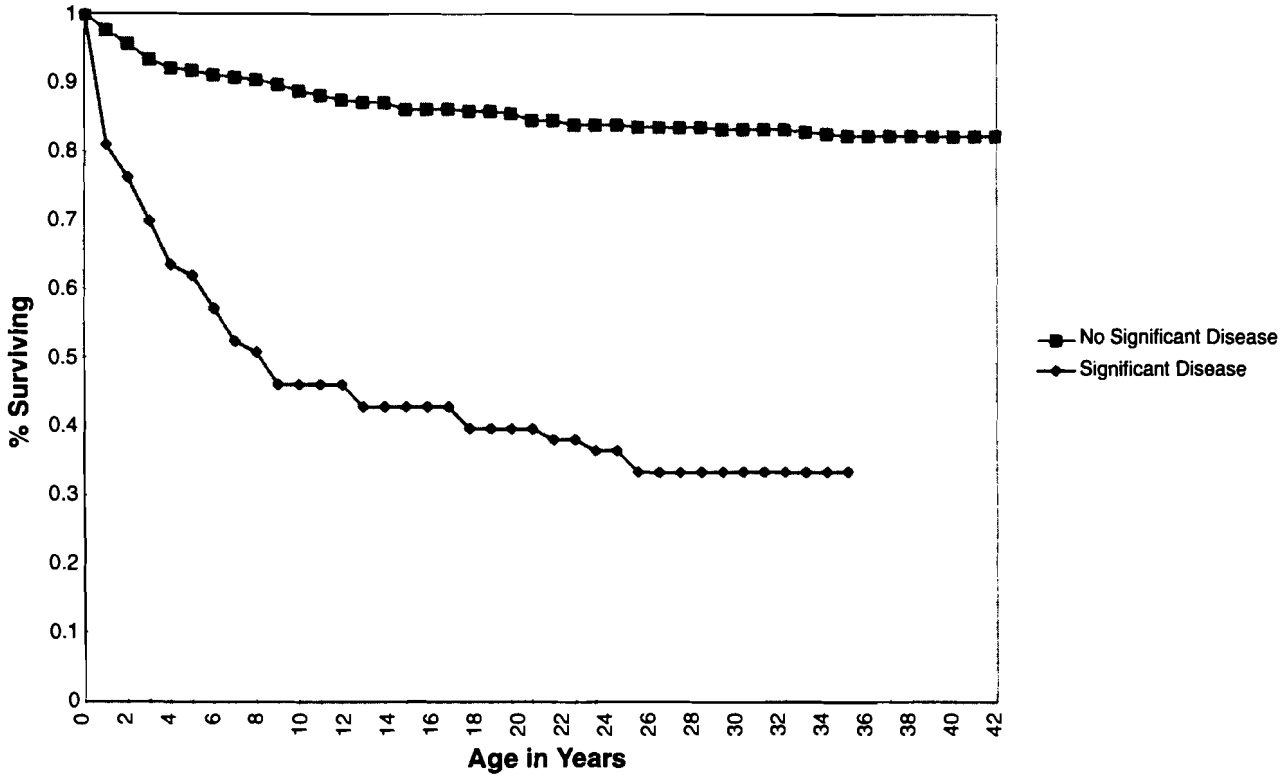

FIGURE 5. Comparison of survival by presence of other significant diseases.

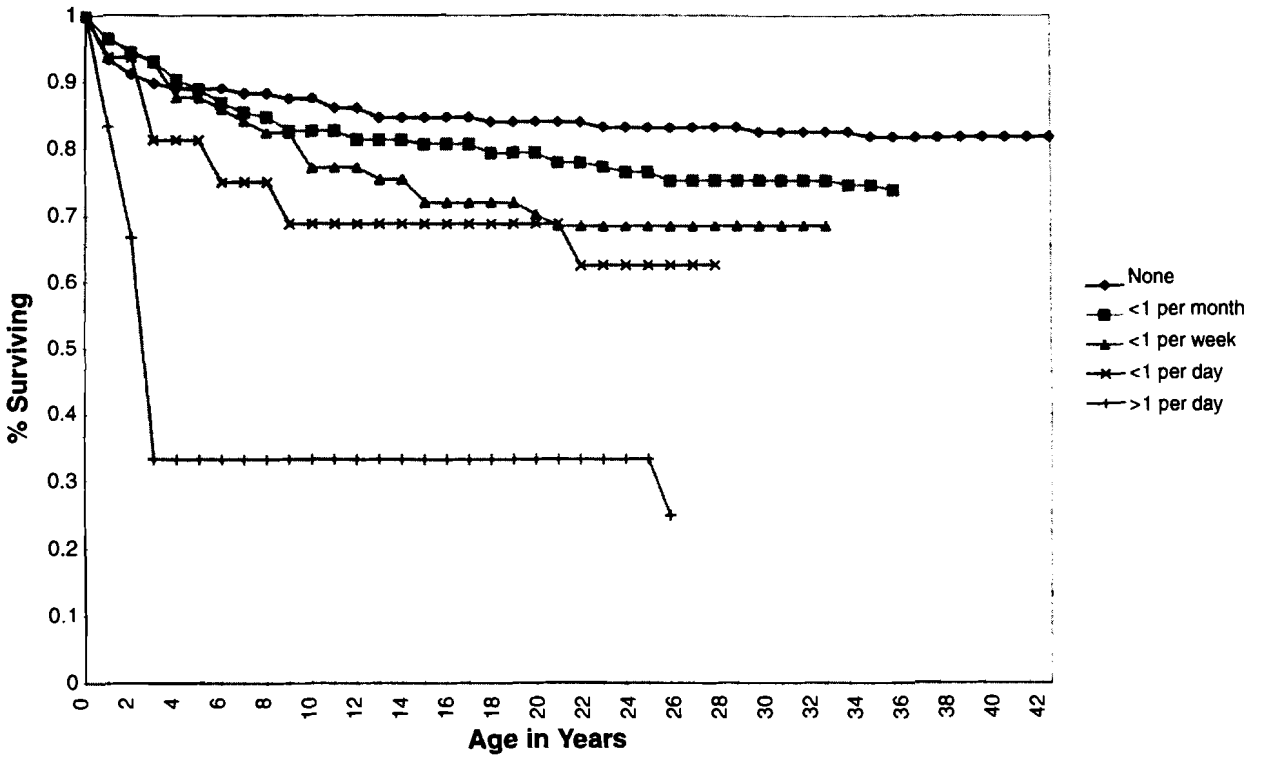

FIGURE 6. Comparison of survival by frequency of seizures.

($P < .001$). Any degree of ability to roll gave a 10-year survival rate of 98%, whereas inability to roll gave a survival rate of 74% ($P < .0001$).

DISCUSSION

The study population consisted of 447 individuals who were extremely neurologically disabled: 83% had cerebral palsy, 11% had a tracheostomy, 93% had profound mental retardation, 52% were fed by gastrostomy tube, 9% were fed by nasogastric tube, 45% had no hand or arm movements, 79% were immobile, and 64% had epilepsy.

As expected, there was no significant difference in survival rate by sex. Decreased survival rates were significantly associated with decreased mobility, inability to roll, presence and severity of epilepsy, and receiving tube feedings. These results are in keeping with previously published findings.[1-4] Two previous studies have reported decreased survival rates in children with more pronounced mental retardation.[3,4] We found a similar trend, but it was not statistically significant. There was no correlation between survival rate and the presence of a tracheostomy tube.

When the study population was limited to coincide with six groups that were previously defined,[1] we obtained survival rates that were significantly better. For example, for the most severely handicapped individuals (group 1), our 8-year survival rate was 66%, compared with 5% in the previous study.[1] For group 2, the rates were 89% and 22%, respectively. For group 3, the rates were 92% and 21%. In all of the six groups that were compared, irrespective of age groupings, we consistently obtained significantly better survival rates.

Our study was specifically designed to identify subgroups of disabled children that would be identical, and thus comparable, to those of Eyman et al.[1] Our population is comparable, but not identical, to the "severe" categories of ambulatory and manual dexterity of Hutton et al.[3] In their report, 10-year survival rates for these two categories were 70%, whereas our 10-year survival rate was 84%. In another study of quadriplegic children with cerebral palsy, the 10-year survival rate was 90%.[4] Although our study population would fit into this general category, that population was certainly much less disabled than ours. It is not possible to directly compare the results of these two studies with ours, since their clinical definitions are too broad. Nevertheless, these two studies report survival rates that are much closer to ours than to those of Eyman et al.[1,2]

The strongest predictor of survival was the presence of other significant diseases. In our study, the 10-year survival rate for healthier

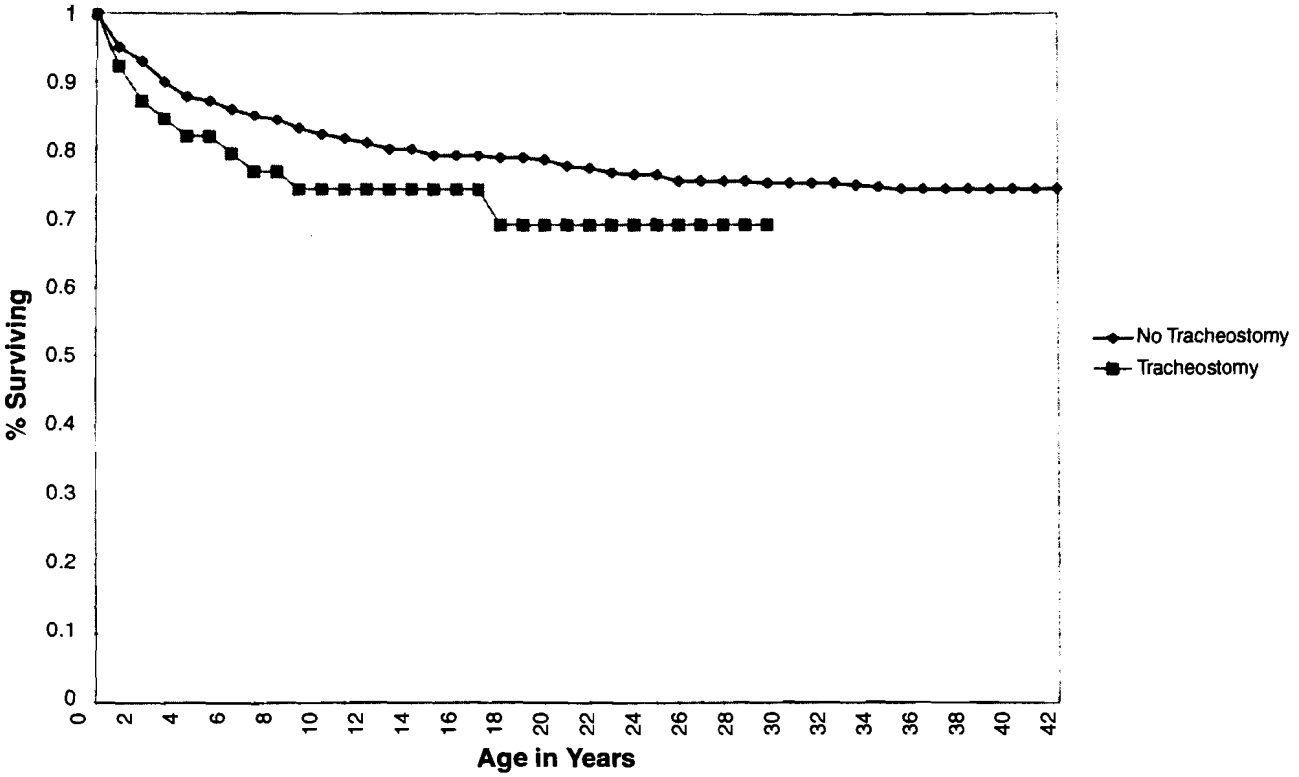

FIGURE 7. Comparison of survival by presence of tracheostomy.

individuals was 90%, but for those with other significant diseases it was 45%. In the previous studies,[1,2] stratification of survival rates according to other concomitant illnesses was not done. The presence of other significant diseases did correlate with the presence of a tracheostomy, seizure frequency, absence of hand and arm movements, inability to roll, and immobility. There was no correlation between the presence of other significant diseases and sex, race, type of cerebral palsy, toileting ability, or degree of mental retardation. Our study is the first one to show that survival of severely disabled children is significantly reduced by a defined set of other significant medical diseases. Conversely, severely disabled children who are otherwise not ill have reasonably good survival rates.

Of considerable importance is that there was a significant correlation between survival rate and feeding technique. Although the survival rate was significantly less in individuals fed by gastrostomy tube than in those fed orally, it was significantly better than in children fed by nasogastric tube. Improvement in outcome with gastrostomy tube placement, thus decreasing aspiration pnemonia and gastroesophageal reflux and bleeding, has been noted previously.[5-7] The incidence of tracheostomy tubes, immobility, and lack of rolling ability was not significantly different between patients fed by gastrostomy tube and those fed by nasogastric tube. The relatively poor outcome among those fed by nasogastric tube may have been due to the higher incidence of other significant diseases in this population. Individuals who are medically fragile may be considered to be too ill for general anesthesia and thus are fed by nasogastric tube. However, when our data were analyzed separately by the presence or absence of other significant diseases, children fed by gastrostomy tube still had a significantly greater survival rate than those fed by nasogastric tube. Thus, the presence of other significant diseases cannot account for our results. The best explanation is that the presence of a nasogastric tube promotes gastroesophageal reflux and vomiting, which can lead to aspiration and death. This is the first report comparing survival rates by gastrostomy tube versus nasograstric feeding.

Since tube feeding has been associated with increased mortality, Kastner et al[8] suggested that feeding tubes not be used. However, in that particular study, outcomes were not separated according to gastrostomy tube or nasogastric tube. Our data indicate that nasogastric

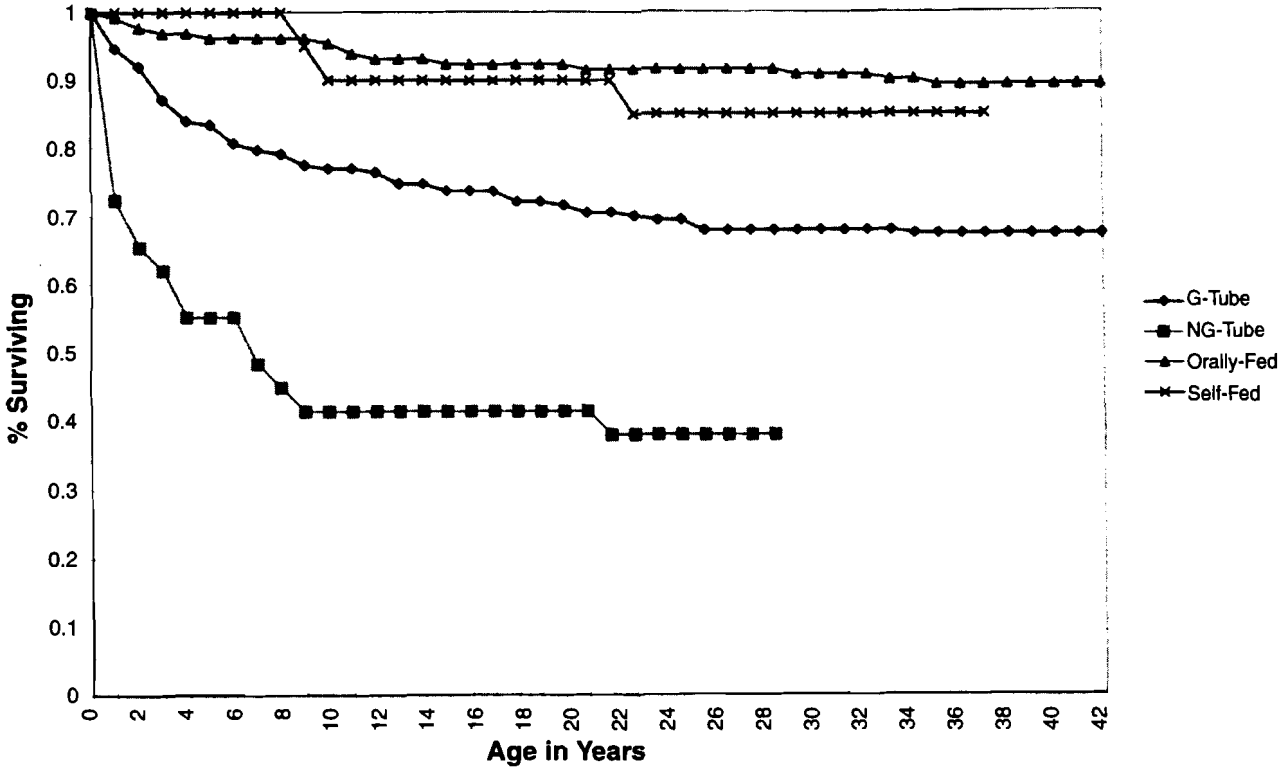

FIGURE 8. Comparison of survival by feeding method.

tubes are associated with a much higher mortality rate than gastrostomy tubes. Thus, when data incorporate both tube feeding approaches, poor survival rates would be expected. The presence of other significant diseases in individuals fed by gastrostomy tube significantly decreases survival rates. In our study, the 10-year survival rate for those with gastrostomy tubes and without other significant diseases is 86%, which is comparable to the 95% survival rate of orally fed individuals. A recent study has reported that in severely disabled children, the presence of a feeding tube actually did not increase mortality.[9] In those with a tracheostomy, tube feedings were associated with increased survival rates.[9] However, in this study, as in others, gastrostomy tube feedings were not distinguished from nasogastric tube use.

It is not possible to claim that our greater survival rates are related to the selective death of younger children who may not have lived long enough to enter a nursing facility. When we excluded children less than 1 year of age from the data analysis and studied those between 1 year and 15 years of age, we obtained significantly better survival rates than those of Eyman et al[1] (Table 4). Likewise, when we limited our study to those more than 15 years of age, we also obtained significantly better survival rates (Table 5). Thus, excluding the youngest and presumably the most ill children from the study populations did not diminish our improved survival rates.

There may be several reasons why our survival rates were better than those of Eyman et al.[1,2] The previously collected data covered the 1980 to 1990 decade, whereas our study was over the 1986 to 1996 decade. There have been many medical advances from 1980 to 1985, but more importantly there was a shift in the public and medical professional attitudes toward the care of disabled individuals. We purposefully limited our study to after 1985 because before that year the approach to the care of disabled children in Chicago was rudimentary in comparison to current standards. For example, at our nursing facilities, gastrostomy tubes and tracheostomies were almost nonexistent before 1985. Almost certainly, attitudes have similarly changed in California during these years. (In the final paragraph of their report, Eyman et al[1] stated that improvement in medical care had produced no effect on life expectancy outcomes in mentally retarded individuals. This conclusion was drawn not from their data but from three cited references that were published in

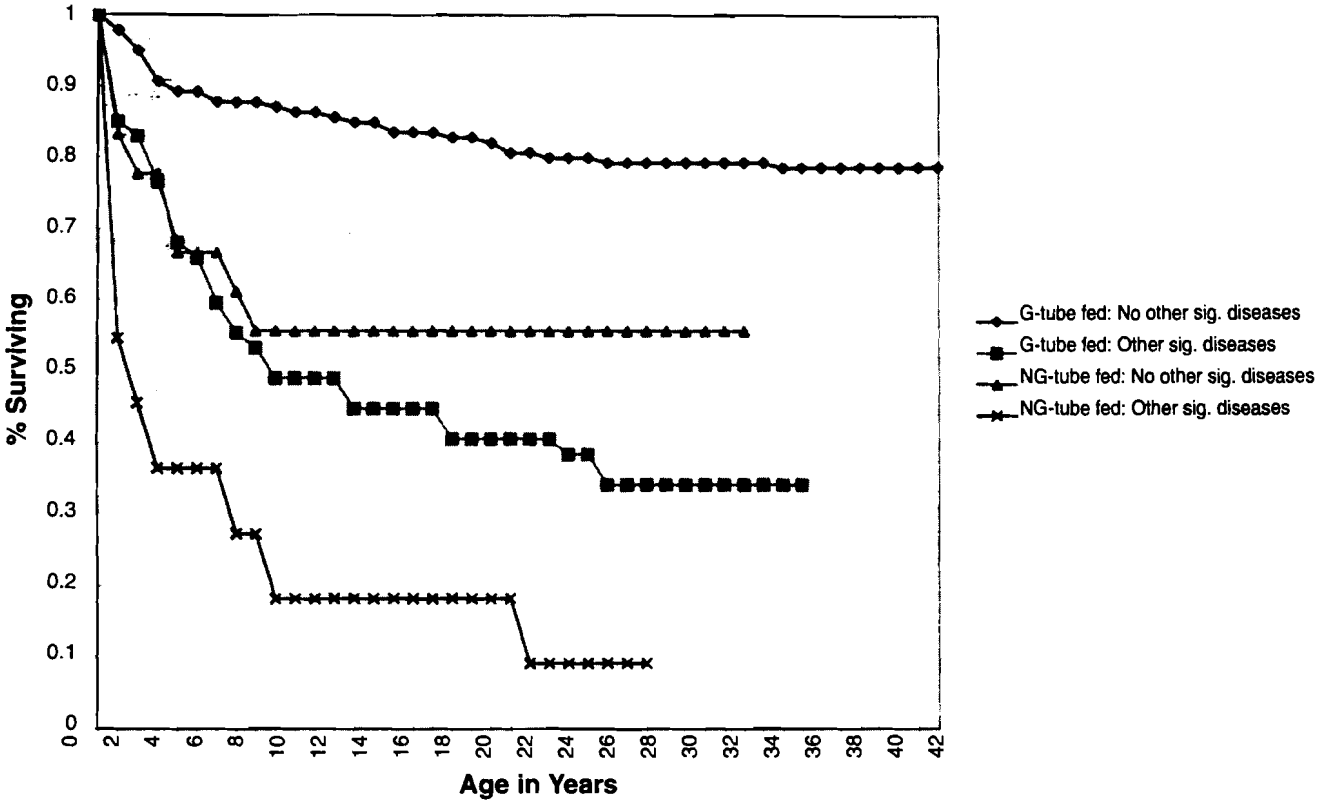

FIGURE 9. Comparison of survival by feeding method and other significant diseases.

1968, 1972, and 1983.) An analogous situation is the observation that survival rates have improved substantially in Down's syndrome, with 10-year survival rates of 37% for the 1945 to 1955 decade versus 86% between 1966 and 1975.[10] The primary reason for this improvement in survival is the surgical correction of congenital defects, particularly cardiac ones. Our study covers a period of much more intense medical management of severely disabled individuals, and this may account for better longevity outcomes.

Although we used the results from three different skilled nursing facilities, they all have been under uniform medical direction over the study period. In the study of Eyman et al,[1] data were collected from 21 regional centers located across California, and the medical care was provided by a large number of physicians and institutions. Such diversity would have produced discrepant quality of medical care. There is no suggestion in their data of any uniformity in medical care provided to the severely disabled.[1,2] Thus, uniformity of medical interventions in our study may be a contributing explanation for improved survival rates.

All of our study individuals were in a skilled nursing facility where registered nurses are available in-house, 24 hours a day, 7 days a week, to provide acute medical care. This intensity of medical service undoubtedly also contributed to better survival rates. Acute illnesses are addressed promptly. In the previous study,[1] 65% of the disabled individuals were residing at home, and only 3.5% were in a skilled nursing facility. It is possible that the poor survival rates, particularly for medically fragile individuals, were due to a lack of prompt medical intervention.

Recently, the California data have been reanalyzed according to place of residence.[11] Survival rates were comparable for residents at home and in group homes. However, when compared with survival rates in state institutions and other health care facilities, home and group home placement resulted in 25% greater mortality, irrespective of the degree of physical handicap. Residents of skilled nursing facilities were excluded from the data analysis. The authors concluded that the best explanation for their results was reduced availability of medical care, especially emergency care, in the home and group home setting. These new survival results,[11] in addition to our results, strongly suggest that the critical factor in determining survival rates in severely disabled children is the availability of acute medical care.

With progressive governmental and private payer budgetary restrictions and the pressure for deinstitutionalization of severely disabled children, our data and the findings from the recent California study are sobering. Placement of profoundly disabled children in environments where acute medical care is not readily available may result in a dramatic increase in mortality rates.

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