Survival Rates of Children With Severe Neurologic Disabilities: A Review

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Knowledge of accurate survival rates of children with neurologic disabilities is important for third-party insurance payers planning future medical expenses. This is of particular importance to pediatric skilled nursing facilities (SNFs) that depend on financial support from governmental sources. Eyman published survival rate results from California that were extremely pessimistic and not in keeping with our clinical impressions. This led us to conduct a thorough review of our survival rates, which were much better than those reported by Eyman. Since the publication of our study, a large number of reports have appeared from many different countries, as well as further information from California using an expanded database. The survival rate data that we obtained remain consistently better than that in most recent reports. In the California results, 10-year survival rates for the most-disabled group (group 1) were reported to be 32% in 1993 and 45% in 1998, compared with 73% in our study. Eight-year survival rates for group 1 from California were reported to be 38% in 1993 and 63% in 2000, compared with our finding of 73%. The reasons for our better survival rates include the fact that all of our patients were in SNFs, where prompt medical care for acute illnesses was always provided, whereas only 3.5% of the study group was in SNFs in California. Also, the California data contained many methodologic and statistical errors, which are reviewed here.

REDUCED SURVIVAL rates of neurologically disabled children have been noted in many studies.[1-13] The most comprehensive and detailed analysis of physically disabled children was performed in California, where a total of 7,226 individuals who received services from the State Department of Developmental Services were studied.[1,2] This investigation reported extremely short life expectancies, a finding at odds with our own clinical experience.

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.

We investigated our own experience with neurologically disabled children at three pediatric skilled nursng facilities (SNFs). The primary 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. An additional focus was to identify factors that would affect survival rates. We paid particular attention to clinical parameters, the presence of other significant diseases, and the use of gastrostomy and nasogastric feeding tubes. The full details of our investigation have been published.[3,14]

This article reviews our survival rate results and compares them with those reported by other investigators. Particular emphasis is placed on a comparison with more recent reports and a critique of the published California research results.

MATERIALS AND METHODS

This section gives a brief review of the methods used in our previously published study,[3] to which the reader is referred for more details. Clinical information was gathered from three pediatric SNFs for children with severe neurologic disabilities (as summarized in Table 1). The study was undertaken and started in 1996. The period of 1985-1996 was chosen because during this time the medical approach to the care of the children was consistent, with the same medical directors providing the care. Previously, much less intensive medical intervention had been used.

In collecting all of the clinical data, particular care was taken to record in detail the clinical findings of each individual in accordance with previously published group definitions (Table 2). Cerebral palsy was defined as a severe impairment of motor movements, the cause of which occurred before 28 days of age. We abstracted clinical information concerning the underlying neurologic disease, presence of tracheostomy, toilet-training

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Table 1. Neurologic Diagnoses by Percentage for the Entire Study Population (n = 447)

Prenatal (congenital) encephalopathy	32.4%
Hypoxic-ischemic encephalopathy at birth	14.3%
Encephalopathy with hydrocephalus	8.7%
Encephalopathy with IVH 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 syndrome	1.8%
Other chromosomal abnormalities	1.6%
Other syndromes	2.5%
Progressive CNS degenerative diseases	3.6%
Other conditions	5.8%
Total	100%

abilities, and type of cerebral palsy. The method of feeding was determined: self-feeding, oral feeding by others, nasogastric tube feeding, and gastrostomy tube (G tube) feeding. The degree of mental retardation was determined from available psychological examinations. Mobility parameters were assessed in terms of ability to walk, to crawl, to creep, to scoot, or not able to move at all (immobile). The ability to roll over was assessed as side-to-side, front-to-back, back-to-front, and not able to roll at all. The ability to use the hands and arms was also assessed. Seizure type and frequency were determined and categorized as fewer than one per month, between one per week and one per month, between one per day and one per week, and more than one per day.

The presence of other significant medical diseases was determined. Other significant diseases were defined as two or more bouts of pneumonia per year, recurrent bowel obstructions, refractory seizures (more than one seizure per day), severe asthma, cardiac arrhythmia, and progressive cardiac failure. Individuals who had progressive neurologic diseases were separately identified, and causes of death were ascertained.

To compare survival rates between our study and previously published results, we abstracted data from published graphs.[2] In these graphs, 8-year survival rates can be determined. Thus, for comparison's sake, we determined our own 8-year survival rates. Also, we further subgrouped the previously published survival rates by individuals under age 1 year, individuals between age 1 and 15 years, and individuals older than 15. We likewise subdivided our groups into these age categories to obtain comparable survival rate data. For statistical analysis, we generated and plotted Kaplan-Meier survival functions, and used the χ^2 test to determine significance.

RESULTS

The total study population consisted of 447 cases. The age range was from 0 to 42 years, with a mean age of 13.6 ± 9.8 years. The neurologic diagnoses of the study population are listed in Table 1. Of this total population, 83% had cerebral palsy, 11% had a tracheostomy, 98% were incontinent, and 93% had profound mental retardation. In terms of feeding, 52% were G tube feeders, 9% were nasogastric tube feeders, 34% were orally fed by others, and 5% were self-feeders. In terms of mobility, 79% were immobile. In terms of seizure activity, 40% had fewer than one seizure per month, 17% had between one per week and one per month, 4% had between one per day and one per week, and 3% had more than one per day. Of those with seizures, 97% experienced generalized seizures and 3% experienced focal seizures.

Of the study population, 27% died during the study period. Of those who died, 77% died from pneumonia, 12% from cardiac arrest, 6% from progressive central nervous system degeneration, 4% from progressive heart failure, and 1% from bowel obstruction. There was no significant difference in the rate of death during each year from 1985 through 1996.

Clinical groups 1 through 6 were defined to

Table 2. Clinical Definition of Groups in Accordance With Eyman[2]

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

NOTE. In all cases the individuals had cerebral palsy and did not have a progressive neurologic disease process. In all cases the individuals had profound mental retardation, were immobile and were incontinent.

Table 3. Eight-Year Survival Rate for Cases Under 1 Year of Age

Group number	Eyman[2]	Plioplys[3]	*P* value
1	5%	66%	<0.001
2	22%	89%	<0.001
3	21%	92%	<0.001
4	30%	91%	<0.001
5	70%	100%	NS
6	50%	100%	NS

Abbreviation: NS, not significant.

correspond exactly to the groupings of Eyman[2] (Table 2). Survival rates for each group are compared to previously published results in Tables 3, 4, and 5. Table 3 compares 8-year survival rates for all of the age groups. Our study found a significantly better survival rate than the previously published results for groups 1 through 4. The most dramatic difference was seen in the most severely disabled group (group 1), in which the previous report showed a 5% survival rate and we obtained a rate of 66%.

Tables 4 and 5 compare the 8-year survival rates limited to those who have survived to age 1 year (Table 4) and to age 15 years (Table 5). Again, we obtained significantly better survival rates than those reported by Eyman.[2]

Of the study population, 18% had other significant diseases. Of these, 66% had recurrent pneumonias, 18% had refractory seizures, 7% had cardiac failure, 5% had recurrent bowel obstruction, and 4% had severe asthma. There was a significant difference in survival rates, with the healthier group of individuals having a 10-year survival rate of 90% and the group of ill individuals having a rate of 45% (P <0.0001).

There was no significant difference in survival rate based on the presence of a tracheostomy tube. The 10-year survival rate was 85% for those with no tracheostomy and 75% for those with a tracheostomy (P = ns).

Table 4. Eight-Year Survival Rate for Cases Between 1 and 15 Years of Age

Group number	Eyman[2]	Plioplys[3]	*P* value
1	35%	73%	<0.001
2	38%	95%	<0.001
3	40%	94%	<0.001
4	60%	91%	NS
5	80%	100%	NS
6	70%	100%	NS

Abbreviation: NS, not significant.

Table 5. Eight-Year Survival Rate for Cases Older than 15 Years of Age

Group number	Eyman[2]	Plioplys[3]	*P* value
1	58%	86%	<0.001
2	No data	79%	no data
3	60%	100%	<0.001
4	76%	82%	NS
5	92%	100%	NS
6	70%	100%	NS

Abbreviation: NS, not significant.

The 10-year survival rate was 95% for self-feeders, 95% for those orally fed by others, 78%, for G tube feeders, and 41% for nasogastric tube feeders. When the orally fed and self-feeder groups were compared with the G tube–fed group, the survival rate difference was statistically significant (P <0.001). Likewise, when the orally fed and self-feeder groups were compared with the nasogastric tube–fed group, the survival rate difference also was statistically significant (P <0.0001). When the G tube–fed group was compared with the nasogastric tube–fed group, a statistically significant difference (P <0.001) in survival rate was also seen.

Survival rates for the G tube– and nasogastric tube–fed groups were separated according to the presence or absence of other significant diseases. The 10-year survival rate for those with no significant disease was 86% for G tube–fed children and 56% for nasogastric tube–fed ones (P <0.01). The 10-year rate for those who had other significant diseases was 49% for G tube–fed children and 18% for nasogastric tube–fed ones.

There were significant associations between degrees of mobility and survival rates. Any degree of immobility was associated with decreased survival rates.

DISCUSSION

We found a significant association between decreased survival rates with immobility and the use of tube feedings. These results are in keeping with previously published findings.[1,2,6-12,15] As would be expected, we found no difference in survival rate when groupings were made on the basis of sex. This finding is in keeping with previous reports that mentioned an analysis by sex.[8-10]

Previous studies have reported decreased sur-

vival rates in children with more pronounced mental retardation.[7,9-13] We found a similar trend, but determined that it was not statistically significant. We found an association with epilepsy, in that those children who experienced more frequent seizures had a much greater mortality rate than those who experienced less frequent seizures. Similar findings had been reported previously.[6-8,10,12]

We found no correlation between survival rates and the presence of a tracheostomy tube. The only other published report concerning the survival rate of children with tracheostomies[16] showed that in those who were tube fed, a tracheostomy slightly improved survival. The explanation for this observation was that the tracheostomy prevented pneumonia that can be caused by aspiration of tube-feeding formula.

Our survival rate results cannot be compared directly to most of the previous studies about the survival rates of children with cerebral palsy.[6-13] In every one of these reports, the most severe category described has a very broad definition such that all 6 groups from our study would fit within that category. In our study, the mortality rate in groups 1 and 2 was fairly large, whereas no deaths occurred in groups 5 and 6. Thus, depending on the mixture of numbers of cases from groups 1 through 6, one can obtain almost any survival rate outcome. Our results can only be compared to studies that are fine-tuned enough to separate different degrees of neurologic disability. The major accomplishment of Eyman[2] was to define groupings of neurologic severity such that subsequent investigators, such as ourselves, could collect comparative data.

When our study population was limited to coincide with the six groups previously defined by Eyman,[2] we obtained survival rates that were significantly better. For example, for the most severely disabled individuals (group 1), our 8-year survival rate was 66%, whereas Eyman's result was 5%. In all of the six groups compared, irrespective of age groupings, we consistently obtained significantly better survival rates (Tables 3, 4, and 5).

The strongest predictor of survival was the presence of other significant medical diseases. In our study, the 10-year survival rate for healthier individuals was 90%, whereas that for those with other significant diseases was 45%. Our study was the first to show that the survival rate of severely disabled children is significantly reduced by a defined set of significant medical diseases. As a corollary, severely disabled children who are otherwise healthy have much better survival rates.

We found a significant correlation between survival rate and feeding technique. Although the survival rate in G tube feeders was significantly lower than in orally fed children, it was significantly higher than in nasogastric tube feeders. Improved outcome with G tube placement, related to decreased aspiration pneumonia and gastroesophageal reflux and bleeding, has been noted previously.[17-19] The relatively poor outcome of nasogastric tube–fed children may have been due to the higher incidence of other significant diseases in this population. Children who are medically fragile may be considered 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, G-tube–fed children still had a significantly greater survival rate than nasogastric tube–fed ones, irrespective of the presence or absence of other medical diseases. The best explanation for our observation is that the presence of a nasogastric tube promotes gastroesophageal reflux and vomiting, which can lead to aspiration and death. Ours was the first study to compare survival rates of G tube– and nasogastric tube–fed children.

It is not possible to claim that our better survival rates are due to the selective death of younger children who may not have lived long enough to enter into a nursing facility. When we excluded children younger than 1 year from the data analysis and studied only those between 1 and 15 years, we obtained significantly better survival rates than those of Eyman[2] (Table 4). Likewise, when we limited our study to those over 15 years, we also obtained significantly better survival rates (Table 5). Thus, excluding the youngest, and presumably the most ill, children from the study population did not diminish our improved survival rates.

Access to Medical Care

There are several likely reasons why we obtained improved survival rates compared to those of Eyman.[2] Eyman's data covered the period 1980-1991, whereas our study period was 1985-1996. Many medical advances occurred between 1980 and 1985, but, more importantly, there was a shift in the public and medical professional attitudes toward the care of disabled individuals. We pur-

posefully 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, G tubes and tracheostomies were almost nonexistent before 1985. An analogous situation has been observed in children with Down syndrome, for whom the 10-year survival rate was 37% for the period 1944-1955 but 86% for the period 1966-1975.[20] The primary reason for this improved survival is the surgical correction of congenital defects, particularly cardiac defects. Our study covers a time period of much more intense medical management of severely disabled individuals, which may account for better outcomes.

Although we used the results from three different SNFs, all of these facilities were under uniform medical direction during the study time period. In Eyman's studies,[1,2] data were collected from 21 regional centers located across California, and the medical care was provided by numerous different physicians and institutions and thus was of variable quality. There is no suggestion in Eyman's data[1,2] of any uniformity in medical care provided to the severely disabled. Thus, uniformity of medical interventions in our study may be a contributing factor to improved survival rates.

All of our studied individuals were in a SNF 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 Eyman's study,[2] 65% of the disabled individuals were residing at home, and only 3.5% were in a SNF. It is probable that the poor survival rates, particularly for medically fragile individuals, were due to a lack of prompt medical intervention.

The California data have been reanalyzed according to place of residence.[21] Survival rates were similar for residents at home and in group homes. However, when compared to survival rates in state institutions and other health facilities, home and group home placement resulted in a 25% higher mortality rate irrespective of the degree of physical disability. It should be noted that in this study, residents of SNFs were excluded from the data analysis. The authors concluded that the best explanation was reduced availability of medical care, especially emergency care, in the home and group home settings.

Another report from California on survival rates of children in the persistent vegetative state[22] between 1981 and 1996 provided further significant information. The mortality rate for these severely disabled children per 1000 person-years was 229 for 1981 to 1987, 149 for 1988 to 1992, and 135 for 1993 to 1996. This study noted that the median survival rate for this population was 1 year for 1980, but 4.2 years for 1993 to 1996. These data strongly suggest that progressively more intensive medical care has been provided to severely disabled children, thus improving survival rates.

These survival results,[21,22] in addition to our own results,[3] strongly suggest that the critical factor in determining survival rate in disabled children is the availability of acute medical care.

Methodologic Errors in the California Data

Shortly after the publication of our survival rate results,[3] we became aware of a publication by Strauss,[23] who further analyzed the California data. In this article, Strauss explained the statistical methods that had been used by Eyman[1,2] as follows: "Any child whose condition improved was removed from the analysis. Since such children would have a better outlook, this procedure results in lower life expectancies than if an actual cohort of children had been followed." In a subsequent article,[24] Strauss elaborated further: "In particular, only children whose condition did not change were included. Because a child observed for a short period is less likely to display change than one observed for a long period, those who die early are more likely to be included. This accounts for the generally pessimistic prognoses in their study." Strauss typified the statistical approach used by Eyman[1,2] as a "methodologic mistake"[24] and as "an arithmetic blunder" that led to "mortality rates about 3 times too high."[25]

In response to Strauss's published statements about their work, Grossman and Eyman[26] fully agreed with the comments concerning the statistical approach that they had used: "Individuals who improved or regressed in their basic skills were no longer included in the original groups." Indeed, in the original Eyman publication of 1993,[2] the following sentence appeared in the methods section: "Subjects whose skills improved in the period of study were excluded from analysis because they

Table 6. Comparison of 5-Year Survival Rates

	Group 1	Group 2	Group 3
Analysis for cases under 1 year of age			
Eyman[2]	16%	30%	25%
Strauss[27]	43%	62%	77%
Plioplys[3]	75%	89%	91%
Analysis for cases over 1 year of age			
Eyman[2]	50%	60%	55%
Strauss[27]	55%	69%	69%
Plioplys[3]	82%	94%	93%

NOTE: The groupings are approximate. See text for details.

would then belong in a less vulnerable group with a better life expectancy."

In their 1998 article, Grossman and Eyman[26] shed further light on the methodologic difficulties surrounding their work. These authors explained that all of the information that they used for their data analysis came from the client development evaluation report (CDER) submitted by regional centers to obtain reimbursement for care provided to disabled individuals. Given the fact that all of the data used in their analysis came from a system meant to provide financial reimbursement, concerns should be raised about the basic validity of their clinical data.

Grossman and Eyman[26] further undermined the validity of their data by commenting that the clinical information on etiology of the disability and other ongoing medical problems was "notoriously unreliable." If the basic clinical data used in the Eyman studies[1,2] were based on "notoriously unreliable" sources, then all of the published results based on these data may also be "unreliable."

Given the clarification of the statistical methods used by Eyman,[1,2] and the fact that Strauss has continued to reanalyze the California data, comparing our results with those subsequently published by Strauss and colleagues is a worthwhile exercise. In the first publication clarifying Eyman's statistical methodology, Strauss[23] reanalyzed the Eyman data and found that of the lowest-functioning group (group 1), children age 5 years, at least 60% lived to age 10 years. From our own published data, the results for this scenario indicate that the survival rate to age 10 would be 72%.[3] Thus, once the statistical methodology is corrected, the California data become more similar to our results.

In another article dealing with this issue, Strauss reanalyzed the California data for group 3.[24] According to Eyman's methods, the 10-year survival rate would be 35% (34% if taken directly from Eyman),[2] whereas if the "methodologic mistake" were avoided, then the survival rate would be 70%. Our results for this category of disability gave a survival rate of 90%.[3] Thus in both groups 1 and 3, once the Eyman methodologic flaw is corrected, the survival rates obtained by Strauss are much more in keeping with our own results.

Using the same data base that Eyman had used, Strauss reanalyzed the California data but with different definitions of functional groups.[27] Because Strauss's group definitions are different, precise comparisons of groups are not possible, but fairly accurate approximations can be made. Group A (tube fed, not able to lift head) corresponds to group 1. Group B (tube fed, can lift head, but cannot roll) corresponds to group 2. Group C (no tube, but cannot lift head) approximates group 3. The 5-year survival rates are summarized in Table 6. These comparative data reveal that Strauss's results are more in keeping with our published results than with the results of Eyman.[2]

In a subsequent study, Strauss again used different groupings.[4] The group that could not lift the head corresponds to group 1; the group that was not tube fed, but was fed by others corresponds to group 3; the group that was fed by others corresponds to group 4. The comparative survival rate results are summarized in Table 7. Again, Strauss's results are much more in keeping with our results than with Eyman's results.[2]

Several comparisons can be made between our data and data obtained from a study of adults with

Table 7. Comparison of 10-Year Survival Rates

	Group 1	Group 2	Group 3	Group 4	Group 5	Group 6
Eyman[2]	32%	36%	37%	50%	78%	68%
Strauss[4]	45%	62%	70%	80%,87%	85%	94%
Plioplys[3]	73%	85%	90%	91%	100%	100%

NOTE: The groupings are approximate. See text for details.

Table 8. Comparison of 8-Year Survival Rates.

	Group 1	Group 2
Eyman[2]	38%	40%
Strauss[5]	63%, 65%	81%
Plioplys[3]	73%	94%

cerebral palsy by Strauss and Shavelle.[28] In that study, for the group that had no ability to lift the head and was tube fed from age 15 years, the life expectancy was 30.4 years for females and 26.7 years for males. In the group that was tube fed but had the ability to lift the head, life expectancy was 36.1 years for females and 31.9 years for males. Our comparable group 1 data suggest a median survival of slightly more than 40 years. Again, the data published by Strauss are much more in keeping with our results.

The most recent work by Strauss et al[5] concerns the survival rates of children with minimally conscious states. Children in the vegetative state and in the immobile minimally conscious state correspond to group 1, and those in the mobile minimally conscious state correspond to group 2. The comparative results are summarized in Table 8. Once again, Strauss's results are much more in keeping with our results than with Eyman's.[2]

A recent brief report[29] claims that the California data provide similar survival rates to those reported in a recent study from Western Australia.[13] It should be noted is that the comparative California result was based on Blair's definition of degrees of severity,[13] with the "severe" group simply not able to ambulate and "moderate" severity group able to walk with an assistive device. The current California data incorporate all of Eyman's data[1,2] and expand on it. In reviewing Eyman's 1993 report,[2] for those six groups with no mobility (not being able to walk or crawl, corresponding to Blair's "severe" category), and with an IQ <20, the studied number of cases is 3157. It should be noted that this is a minimum number of "severe" category of cases within the California database, as of the 1993 report. Since the publication of Eyman's report,[2] the California database has more than doubled in size. Thus the expected number of cases corresponding to Blair's "severe" category, and with an IQ of <20, must be, at a bare minimum, more than 6,000 cases, and more realistically more than 10,000 cases. However, in the report claiming similar outcomes, the California results are based only on 974 cases.[29] Thus this report was based on a highly selected set of cases from the California database. As already mentioned, our survival rate results[3] vary greatly with the degree of severity of disability. Those who are most severely affected (group 1) have a markedly reduced life expectancy, but no deaths occurred in groups 5 and 6. Thus, based on the mixture of cases from groups 1 through 6 (all of whom fit into Blair's "severe" category), one can generate any survival rate outcome. Thus, the claim of similar outcome results[29] cannot be accepted as presented.

In summary, Strauss and colleagues have reanalyzed the Eyman[1,2] data correcting methodologic problems and further expanded on these investigations. In all cases, the survival rates as reported by Strauss are higher than those reported by Eyman and much closer to those that we found.[3] Nevertheless, in all cases, our survival rate data provide better outcomes than those in the most recent reports by Strauss and colleagues (Tables 6, 7, and 8). These differences are due to a number of factors, including more intensive medical care provided to disabled children since 1985, uniformity of medical and nursing care provided, and the availability of acute medical care in the SNF as opposed to the home or group home setting.

Critique of the California Data

It would be of value to discuss in more detail the methodologic difficulties underlying the epidemiologic studies reported from California. The database used in the California studies was started in 1980 and to date contains information on over 200,000 individuals with various disabilities. This database was used by Eyman, Strauss, and their colleagues in the studies mentioned earlier. Given the large population, these authors' numerous publications have had a great impact in medical, legislative, and legal circles. However, their database has some very significant limitations. Given the influence of tube feeding has on mortality rates, it is surprising that the California data were collected in such a way that they cannot distinguish between G tube and nasogastric tube feedings.[4,16,27] In our study, 9% of the children were fed chronically by nasogastric tube, a feeding method associated with a lower survival rate than that associated with G tube feeding.[3] Thus, without specifying the tube feeding method used, the survival rate cannot be accurately determined.

The information in the California database was collected at 21 regional centers. The clinical information and the CDER were updated on a yearly basis. The CDER form contains more than 200 entries concerning medical, behavioral, and adaptive skills and limitations. Filling out the form correctly requires a considerable amount of medical knowledge, in particular a firm knowledge of ICD-9 coding. Given the complexity of the ICD-9 coding system, this skill is not easily mastered, even by medical professionals. Eyman[2] explained that the CDER was filled out by caseworkers in the community and by psychiatric technicians in institutions. Because most of the reported individuals resided at home, caseworkers provided the majority of clinical information. Even though the caseworkers had been trained in filling out the forms, it is not conceivable that they had the necessary medical knowledge and expertise to fill out the complicated forms fully and accurately. Thus it is not surprising that in Eyman's report[2] in the study population of 7226 neurologically disabled children, the presence or absence of epilepsy could not be determined in 2572 cases (36%). Epilepsy is an integral part of a person's neurologic disability that should have been easily ascertainable even to those with little medical background.

Strauss and Shavelle[28] gave a slightly different explanation about the source of the CDER data. They explained that the CDER was filled out by a social worker, who obtained the information from the caregiver. Because most of the studied individuals lived at home, the caregiver was a family member. Both the sources of the data and the data collectors had no medical training or background. Therefore, it is not surprising that in Strauss and Shavelle's report on 23,795 adults with developmental disabilities, there was no basic neurologic diagnosis in 14,376 cases (60%) and an "ill defined" diagnosis in 4325 cases (18%).[28] Thus in this study, fully 78% of the subjects had no neurologic diagnosis for their disability. The same basic diagnostic deficits appear in a study of 13,378 children with cerebral palsy with no diagnosis in 8449 cases and an "ill-defined" diagnosis in 2554 cases, resulting in 82% of children having no neurologic diagnosis for their disability.[4] Thus in the California database, 78% of adults and 82% of children had no neurologic diagnosis for their disabilities. This medical database is markedly deficient.

Both of the above-cited studies[4,28] covered the period 1980-1995. It may be argued that the older data were more imprecise and incomplete. However, a more recent report, dealing with 5075 severely neurologically disabled children from 1988 to 1997, resulted in no basic neurologic diagnosis in 39.7% of cases.[5] Thus the problem lies not with the time period from which the data were collected, but rather in the data collecting system itself.

Besides immobility and tube feeding, we found that other serious medical conditions are very important predictors of survival rates.[3] Eyman[1] also paid attention to this issue and in the CDER defined a number of serious medical conditions as "diabetes, heart disease, chronic respiratory infection, or hepatitis." This list of illnesses appears on the CDER itself. A subsequent report by Eyman[2] listed other serious medical conditions but did not explain them. Strauss[22] explained that the identification of severe medical conditions was from a separate listing of a "severe" identification on the CDER next to the disease code number. Indeed, on the CDER itself there is a "condition impact" modifier space next to the medical illness ICD-9 code. However, without a definition of what the term "severe" means in association with any given medical condition, the use of this designation is at best entirely unreliable and at worst completely capricious. It is thus not surprising that in a series of reports,[1,2,22] the presence of other serious medical conditions is specifically noted to have no impact on survival rates. Such results are completely contrary to our findings[3] and are totally contrary to common clinical experience. A child with, say, frequent bouts of pneumonia cannot have a life expectancy equivalent to that of a child who is otherwise healthy. In comparison, we precisely defined a set of serious medical conditions, and found a strong association between these illnesses and survival rates.[3]

In one specific serious medical problem, refractory epilepsy, we found a markedly decreased survival rate in those who experienced more than one seizure per day.[3] Our results are in keeping with previous reports on decreased survival expectations associated with epilepsy in children with cerebral palsy.[6-8,12] Furthermore, in otherwise neurologically and medically healthy individuals, epilepsy itself has been shown to decrease survival rates.[30,31] Given these results, it is surprising to

repeatedly find the California data stating that epilepsy and the frequency of seizures have no impact on survival rates.[1,2,22] The only possible explanation for this discrepancy is that the data used in the California reports are inaccurate.

Given all of the aforementioned difficulties with the California data, it is not surprising that Grossman and Eyman[26] characterized the clinical information collected as being "notoriously unreliable." As they themselves explained, the information was collected for financial reimbursement purposes. Reimbursement was based solely on one section of the CDER dealing with physical and cognitive impairment scales, not with the sections dealing with background medical and neurologic information. Financial pressures easily overcome the need for careful and accurate filling out of lengthy and complex governmental forms.

All of the published reports from California mention the statistical reliability of the CDER. However, in the cited references, statistical evaluations had been performed only on the 66 questions concerning functional impairment.[32] The statistical reliability of the basic medical and neurologic information contained in the CDER has never been formally investigated and thus remains "notoriously unreliable."[26]

Although the number of cases in our study was not large (447 patients),[3] all of the cases were thoroughly examined, and their records reviewed by medical personnel with many years of experience taking care of neurologically disabled children (the Directors of Nursing of the SNFs, and AVP). This hands-on, professional approach produced an accurate picture of each child entered into the study. Thus the survival rate results from our study are fully reliable.

Strauss and Shavelle[33] correctly argued that insurance companies, in providing life insurance policies, rely equally on the reports of actuaries and physicians. Life expectancy estimates vary greatly depending on the medical illnesses that the life insurance applicant may have, and the ascertainment of such illnesses is the physician's responsibility. When conducting epidemiologic studies and making life expectancy predictions for disabled children, accurate medical information is just as necessary. Unfortunately, the California database, which has generated so many studies and reports, incorporated neither accurate nor adequate medical information.

CONCLUSION

Our published results[3] have given much better survival rates for severely disabled children than those previously reported. This article has discussed the reasons for this improved survival and compared our rates to those from other studies, particularly those using the California database. The major factors associated with improved survival are more intensive medical care provided to children with disabilities and access to prompt medical interventions in the SNF. We believe that our results are not unique to our geographic area, but rather are representative of those seen at pediatric SNFs across North America.

Given today's climate of increased governmental and private payer budgetary restrictions and mounting pressure for deinstitutionalization of severely disabled children, our results are very sobering. Placement of severely disabled children in environments where acute medical care is not readily available will result in a dramatic increase in mortality rates.

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