

# LIFE EXPECTANCY OF SEVERELY DISABLED CHILDREN: A BRIEF REVIEW

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## INTRODUCTION

The term *life expectancy* is frequently misunderstood. When a determination of life expectancy is made, it is not a precise prediction of how long a person will live; rather, it is a statistical average of future life spans. For example, the current life expectancy of women in the United States is 79 years. That does not mean that

all women will live to 79 years of age; rather, it means that the average life span for all women is 79 years. Out of 100 healthy 20-year-old women identified today, approximately 50 will have died and 50 will still be living 59 years from now. That means some may have died in their 20s, 30s, etc., and some will live into their 80s and beyond. The term *life expectancy* always refers to an average life span expectation for a group of individuals and can never be an exact prediction of an individual's life span.

In statistical terms, life expectancy is the arithmetic average of life spans of a group of individuals, and median survival time is the midpoint of a group of life spans. Because some individuals within any group will live to a much older age, median survival time is always less than life expectancy. The survival rate is expressed as the percentage of a group's members who are still alive at a given time. In estimating future medical care costs, life expectancy determinations are the most appropriate.

Life expectancy data are collected and analyzed in the United States by the National Center for Health Statistics. This extremely large database consists of the entire population of the United States; however, when one is dealing with disabled children, the numbers that have been studied and reported are, in comparison, extremely small. Because many medical factors play a significant role in the life spans of all disabled children, the following presents a brief review of survival rate results from our work with such children in the Chicago, IL, area. It also defines many of the clinical factors that require evaluation and offers a critical review of literature that has been published on this subject.

## SURVIVAL RATE RESULTS

Reduced survival rates of neurologically disabled children have been noted in many studies.<sup>1-11</sup> The most comprehensive and detailed analysis of physically disabled children was performed by Eyman and his colleagues, who studied a total of 7226 individuals in California.<sup>4,5</sup> These investigations reported extremely short life expectancies, which were at odds with our own clinical experience. It should be emphasized that the groupings as defined by Eyman are extremely important ones.<sup>5</sup> It is common medical experience that the more medically compromised a child is, the shorter the life expectancy, but Eyman made the first attempt to stratify and fine tune degrees of disability as factors in life expectancy. As a common basis for the groupings, all of the individuals were immobile and incontinent and had profound mental retardation. In addition, the factors that separated the individual groups dealt with tube feeding, ability to roll, and the presence of functional hand or arm use.

The primary focus of our study was to identify subgroups of severely disabled children who would be identical to groups previously described by Eyman (Table 37.1) and to determine their survival rates. We investigated our own experience with neurologically disabled children at three skilled nursing facilities (SNFs) in the Chicago area and tried to identify additional clinical factors that would affect survival rates. The full details of our investigations have been published,<sup>12,13</sup> and reviews and more detailed comparisons of our results are also in print.<sup>14,15</sup>

When we did our first statistical analysis, we were stunned by our results. The 8-year survival rates are compared in Table 37.2. We found statistically significantly

**Table 37.1 Clinical Definition of Groups in Accordance with Eyman et al.<sup>5</sup>**

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

*Note:* In all cases, the individuals had cerebral palsy and did not have a progressive neurological disease process. All cases had profound mental retardation, were immobile, and were incontinent.

**Table 37.2 Eight-Year Survival Rate for Cases Less Than One Year of Age**

Group Number	8-Year Survival Rate (%)		P Value
	Eyman et al. <sup>5</sup>	Plioplys <sup>12</sup>	
1	5	66	<0.001
2	22	89	<0.001
3	21	92	<0.001
4	30	91	<0.001
5	70	100	Not significant
6	50	100	Not significant

better survival rates in comparison to Eyman's results in groups one through four. The most dramatic difference was in group one, the most disabled group, in which the previous report showed a 5% 8-year survival rate while we obtained a survival rate of 66%.

## MOBILITY

In our study, we found significant associations between degree of mobility and survival rates. Those who were totally immobile had a 10-year survival rate of 76%, whereas any degree of mobility (i.e., walking, crawling, creeping, or scooting) gave a survival rate of 96% ( $P < 0.001$ ). These results are in keeping with many other published reports.<sup>2-9,11,16</sup>

## MENTAL RETARDATION

Many studies have reported decreased survival rates in children with more pronounced mental retardation.<sup>1,2,6-10</sup> Although we also found an association between the degree of mental retardation and the survival rate, this association was not statistically significant.

## OTHER SIGNIFICANT MEDICAL ILLNESSES

In our study, we determined the presence of other significant medical diseases.<sup>12</sup> We defined these as two or more bouts of pneumonia per year, recurrent bowel obstructions, refractory seizures (i.e., greater than one seizure per day), severe asthma, cardiac arrhythmia, and progressive cardiac failure. Once the statistical analysis was completed, only pneumonias and refractory seizures were significant determinants because we found extremely few cases that presented with the other defined medical illnesses. A significant difference was found in survival rates, with the healthier group of individuals having a 10-year survival rate of 90% compared to the group of ill individuals whose rate was 45% ( $P < 0.0001$ ). In the previous studies by Eyman, stratification of survival rates according to other concomitant illnesses was, in fact, not done.<sup>4,5</sup> 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, we also showed that severely disabled children, who are otherwise healthy, have much better survival rates.

## EPILEPSY

We found an association with epilepsy — those children with more frequent seizures had a much greater mortality rate than those with less frequent seizures. Those who did not have seizures had a 10-year survival rate of 87%, whereas those who averaged more than one seizure per day had a rate of 33% ( $P < 0.001$ ). Similar associations between epilepsy and decreased survival rates in neurologically disabled children have been reported.<sup>2,3,7,9,11</sup>

## FEEDING

In our study, the 10-year survival rate for self-feeders was 95%; for those fed orally by others, it was also 95%. Survival rates for individuals fed by gastrostomy tube and nasogastric tube were 78% and 41%, respectively. These results, which demonstrate decreased survival rates with the use of tube feedings, are in keeping with other published reports.<sup>2-7,9,11,16</sup> We found a significant correlation between survival rate and feeding technique. Although the survival rate was significantly less in gastrostomy-tube-fed individuals when compared to those who were fed orally, it was significantly better when compared to nasogastric-tube-fed children. This result agrees with previous findings that have demonstrated that gastrostomy tube placement improves outcomes because it decreases the incidence of aspiration pneumonias.<sup>17-19</sup>

## TRACHEOSTOMY

In our study, no significant difference was found in survival rate depending on the presence of a tracheostomy tube. The only other published report about the survival rate of children with tracheostomies showed that tracheostomy caused slightly improved rates of survival in those who were tube fed.<sup>20</sup> The authors of that report felt that tracheostomies helped prevent aspiration pneumonias.

## MORE INTENSE MEDICAL AND NURSING CARE

Whereas the Eyman data covered the decade from 1980 through 1991, we conducted our study during the decade from 1985 through 1996. Not only had many medical advances occurred between 1980 and 1985 — the years these studies were launched — but, more importantly, the attitudes of both the public and medical professionals regarding the care of disabled individuals underwent a profound shift during this 5-year period. We purposely limited our study to the years after 1985 because, prior to 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, prior to 1985, gastrostomy tubes and tracheostomies were almost nonexistent; many disabled children were simply provided comfort care, and acute medical illnesses were left untreated.

In 1982, the Baby Doe legal case drew considerable public attention. This was a child with Down syndrome who had intestinal atresia, was denied surgery, and died. Subsequently, the Baby Jane Doe case of a child with untreated spina bifida was reviewed by the U.S. Supreme Court. In a 1984 response to these extremely high-profile cases, the U.S. Department of Health and Human Services made a ruling that all disabled infants and children should have full access to medical care. This ruling, made under the auspices of the Child Abuse and Protection Act, held that not providing medical treatment to a disabled child was equivalent to child abuse; therefore, medical practitioners who refused to treat these children could face criminal prosecution and incarceration. Also, in 1985 the federal Children with Special Health Care Needs (CSHCN) program was established. Thus, in 1985 in Chicago, California, and across the entire United States, medical care of disabled children advanced dramatically.

The trend for improved survival rates continues in more recent publications. In a study of children in the persistent vegetative state, the mortality rate of these severely disabled children progressively declined over the 1981 to 1996 time period.<sup>21</sup> This drop in mortality rates strongly suggests that more intensive medical care has also been provided to this population.

Our study covers a time period of much more intense medical management of severely disabled individuals and this, in part, accounts for our better outcomes. An analogous situation is the observation that survival rates have improved substantially in Down syndrome. From 1944 to 1955, the 10-year survival rate for individuals with Down syndrome was 37%; from 1966 to 1975, the survival rate was 86%.<sup>22</sup> The primary reason for this improvement in survival is the surgical correction of congenital defects, particularly cardiac ones.

## UNIFORM HIGH-QUALITY MEDICAL CARE

Although we used the results from three different SNFs, they all were under uniform medical and nursing direction and continuously provided care of the highest quality throughout the study period. In Eyman's studies, data were collected from 21 regional centers located across California, and the medical care was provided by a very large number of different physicians and institutions.<sup>4,5</sup> Such a large number of diverse medical care providers would have produced discrepancies in the quality of medical care. In Chicago's general medical community, many practitioners have believed, and still do, that these disabled children should be allowed to die. If one were to stop feeding and providing liquids to the most disabled group, life expectancy would be measured in days, not years. This attitude that espouses the neglect of disabled children has found a notable proponent in Peter Singer, Professor of Bioethics, at Princeton University. He has published several books in which he argues that the active euthanasia of disabled individuals is ethically totally acceptable. Given the prominence of his position and institution, Singer's words are being echoed in hospital ethics committees across the country. I find these attitudes to be abhorrent and, with the support of the Pediatric Long-Term Care Section, introduced a resolution to the House of Delegates of the American Medical Directors Association (AMDA) in 2000.<sup>23</sup> The resolution, which was accepted unanimously, stated, "AMDA opposes any physician involvement in assisted suicide or active euthanasia of any person regardless of age." Almost certainly, across the state of California, there were and are many physicians with this attitude whose practice was and is to limit access of disabled children to medical care, thus producing decreased survival rates. The decrease in survival rates that necessarily arises from such practice is the most important factor that cannot be accounted for in the California database, but the consistently high quality of care provided throughout our study is a guarantee of a more uniform and scientifically valid outcome.

## ACCESS TO ACUTE MEDICAL CARE

All the individuals we studied were cared for in SNFs where registered nurses are available in-house to provide acute medical care 24 hours a day, 7 days a week. This intensity of medical service undoubtedly also contributed to better survival rates. Acute illnesses are addressed promptly. In Eyman's study, 65% of the disabled individuals were residing at home, and only 3.5% were in a SNF.<sup>5</sup> 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.<sup>24</sup> Home and group home placement resulted in a 25% greater mortality rate, when compared to placement in state institutions. This result was the same irrespective of the degree of physical disability. The authors concluded that the best explanation was reduced availability of medical care, especially emergency care, in home settings.

## COMPARISON OF SURVIVAL RATES

The major accomplishment of Eyman was to define groupings of neurological severity such that subsequent investigators, such as ourselves, could collect

comparative data.<sup>5</sup> Consequently, our survival rate results cannot be compared directly to most of the previous studies about survival rates of children with cerebral palsy because these reports define the most severe category so broadly that all six groups within our study would fit within it.<sup>1-3,6-11,16</sup> Although we found the mortality rates in groups one and two were fairly large, no deaths occurred in groups five or six. Thus, depending on the mixture of numbers of cases from groups one through six that researchers employ, they can obtain almost any survival rate outcome. For this reason, our results can only be compared to studies that are fine tuned enough to separate different degrees of neurological disability. As a corollary, none of the aforementioned studies can be used to make life expectancy determinations. In all cases, the definition of the severe category of disability is too broad and imprecise. Without knowing the exact mixture of patients from groups one through six included in the reports, the results cannot be applied to children with multifactorial disabilities; therefore, the only studies that are fine tuned enough to be useful in the issue of survival rates of children with disabilities are those from California and our own report.<sup>12</sup>

### COMPARISON OF SURVIVAL RATES: CALIFORNIA DATA

Shortly after the publication of our survival rates results, we became aware of a publication in the *Lancet* by Strauss, who was continuing further analysis of Eyman's California data.<sup>25</sup> In this article, Strauss explained the statistical methods that had been used by Eyman 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."<sup>25</sup> In a subsequent article, 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."<sup>26</sup> Strauss typified the statistical approach used by Eyman as a "methodologic mistake" and as "an arithmetic blunder" that led to "mortality rates about three times too high."<sup>4,5,20,27</sup>

In response to Strauss's published statements about their work, Grossman and Eyman 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."<sup>28</sup> Indeed, in reviewing the original Eyman publication of 1993, the following sentence appeared in the methods section: "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."<sup>5</sup>

In their 1998 article, Grossman and Eyman shed further light on the methodological difficulties surrounding their work. They explained that all of the information that they used for their data analysis came from the Client Development Evaluation Report (CDER) that was submitted by regional centers in order to obtain reimbursement for care provided to disabled individuals. Given the fact that all the data used in their analysis came from a system meant to provide financial reimbursements, concern should be raised about the basic validity of their data.

Grossman and Eyman further undermined the validity of their data by commenting that the clinical information regarding etiology of the disability and other ongoing medical problems was “notoriously unreliable.”<sup>28</sup> If the basic clinical data used in the Eyman studies were based on “notoriously unreliable” sources, then all the published results based on this data may also be unreliable.<sup>4,5</sup>

Given the clarification of the statistical methods used by Eyman et al. and the fact that Strauss has continued to reanalyze the California data, it is worthwhile comparing our results to those subsequently published by Strauss and his colleagues.

In the first publication clarifying Eyman’s statistical methodology, Strauss reanalyzed the Eyman data and found that at least 60% of the children in group one (the group with the lowest functioning) lived to 10 years of age.<sup>25</sup> From our own published data, the results for this scenario indicate that the survival rate would be 72% to the age of 10 years.<sup>12</sup> 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 three.<sup>29</sup> According to Eyman’s methods, the 10-year survival rate would be 35% (34% if taken directly from Eyman), whereas if the “methodologic mistake” were avoided, the survival rate would be 70%.<sup>5</sup> Our results for this category of disability, gave a survival rate of 90%.<sup>12</sup> Thus, in both groups one and three, once the Eyman methodological flaw is corrected, the survival rates obtained by Strauss are much more in keeping with our own results.

Using the same database that Eyman had used, Strauss reanalyzed the California data but with different definitions of functional groups.<sup>30</sup> Because it is difficult to compare our results to those of Strauss with these different definitions, only approximations can be made. The 5-year survival rates of the comparative studies are presented in Table 37.3. This table indicates that the Strauss results are more in keeping with our published results than with Eyman’s.<sup>5</sup> In a subsequent study, Strauss again used new groupings.<sup>26</sup> These comparative results are presented in

**Table 37.3 Comparison of Five-Year Survival Rates**

Study	5-Year Survival Rate (%)		
	Group 1	Group 2	Group 3
<b>Analysis for Cases &lt;1 Year of Age</b>			
Eyman et al. <sup>5</sup>	16	30	25
Strauss et al. <sup>30</sup>	43	62	77
Plioplys <sup>12</sup>	75	89	91
<b>Analysis for Cases &gt;1 Year of Age</b>			
Eyman et al. <sup>5</sup>	50	60	55
Strauss et al. <sup>30</sup>	55	69	69
Plioplys <sup>12</sup>	82	94	93

Note: Groupings are approximate.



**Table 37.4 Comparison of Ten-Year Survival Rates**

Study	10-Year Survival Rate (%)					
	Group 1	Group 2	Group 3	Group 4	Group 5	Group 6
Eyman et al. <sup>5</sup>	32	36	37	50	78	68
Strauss et al. <sup>26</sup>	45	62	70	80, 87	85	94
Plioplys <sup>12</sup>	73	85	90	91	100	100

Note: Groupings are approximate.

Table 37.4 and, again, are much more in keeping with our results than with Eyman's.<sup>5</sup>

A more recent study concerning the survival rates of children with minimally conscious states was published by Strauss et al. in 2000.<sup>31</sup> It found a 63% 8-year survival for those in a *vegetative state*, and our comparable group one survival rate was 73%. For the *immobile, minimally conscious state*, the Strauss study found a 65% survival rate compared with our group one rate of 73%, and, for the *mobile, minimally conscious state*, Strauss found an 81% survival rate compared with our group two result of 94%. Obviously, the Strauss results are, again, much more in keeping with our results than are the Eyman results, which would be, respectively, 38, 38, and 40%.<sup>5</sup>

In summary, Strauss and his colleagues have reanalyzed the Eyman data by correcting its methodological problems and have further expanded on these investigations.<sup>4,5</sup> 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;<sup>4,5,12</sup> however, in all cases, our survival rate data provide better survival outcomes than those reported by Strauss and his colleagues. We attribute our more favorable results to both the more intensive and uniform medical and nursing care provided the medically fragile child since 1985 and the acute medical care provided by the SNFs that is not available in the home or group settings.

## METHODOLOGICAL DIFFICULTIES IN THE CALIFORNIA DATABASE

The database used in California was started in 1980 and, to date, encompasses information on over 200,000 individuals with various disabilities. This database was used by Eyman, Strauss, and colleagues in the studies mentioned above. Given the large population represented in this data, their numerous publications have generated a substantial impact in medical, legislative, and legal circles; however, the methodological difficulties that underlie these epidemiological studies impose very significant limitations on this database.

The information contained in the California database was drawn from annual updates of clinical information and CDER forms collected from 21 regional centers. Even though a considerable degree of medical knowledge is necessary to complete the CDER forms, which contain over 200 entries regarding individuals' medical,

behavioral, and adaptive skills and limitations, Eyman explained the CDER data he accessed was filled out by psychiatric technicians in institutions and by community caseworkers.<sup>5</sup> The majority of individuals in his study resided at home, which indicates caseworkers provided the bulk of the clinical information. Although these caseworkers had been trained in filling out the forms, it is inconceivable that they had the necessary medical knowledge and expertise to fully and accurately complete such complicated forms. It is, therefore, not surprising that, within Eyman's study population of 7226 neurologically disabled children, the presence or absence of epilepsy could not be determined in 2572 (36%) cases.<sup>5</sup> Epilepsy is an integral part of a person's neurological disability, which should be easily identified even by someone with only a minimal medical background. Strauss and Shavelle gave a slightly different explanation about the source of the CDER data.<sup>32</sup> They explained that, because the majority of subjects in the study lived at home, the CDERs were filled out by social workers based on information supplied by caregivers, who were family members. This means that both the source of the collected data and the collectors of the data were individuals without medical training or background. Thus, it is not surprising that, in their report concerning 23,795 adults with developmental disabilities, no basic neurological diagnosis was noted in 14,376 cases and an "ill-defined" diagnosis was noted in 4325 cases.<sup>32</sup> Consequently, in this study, fully 79% of the studied cases had no identifiable neurological diagnoses for their disabilities.

The same basic diagnostic deficits appear in a large study of 13,378 children with cerebral palsy; 8449 cases had no and another 2554 cases had "ill-defined" neurological diagnoses. This represents a full 82% of these children for whose disability no identifiable neurological diagnosis was identified.<sup>26</sup> Thus, because 79% of adults and 82% of children did not have neurological diagnoses for their disabilities, the medical database used in these reports is transparently deficient.

Both of the above-cited studies covered the 1980 to 1995 time period,<sup>26,32</sup> and 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 identifiable basic neurological diagnosis in 39.7% of cases.<sup>31</sup> The problem, therefore, 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. Eyman et al. also paid attention to this issue, and in the CDER they had defined a number of serious medical conditions as "diabetes, heart disease, chronic respiratory infection, or hepatitis."<sup>4</sup> It is thus extremely surprising that in a series of reports by both Eyman and Strauss the presence of other serious medical conditions is specifically noted to have no impact on survival rates.<sup>4,5,21</sup> These results are completely contrary to our findings and to common clinical experience.<sup>12</sup> A severely disabled child who has very frequent pneumonias cannot have a life expectancy equivalent to that of a child who is otherwise healthy. In contrast, we precisely defined a set of serious medical conditions and found these illnesses had a strong impact on survival rates.

In one specific serious medical problem, that of refractory epilepsy, we found decreased survival rates in those who had more than one seizure per day.<sup>12</sup> Our results are in keeping with previous reports about epilepsy decreasing survival

expectations in children with cerebral palsy.<sup>2,3,9,11</sup> Furthermore, in otherwise neurologically healthy individuals, epilepsy has been shown to decrease survival rates.<sup>33,34</sup> Given these results, it is surprising to repeatedly find that the California data show that epilepsy and the frequency of seizures have no impact on survival rates.<sup>4,5,21</sup> It is only recently, in studying a mildly disabled group of patients, that the California data have indicated that epilepsy has an impact on life expectancy.<sup>35</sup>

The only possible explanation for all of these discrepancies in the California results is that the database used in these studies is incomplete and/or inaccurate.

## **COMPARISON OF THE CALIFORNIA DATA TO OTHER STUDIES**

It has recently been claimed that the California data provide survival rates similar to data reported from Western Australia.<sup>1,36</sup> The comparison was based on Blair's definition of "severe" disability as not being able to ambulate. The current California data incorporate and expand upon all of Eyman's data.<sup>5</sup> In reviewing the 1993 report for the six groupings with no mobility and with an IQ of less than 20, the number of studied cases was 3157. This is a minimum number, as many others in the 1993 report would have also fit Blair's "severe" category. Since 1993, the California database has more than doubled. Thus, the expected number of "severe" cases must be, at a bare minimum, well over 6000. It may be claimed that, because the reported comparison was started with patients at 5 years of age, the available database is smaller; however, another report of adults with cerebral palsy reveals that the available numbers are very large.<sup>32</sup> A total of 23,795 adults with cerebral palsy were studied, and the number with "severe" cerebral palsy (although this term was not clearly defined) was 8093. The number of cerebral palsied adults with profound mental retardation was 6939; thus, the available numbers for study are indeed large. However, the comparison of the California data to Blair's was based on only 974 cases, just a small fraction of available cases. Thus, this result of comparable outcomes was based on a highly selected set of cases from the California database. As mentioned above, depending on the mixture of severity of the disability studied, almost any survival rate outcome can be obtained. The claim of similar outcome results cannot be accepted as presented.<sup>36</sup> The best explanation for this result lies in a careful selection of cases studied. The same letter stated that the California data were compared to data from Great Britain and that similar results were obtained; however, no data have been published to substantiate this claim.<sup>6,36</sup>

## **RESPONSE TO 2004 ANALYSIS BY STRAUSS**

On February 28, 2004, Strauss posted criticism of our study on his website.<sup>37</sup> This criticism was posted exactly 6 years after the publication date of our article. He claims that "we made a serious methodologic error." These claims of supposed "errors" must be addressed.

Strauss gave an example of a hypothetical case where, in the first year of life, 90% would die, and between ages 1 and 2, 50% would die. Graphs can be generated using techniques identical to those in our publication.<sup>12</sup> From birth to

2 years of age results in a death rate of 95%, which is exactly what Strauss wrote should happen. Strauss incorrectly claimed that our cumulative death rate would have been 90.5%. Comparing the results from 1 year to 2 years of age, our graphic techniques result in a death rate of 50%. Strauss incorrectly claims that our methods would result in a death rate of 5%. Our techniques result in a death rate that is exactly what Strauss wrote should take place.<sup>37</sup>

Strauss's discussion of data presented in our Figure 4 (group 4)<sup>12</sup> is erroneous. He states that there were six individuals over the age of 15. He claims that none of them was "exposed" to death at age 4. In order for anyone to have reached the age of 15, they would also have reached the ages 1, 2, 3, 4, etc., through 15. So, the longer surviving individuals in group 4 were exposed to death every day of their lives, including their fourth birthday.

Strauss further claims that we had "assumed survival to age 15" and had "guaranteed these six could not die prior to age 15." This claim is incomprehensible. We provided the best medical care that we could to these disabled individuals, and they lived well past their 15th birthday. If providing good medical care results in a good clinical outcome, that is desirable and commendable, but it cannot be guaranteed. We made no "assumptions" and made no "guarantees" outside of providing the best medical care that we could. We just reported what happened. In contrast, the California database that Strauss has been using is seriously flawed. See the text above and references 14 and 15 for further details.

Strauss corrected Eyman's "methodologic mistake" and posted these results on his website.<sup>37</sup> Strauss's correction starts from 1 year of age and provides data in 5-year increments. Only information about groups 1 through 3 is provided. Comparative results can be obtained and these are presented in Table 37.5. In generating this table we used the more extensive database used by Eyman in his 1993 publication.<sup>4</sup>

Table 37.5 shows that Strauss's correction of the Eyman data results in much better survival rates. However, the results from our study continue to give better outcomes. It should be noted that in group 3, the survival rates from our study are identical to those of the Strauss corrections. The similarity in group 3 outcomes continues until 30 years of age. If our methodologic techniques were flawed, as claimed by Strauss in discussing our group 4 results, then it is inconceivable that our group 3 results (a more disabled group than group 4) would be basically identical to those of Strauss's correction. This similarity in outcomes in group 3 gives independent confirmation of the reliability of our data.

Our data differ significantly from the Strauss correction for the more disabled groups 1 and 2 (Table 37.5). The greatest difference is with the most disabled group 1. In our study, we provided consistent, high-quality medical care on a continuous basis to all of our patients, in pediatric skilled nursing facilities. In contrast, the Strauss and Eyman data are based on a state-wide data collection system which includes discrepant qualities of administered medical care. In the Strauss data, only 3.5% resided in skilled nursing facilities, the vast majority residing at home, where access to acute medical care is problematic. Providing consistent, high-quality medical care, as was done in our study, can fully account for the observed differences.

**Table 37.5 Comparison to Strauss's Correction of Eyman's "Methodologic Mistake"**

	<i>Group 1</i>	<i>Group 2</i>	<i>Group 3</i>
<b>Analysis for cases from 1 year of age to 5 years of age</b>			
Eyman et al. <sup>4</sup>	55%	63%	60%
Strauss (correction) <sup>37</sup>	66%	84%	<u>94%</u>
Plioplys et al. <sup>12</sup>	82%	94%	<u>93%</u>
<b>Analysis for cases from 1 year of age to 10 years of age</b>			
Eyman et al. <sup>4</sup>	35%	38%	38%
Strauss (correction) <sup>37</sup>	41%	67%	<u>88%</u>
Plioplys et al. <sup>12</sup>	75%	94%	<u>93%</u>
<b>Analysis for cases from 1 year of age to 15 years of age</b>			
Eyman et al. <sup>4</sup>	No data available		
Strauss (correction) <sup>37</sup>	27%	54%	<u>82%</u>
Plioplys et al. <sup>12</sup>	72%	85%	<u>82%</u>

## SUMMARY

When making life expectancy determinations in children with severe disabilities, many factors must be taken into account. These are listed in Table 37.6. Depending on the severity of disabilities, the survival rates will vary considerably. Beside the California studies and our own report, all of the published studies of life expectancy in cerebral palsy use categories so broad that, depending on the mixture of patients included, any outcome whatsoever can be obtained, which renders them useless for life expectancy determinations.<sup>12</sup> Our data gave much better survival rates for severely disabled children than those reported from California.<sup>12</sup> The Eyman studies appear to have had extremely serious methodological problems, so these reports must be considered totally erroneous. The more recent studies from California report much better life expectancies, results that are much more comparable to our findings than to those of Eyman; however, the methodological difficulties in the California database leave the more recent California results in question. The possible reasons for our better outcomes in comparison to the more recent California results include the more intensive medical care provided to the medically fragile child since 1985, the uniformity of high-quality medical and nursing care those children received, and the availability of acute medical care in SNFs that is not available in the home or group home setting. We believe that our results are not unique to our geographic area but rather are representative of those seen at high-quality pediatric skilled nursing facilities across North America.

**Table 37.6 Important Factors in Life Expectancy Determinations of Children with Severe Neurological Disabilities**

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Mobility
Ability to roll
Degree of mental retardation
Presence of other significant medical illnesses
Pneumonias
Refractory seizures
Feeding methods
Self-feeding
Fed by others
Tube fed
Gastrostomy tube fed
Nasogastric tube fed
Tracheostomy
Intensity of medical and nursing care
Uniformity of high quality medical care
Access to acute medical care

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