For the past 12 years, as the medical director of four pediatric skilled nursing facilities (SNFs) in the Chicago area, and as the founder and chairman of the Pediatrics Long-Term Care Section of the American Medical Directors Association, I have had considerable experience in caring for children with severe neurologic disabilities.

Despite the severity of neurologic disability and the accompanying medical illnesses, these children do have life and spirit. They respond to being held, cuddled, spoken to. They respond to visual stimuli, to sounds, to their names being called. They smile, laugh, coo, and in some cases speak words. These are the most disabled individuals in our society and they fully deserve our care and medical attention.

The neurologic diagnoses of the children that I care for are presented in Table 7.1. This diagnostic listing appears to be fairly representative of children who reside in pediatric SNFs across the United States. According to the most recent data compiled by Sister Katherine Smith of Portland, Oregon (personal communication), the total number of children cared for in the United States in pediatric long-term care facilities, with 24-hour-per-day nursing care, is over 4,200. There are 89 of these facilities and they are located in 28 states. The designations given to these facilities are varied and include SNFs, intermediate care facilities, residential schools, rehabilitation centers, and hospitals. Approximately 50% of these centers are independent, and the others are run as part of a geriatric nursing facility. All of the children in these facilities are medically fragile, with multiple medical problems in addition to their underlying neurologic difficulties. Infections (primarily pulmonary), feeding difficulties, worsening spasticity with joint contractures and scoliosis, disuse osteoporosis,
TABLE 7.1 Neurological Diagnoses by Percentage for the Entire Study Population \((N = 447)\)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prenatal (congenital) encephalopathy</td>
<td>32.4</td>
</tr>
<tr>
<td>Hypoxic-ischemic encephalopathy at birth</td>
<td>14.3</td>
</tr>
<tr>
<td>Encephalopathy with hydrocephalus</td>
<td>8.7</td>
</tr>
<tr>
<td>Encephalopathy with intraventricular hemorrhage after birth</td>
<td>6.5</td>
</tr>
<tr>
<td>Congenital infection</td>
<td>5.7</td>
</tr>
<tr>
<td>Head injury</td>
<td>5.4</td>
</tr>
<tr>
<td>Central nervous system malformation</td>
<td>4.9</td>
</tr>
<tr>
<td>Meningitis</td>
<td>4.3</td>
</tr>
<tr>
<td>Encephalitis</td>
<td>2.5</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>1.8</td>
</tr>
<tr>
<td>Other chromosomal abnormalities</td>
<td>1.6</td>
</tr>
<tr>
<td>Other congenital syndromes</td>
<td>2.5</td>
</tr>
<tr>
<td>Progressive central nervous system degenerative diseases</td>
<td>3.6</td>
</tr>
<tr>
<td>Other conditions</td>
<td>5.8</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
</tr>
</tbody>
</table>

and fractures are just several of the categories of medical problems that these children face.

Reduced survival rates of neurologically disabled children have been noted in many studies (Blair, Watson, Badawi, & Stanley, 2001; Crichton, Mackinnon, & White, 1995; Evans, Evans, & Alberman, 1990; Eyman, Grossman, Chaney, & Call, 1990, 1993; Hutton, Colver, & Mackie, 2000; Hutton, Cooke, & Pharoah, 1994; Hutton & Pharoah, 2002; Kudrjavcev, Schoenberg, Kurland, & Groover, 1985; Patja, Iivanainen, Vesala, Oksanen, & Ruoppila, 2002; Roboz, 1972). The most comprehensive and detailed analysis of physically disabled children was performed in California where a total of 7,226 individuals were studied (Eyman et al., 1990, 1993). This investigation reported extremely short life expectations, which were 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 SNFs in the Chicago area. The primary focus of our study was to identify subgroups of severely disabled children who would be identical to the previously described groups (Eyman et al., 1993) 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 (Plioplys, Kasnicka, Lewis, & Moller, 1998a, 1998b). This chapter reviews our survival rate results and compares them to those of other investigators. Particular emphasis is placed on a comparison with more recent reports, and a critique of the published California research results.

TECHNICAL TERMINOLOGY

The term “life expectancy” is frequently misunderstood. This 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 men in the United States is 72 years. That does not mean that all men will live to 72 years of age. Rather it means that the average life span will be 72 years. For example, if one identified 100 healthy men, all 20 years old, and followed them, 52 years from now approximately 50 would have died and 50 would still be alive. That means that some may have died in their 30's, 40's, and so on, and that some will live into their 80's and 90's.

It is worthwhile to clearly define technical terms that are used. Median survival time is the middle (median) of a group of life spans. Life expectancy is the arithmetic average of life spans of a group of individuals. Because some individuals within any group will live to an older age, life expectancy is always longer than median survival time. Survival rate is the percentage of a group living at a given time (such as: 70% are alive at 20 years of age).

OVERALL SURVIVAL RATE RESULTS

The vast majority of our studied population had cerebral palsy, profound mental retardation, were immobile, were incontinent, and did not have a
progressive central nervous system degenerative disease (Plioplys et al., 1998a). This set of patients was divided into clinical groups 1 through 6 (Table 7.2), which correspond exactly to groups 1 through 6 of Eyman (1993).

The 8-year survival rate of the entire study group, starting with less than 1 year of age, is analyzed and compared in Table 7.3. There is a statistically significantly better survival rate in our study as compared to the previously published results in groups 1 through 4. The most dramatic difference was in the most disabled group (group 1), in which the previous report showed a 5% survival rate and we obtained a rate of 66%. Likewise, for the next most severely disabled 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% whereas published results indicate a rate of 30%.

### TABLE 7.2 Clinical Definition of Groups in Accordance with Eyman et al. (1993)*

<table>
<thead>
<tr>
<th>Group</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1</td>
<td>tube fed, not rolling, no hand or arm use</td>
</tr>
<tr>
<td>Group 2</td>
<td>tube fed, not rolling, hand or arm use present</td>
</tr>
<tr>
<td>Group 3</td>
<td>fed by others, not rolling, no hand or arm use</td>
</tr>
<tr>
<td>Group 4</td>
<td>fed by others, not rolling, hand or arm use present</td>
</tr>
<tr>
<td>Group 5</td>
<td>fed by others, able to roll, hand or arm use present</td>
</tr>
<tr>
<td>Group 6</td>
<td>tube fed, able to roll, hand or arm use present</td>
</tr>
</tbody>
</table>

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

### MEDICAL FACTORS

#### Mobility

In collecting all of our clinical data (Plioplys et al., 1998a), we focused particular attention on detailing the clinical findings of each individual in accord with previously published group definitions (Table 7.2; Eyman et al., 1993). Cerebral palsy was defined as a severe impairment of motor movements, the cause of which occurred prior to 28 days of age. We abstracted clinical information concerning the type of cerebral palsy (spastic quadriplegia, spastic diplegia, athetoid, ataxic, and hypotonic). Mobility parameters were assessed as being able to walk, crawl, creep, 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 presence of any functional hand or arm use was also assessed.

Of our total studied population, 83% had cerebral palsy, 45% displayed no hand or arm movement, 21% had non-purposeful hand or arm movements, and 34% had purposeful hand or arm movements; 79% were immobile.

In our study (Plioplys et al., 1998a), 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% (p < 0.001). Any degree of being able to roll gave a 10-year survival rate of 98%, whereas being unable to roll gave a survival rate of 74% (p < 0.0001).
Our result of decreased survival rates with immobility is in keeping with many other published reports (Crichton et al., 1995; Evans et al., 1990; Eymad et al., 1990, 1993; Hutton et al., 1994, 2000, 2002; Kudrjavcev et al., 1985; Roboz, 1972; Williams & Alberman, 1998).

**Mental Retardation**

In our study the degree of mental retardation was determined from available psychologic examinations. Every resident in a pediatric SNF in Illinois must have a recent professional psychologic examination performed. Profound mental retardation was defined as an IQ < 20, severe mental retardation as an IQ between 20 and 35, and moderate mental retardation as an IQ between 36 and 50. Of our total studied population, 93% had profound mental retardation.

Many studies have reported decreased survival rates in children with more pronounced mental retardation (Blair et al., 2001; Crichton et al., 1995; Hutton et al., 1994, 2000, 2002; Kudrjavcev et al., 1985; Patja et al., 2000). We also found 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.

**Other Significant Medical Illnesses**

In our study, 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 obstructions, refractory seizures (more than one seizure per day), severe asthma, cardiac arrhythmia, and progressive cardiac failure. Of our studied 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% as compared to the group of ill individuals whose rate was 45% (p < 0.0001).

In the previous studies by Eymad (1990, 1993) stratification of survival rates according to other concomitant illnesses was not done. 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.

**Epilepsy**

In our study, 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. Of our total studied population, 40% had < 1 seizure per month, 17% had between 1 seizure per week and 1 per month, 4% had between 1 seizure per day and 1 seizure per week, and 3% had > 1 seizure per day. Of those with seizures, 97% had generalized seizures and 3% had focal seizures. 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%. 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 < 0.001). Similar associations between epilepsy and decreased survival rates in neurologically disabled children have been reported (Crichton et al., 1995; Evans et al., 1990; Hutton et al., 2000; Kudrjavcev et al., 1985; Roboz, 1972).

**Feeding**

In our study, the method of feeding was determined: self-feeder, orally fed by others, nasogastric tube, and gastrostomy tube (G-tube). Of our total studied population, 52% were fed by G-tubes, 9% by nasogastric tubes, 34% were orally fed by others and 5% were self-feeders.

The 10-year survival rate for self-feeders was 95%, for those orally fed by others 95%, G-tube fed individuals 78%, and nasogastric tube fed individuals 41%. When the orally fed and self-feeder groups were compared to the G-tube fed children, the survival rate difference was statistically significant (p < 0.001). Likewise, when the orally fed and self-feeder groups were compared to the nasogastric tube fed children, the survival
rate difference was statistically significant \((p < 0.0001)\). When the G-tube fed children were compared to those with nasogastric tube feedings, there also was a statistically significant difference \((p < 0.001)\).

Survival rates for G-tube and nasogastric tube fed individuals 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.

Our result of decreased survival rates with the use of tube feedings is in keeping with other published reports (Crichton et al., 1995; Evans et al., 1990; Eyman et al., 1990, 1993; Hutton et al., 1994, 2000; Kudrjavcev et al., 1985; Roboz, 1972; Williams & Alberman, 1998). We found a significant correlation between survival rate and feeding technique. Although the survival rate was significantly less in G-tube fed individuals when compared to orally fed ones, it was significantly better when compared to nasogastric tube fed children. Improvement in outcome with G-tube placement, by decreasing aspiration pneumonia and gastroesophageal reflux and bleeding, has been noted previously (Bui, Dang, Chaney, & Vergara, 1989; McGrath, Splaingard, Alba, Kaufman, & Glicklick, 1992; Raventos, Kralemann, & Gray, 1982). The relatively poor outcome of nasogastric tube fed children may have been due to the higher incidence of other significant diseases in this population. Those 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 was 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 comparing survival rates of G-tube and nasogastric tube fed children.

### Tracheostomy

Of our total studied population, 11\% had a tracheostomy. There was no significant difference in survival rate depending on the presence of a tracheostomy tube. The 10-year survival rate for those with no tracheostomy was 83\% and with a tracheostomy was 75\% \((p = \text{NS})\).

The only other published report about the survival rate of children with tracheostomies (Strauss, Kastner, Ashwal, & White, 1997) showed that in those who were tube fed, tracheostomies slightly improved survival. The explanation for this observation was that tracheostomies prevented pneumonias that may have arisen from aspiration of tube feeding formula.

### Age and Survival

Previously published survival rates were further subgrouped by: (a) individuals less than 1 year of age, (b) individuals between 1 and 15 years of age, and (c) individuals older than 15 years of age (Eyman et al., 1993). We likewise subdivided our groups into these age categories to obtain comparable survival rate data.

Tables 7.4 and 7.5 compare the 8-year survival rates of the groups that are limited to those who have survived 1 year of age (Table 7.4) and those who have survived 15 years of age (Table 7.5). Again, we obtained statistically significantly better survival rates in groups 1 through 3 for these age subgroups when compared to the results of Eyman (1993).

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 less than 1 year of age from the data analysis and studied those between 1 and 15 years of age, we obtained significantly better survival rates than those of Eyman (1993; Table 7.4). Likewise, when we limited our study to those over 15 years of age, we also obtained significantly better survival rates.

<table>
<thead>
<tr>
<th>Group number</th>
<th>Eyman et al. (1993) (%)</th>
<th>Plioplys et al. (1998a) (%)</th>
<th>(p) value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>35</td>
<td>73</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>2</td>
<td>38</td>
<td>95</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>3</td>
<td>40</td>
<td>94</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>4</td>
<td>60</td>
<td>91</td>
<td>NS</td>
</tr>
<tr>
<td>5</td>
<td>80</td>
<td>100</td>
<td>NS</td>
</tr>
<tr>
<td>6</td>
<td>70</td>
<td>100</td>
<td>NS</td>
</tr>
</tbody>
</table>
TABLE 7.5 Eight-Year Survival Rate for Cases Older Than 15 Years of Age

<table>
<thead>
<tr>
<th>Group number</th>
<th>Eyman et al. (1993) (%)</th>
<th>Plioplys et al. (1998a) (%)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>58</td>
<td>86</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>no data</td>
<td>79</td>
<td>no data</td>
<td>NS</td>
</tr>
<tr>
<td>3</td>
<td>60</td>
<td>100</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>4</td>
<td>76</td>
<td>82</td>
<td>NS</td>
</tr>
<tr>
<td>5</td>
<td>92</td>
<td>100</td>
<td>NS</td>
</tr>
<tr>
<td>6</td>
<td>70</td>
<td>100</td>
<td>NS</td>
</tr>
</tbody>
</table>

(Table 7.5). Thus, excluding the youngest and presumably the most ill children from the studied populations did not diminish our improved survival rates.

Access to Medical Care

There are many reasons we obtained improved survival rates as compared to those of Eyman (1993). Eyman's data covered the 1980 through 1991 decade, whereas our study was over the 1985 through 1996 decade. There were 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 purposely limited our study to after 1985 because prior to that year, in Chicago, the approach to the care of disabled children was rudimentary in comparison to current standards. For example, at our nursing facilities prior to 1985, G-tubes and tracheostomies were almost nonexistent. An analogous situation is the observation that survival rates have improved substantially in Down syndrome. Ten-year survival rates for Down syndrome individuals for the 1944–1955 decade was 37%, and in the 1966–1975 decade was 86% (Fryers, 1986). The primary reason for this improvement in survival is the surgical correction of congenital defects, particularly cardiac ones. Our study covers a time period of much more intense medical management of severely disabled individuals and this may account for better outcomes.

In 1982 Baby Doe was a child with Down syndrome, who had intestinal atresia, was denied surgery and died. This legal case drew considerable public attention. Subsequently the Baby Jane Doe case of a child with untreated spina bifida was reviewed by the U.S. Supreme Court. In response to these extremely high profile cases, in 1984 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 was made under the auspices of the Child Abuse and Protection Act. Thus, not treating a disabled child became equivalent to child abuse. Medical practitioners who did not treat these children could face criminal prosecution. Also, in 1985 the Children with Special Health Care Needs (CSPCHN) program was established by the U.S. federal government. Thus, in Chicago, as across the entire United States, medical care to disabled children advanced dramatically in 1985.

Although we used the results from three different SNFs, they all have been under uniform medical direction over the study time period. In Eyman’s studies (1990, 1993) data was collected from 21 regional centers located across California, and the medical care was provided by a very large number of physicians and institutions. A large number of diverse medical care providers would have produced discrepant quality of medical care. There is no suggestion in Eyman’s data (1990, 1993) of any uniformity in medical care provided to the severely disabled. Thus, uniformity of medical interventions in our study may be a contributing explanation for improved survival rates.

All of our studied individuals were in an 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 (1993) 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.

In a report concerning survival rates of children in the persistent vegetative state (Strauss, Shavelle, & Ashwal, 1999) the mortality rate of these severely disabled children was analyzed over the 1981–1996 time period. The mortality rate per 1,000 person years from 1981–1987 was 229, from 1988-1992 was 149, and from 1993–1996 was 135. Strauss and colleagues also noted that in 1980 the median survival rate for this population was 1 year, whereas in 1993–1996 it was 4.2 years. This data strongly suggests that over this time period progressively more intensive medical care has
been provided to this population and thus mortality rates have dropped, as was hypothesized in our study (Plioplys et al., 1998a).

The California data has also been re-analyzed according to place of residence (Strauss, Eyman, & Grossman, 1996). 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% greater 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. These new survival results (Strauss et al., 1996), in addition to our own results, strongly suggest that the critical factor in determining survival rates in severely disabled children is the availability of acute medical care.

The medical care provided at our SNFs was continuously of the highest quality. In Chicago’s medical community, there are many practitioners whose attitude is that severely disabled children should be allowed to die. If one were to stop feeding these children, life expectancy would be measured in days, not years. This attitude of neglect toward disabled children has found a notable proponent, Peter Singer, Professor of Bioethics at Princeton University. He has published several books in which he argues that even active euthanasia of disabled individuals is ethically acceptable. Given the prominence of his position and institution, Singer’s words are being echoed during hospital ethics committees across the country. I found 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 in March 2000. The resolution was accepted unanimously and it 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 an attitude of limiting access of medical care to disabled children, thus producing decreased survival rates. This most important factor cannot be accounted for in the California data base. In our own study, high-quality medical and nursing care was provided uniformly and consistently.

### COMPARISON OF SURVIVAL RATE RESULTS

Our survival rate results cannot be compared directly to most of the previous studies about the survival rates of children with cerebral palsy (Blair et al., 2001; Crichton et al., 1995; Evans et al., 1990; Hutton et al., 1994, 2000, 2002; Kudrjavcev et al., 1985; Patjia et al., 2000; Roboz, 1972; Williams & Alberman, 1998). In every one of these reports the most severe category described has a very broad definition such that all six groups from our study would fit within that category. In our study, the mortality rate in groups 1 and 2 was fairly large, whereas in groups 5 and 6 no deaths occurred. Thus, depending on the mixture of numbers of cases from group 1 through group 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 and colleagues (1993) was to define groupings of neurologic severity such that subsequent investigators, such as ourselves, could collect comparative data.

Using the same database that Eyman had used, Strauss reanalyzed the California data but with different definitions of functional groups (Strauss, Ashwal, Shavelle, & Eyman, 1997). It is difficult to compare our results to those of Strauss because the group definitions are different. Only approximations can be made. It appears that group A in this study (tube fed, not able to lift head) would correspond to group 1. Group B (tube fed, can lift head, but cannot roll) would correspond to group 2. Group C (no tube, but cannot lift head) would approximately correspond to group 3. The 5-year survival rates of the comparative studies are presented in Table 7.6. A review of this table indicates that Strauss’s results are more in keeping with our published results than with Eyman’s (1993).
In a subsequent study, Strauss again used new groupings (Strauss, Shavelle, & Anderson, 1998). The comparative results are presented in Table 7.7. Again, Strauss’s results are much more in keeping with our results than with Eyman’s (1993).

In a study of adults with cerebral palsy (Strauss & Shavelle, 1998a), several comparisons to our data can be made. From Table 3 in the Strauss and Shavelle article, in the group that cannot lift the head and is tube fed, from the age of 15 years the life expectancy for females was 30.4 years and for males 26.7 years. In the group that is tube fed but can lift the head, the life expectancy for females was 36.1 years and for males 31.9 years. Our comparable group 1 data suggest a median survival of slightly over 40 years. Again, the data published by Strauss is much more in keeping with our results.

The most recent work by Strauss and colleagues concerning the survival rates of children with minimally conscious states was published in 2000 (Strauss, Ashwal, Day, & Shavelle, 2000). Although the groupings as presented are different from our own published results, approximate comparisons can be made. The 8-year survival of the vegetative state was 63% (our comparable group 1 result was 73%), for the immobile minimally conscious state 65% (our comparable group 1 result was 73%), and mobile minimally conscious state, 81% (our group 2 result was 94%). The Eyman and colleagues (1993) results would be respectively 38%, 38%, and 40%. These comparative results are presented in Table 7.8. Again, Strauss’s results are much more in keeping with our results than with Eyman’s results.

There is a recent letter from California (Shavelle, Strauss, & Day, 2001) claiming that the California data provides similar survival rates when compared to those reported from Western Australia (Blair et al., 2001). It should be noted that the comparative California result was based on Blair’s definition of degrees of severity, the “severe” group simply not being able to ambulate (members of the next group of “moderate” severity could still walk with assistive devices). The current California data incorporate all of Eyman’s data (1993) and expands upon it. In reviewing Eyman’s report of 1993, for the six groupings with no mobility (not being able to walk or crawl, thus corresponding to Blair’s “severe” category), and with an IQ of less than 20, the studied number of cases was 3,157. It should be noted that this is a minimum number of “severe” category of cases within the California database, as of the 1993 report. It would be expected that there were many more cases that did not fit into Eyman’s six groups of severity, but would still have been categorized as “severe” by Blair.

Since Eyman’s report of 1993, the California database has more than doubled. Thus, the expected number of cases corresponding to Blair’s “severe” category, and with an IQ of less than 20, must be, at a bare minimum, well over 6,000. However, in the report claiming similar outcomes, the California results are based on only 974 cases. Thus, this letter (Shavelle et al., 2001) was based on a highly selected set of cases from the California database. It may be claimed that since the reported comparison was started with patients at 5 years of age, the available data base may be smaller. However, a review of a study published by the same California researchers concerning adults with cerebral palsy reveals that the available numbers are, indeed, very large (Strauss & Shavelle, 1998a). In this 1998 report, a total of 23,795 adults with cerebral palsy was studied. The number with “severe” cerebral palsy (although this term was not
clearly defined in the report) was 8,093. The number of cerebral palsied adults with profound mental retardation was 6,939. Thus, the available numbers for study are indeed large.

Our own survival rate results (Plioplys et al., 1998a, 1998b) vary greatly with the degree of severity of disability. Those who are most severely affected, group 1, had a reduced life expectancy (the 8-year survival rate for this group was 66%). However, those with lesser degrees of impairment, groups 5 and 6, had no deaths over the studied time period. 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 whatsoever. The claim of similar outcome results (Shavelle et al., 2001) cannot be accepted as presented. The best explanation for similarity in outcome results lies in a careful preselection of cases studied. The same report (Shavelle et al., 2001) stated that the California data were compared to those from Great Britain (Hutton et al., 1994) and that comparable results were obtained. However, no data have been published to substantiate this claim.

**CRITIQUE OF THE CALIFORNIA DATA**

Shortly after the publication of our survival rates results (Plioplys et al., 1998a), we became aware of a publication in the *Lancet* by Strauss (1997), who was continuing further analysis of the California data. In this article, Strauss explained the statistical methods that had been used by Eyman (1990, 1993) 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” (p. 283). In a subsequent article (Strauss & Shavelle, 1998b), 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” (p. 243). Strauss typified the statistical approach used by Eyman (1990, 1993) as a “methodologic mistake” (Strauss & Shavelle, 1998b) and as “an arithmetic blunder” that led to “mortality rates about 3 times too high” (Strauss, 2001).

In response to Strauss’s published statements about their work, Grossman and Eyman (1998) 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” (p. 243). 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” (p. 330).

In their 1998 article, Grossman and Eyman shed further light on the methodologic difficulties of their work. They explained that all of the information they used for their data analysis came from the client development evaluation report (CDER), which was submitted by regional centers in order to obtain reimbursement for care provided to disabled individuals. Given the fact that all of the data that was used in their analysis came from a system meant to provide financial reimbursements, concern should be raised about the basic validity of their data. Financial motivations are very strong ones.

Grossman and Eyman (1998) 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.” If the basic clinical data that was used in the Eyman studies (1990, 1993) was based on “notoriously unreliable” sources, then all of the published results based on this data may also be “unreliable.”

Given the clarification of the statistical methods used by Eyman and colleagues (1990, 1993), 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 (1997) reanalyzed the Eyman data and found that for the lowest functioning group (group 1), those children who were 5 years of age, at least 60% lived to the age of 10 years. From our own published data, the results for this scenario indicate that the survival rate to the age of 10 years would be 72% (Plioplys et al., 1998a). Thus, once the statistical methodology is corrected, the California data become more similar to our results.

In another article dealing with this issue (Strauss & Shavelle, 1998b), the authors reanalyzed the California data for group 3. According to Eyman’s methods, the 10-year survival rate would be 35% (34% if taken directly from Eyman [1993]), whereas if the “methodologic mistake” were avoided, the survival rate would be 70%. Our results for this category of
21 regional centers. The clinical information and the CDER were updated on a yearly basis. The CDER is a form with over 200 entries concerning medical, behavioral, and adaptive skills and limitations. A considerable degree of medical knowledge is necessary to fill the form out correctly. Eyman (1993) explained that the CDER was filled out by caseworkers in the community and by psychiatric technicians in institutions. Since the majority of 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. It is, therefore, not surprising that in the Eyman report (1993), in the studied population of 7,226 neurologically disabled children, the presence or absence of epilepsy could not be determined in 2,572 cases, or 36%. Epilepsy is an integral part of a person’s neurologic disability, and should have been easily ascertainable even by those with little medical background.

Strauss and Shavelle (1998a) 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. Since the majority of studied individuals lived at home, the caregiver was usually a family member. Both the source of the collected data and the collectors of the data were mostly individuals without medical training or background. Thus, it is not surprising that in their report concerning 23,795 adults with developmental disabilities (Strauss & Shavelle, 1998a), there was no basic neurologic diagnosis in 14,376 cases, and an ill defined diagnosis in 4,325 cases. Thus, in this study, fully 79% of the studied cases had no identifiable neurologic diagnosis for their disability. The same basic diagnostic deficits appear in a large study of 13,378 children with cerebral palsy (Strauss et al., 1998) where no diagnosis appeared in 8,449 cases and an ill defined one in 2,554 cases, yielding a result that fully 82% of children carried no identifiable neurologic diagnosis for their disability. Thus, 79% of adults and 82% of children did not have a neurologic diagnosis for their disabilities. The medical database used in these reports is deficient.

Both of the above-cited studies (Strauss et al., 1998; Strauss & Shavelle, 1998a) covered the 1980–1995 time period. It may be argued that the older data was more imprecise and incomplete. However, a more recent report, dealing with 5,075 severely neurologically disabled children from 1988 to 1997 (Strauss et al., 2000), resulted in no identifiable basic

In summary, Strauss and his colleagues have reanalyzed the Eyman (1990, 1993) data correcting methodologic problems, and have further expanded on these investigations. In all cases the survival rates as reported by Strauss are higher than those reported by Eyman (1990, 1993) and much closer to those that we found. However, in all cases, our survival rate data provides better survival outcomes than those reported by Strauss and his colleagues. This difference is most likely due to a number of factors that were discussed above. They include more intensive medical care provided to the medically fragile child since 1985, uniformity of medical and nursing care provided, and the availability of acute medical care in the SNF that is not available in the home or group home setting.

**METHODOLOGIC DIFFICULTIES IN THE CALIFORNIA DATABASE**

It would be of value to discuss in more detail the methodologic difficulties underlying the epidemiologic studies that have been reported from California. The database used in California was started in 1980 and to date has 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 number, their numerous publications have produced a great impact in medical, legislative, and legal circles. However, there are very significant limitations to their database.

Given the impact that 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 (Strauss, Ashwal, et al., 1997; Strauss, Kastner, et al., 1997; Strauss, Shavelle, & Anderson, 1998). In our study, 9% of the children were chronically fed by nasogastric tube; this form of feeding was associated with decreased survival rates. Thus, without specifying the type of tube feeding used, the survival rates cannot be accurately determined.

The information contained in the California database was collected at 21 regional centers. The clinical information and the CDER were updated by nasogastric tube; this form of feeding was associated with decreased survival rates. Thus, without specifying the type of tube feeding used, the survival rates cannot be accurately determined.
Besides immobility and tube feeding, we found that other serious medical conditions are very important predictors of survival rates (Plioplys et al., 1998a). Eyman and colleagues (1990) 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. In a subsequent report by Eyman and colleagues (1993) other serious medical conditions were listed in their table but were not explained. Strauss and associates (1999) explained that the identification of severe medical conditions was from a separate listing of a “severe” identification on the CDER itself next to the disease code number. 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 (Eyman et al., 1990, 1993; Strauss et al., 1999) the presence of other serious medical conditions is specifically noted to have no impact on survival rates. These results are completely contrary to our findings (Plioplys et al., 1998a) and to common clinical experience. 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 comparison, we precisely defined a set of serious medical conditions and found a strong impact of these illness on survival rates (Plioplys et al., 1998a).

In one specific serious medical problem, that of refractory epilepsy, we found a markedly decreased survival rate in those who had more than one seizure per day (Plioplys et al., 1998a). Our results are in keeping with previous reports about epilepsy decreasing survival expectations in children with cerebral palsy (Crichton et al., 1995; Evans et al., 1990; Kudrjavcev et al., 1985; Roboz, 1972). Furthermore, in otherwise neurologically healthy individuals, epilepsy has been shown to decrease survival rates (Nilsson, Tomson, Farahmand, Diwan, & Persson, 1997; Shackleton, Westendorp, Trenite, & Vandenbroucke, 1999). 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 (Eyman et al., 1990, 1993; Strauss et al., 1999). It is only recently, in a study of mildly disabled individuals, that the California data has shown an impact of epilepsy on life expectancy (Strauss et al., 2003). The only possible explanation for this discrepancy is that the data used in the California reports are incomplete and/or inaccurate.

Given all of the previously enumerated difficulties with the California data, it is not surprising that Grossman and Eyman (1998) typified the clinical information collected as being “notoriously unreliable.” As they themselves explained, the information was collected for the purpose of financial reimbursement. Reimbursement was based solely on one section of the CDER that dealt with physical and cognitive impairment scales, not with the sections dealing with medical and neurologic information. Financial pressures easily overcome the need for careful and accurate filling out of lengthy and complex government forms. In the methods section of all of the published reports from California, the statistical reliability of the CDER is mentioned. However, in the cited reliability references, the statistical evaluation was determined only for the 66 questions concerning functional impairments (Harris, Eyman, & Mayeda, 1982). It is clear that the statistical reliability of the basic medical and neurologic information contained in the CDER has never been formally determined.

CONCLUSION

Although the number of cases in our study was relatively small (447; Plioplys et al., 1998a), 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 me). 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.

In one of their publications, Strauss and Shavelle (1998c) argue quite correctly that life insurance companies, in providing policies, rely equally on the reports of actuaries and physicians. Life expectancy estimates vary greatly, depending on the medical illnesses that the applicant may have, and the ascertainment of such illnesses is the role of the physician. For researchers who conduct epidemiologic studies and make 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.

Our published data (Plioplys et al., 1998a) has given much better survival rates for severely disabled children than those previously reported. The reasons for the improved survival were discussed, and compared
particularly to the California data. The major factors improving 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. In light of progressive governmental and private payor budgetary restrictions and the 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.

REFERENCES

Pediatric Skilled Nursing Facilities


