SURVIVAL RATES OF CHILDREN WITH SEVERE NEUROLOGIC DISABILITIES IN PEDIATRIC SKILLED NURSING FACILITIES

AUDRIUS V. PLIOPLYS MD, FRCPC, FAAP, CMD

DIVISION OF NEUROLOGY
MERCY HOSPITAL
2600 S. MICHIGAN AVE.
CHICAGO, ILLINOIS 60616
708-445-5060
FAX: 773-445-0123

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SURVIVAL RATES OF CHILDREN WITH SEVERE NEUROLOGIC DISABILITIES
A COMPARISON OF PUBLISHED DATA

Audrius V. Plioplys MD, FRCPC, FAAP, CMD; Chicago, IL

Introduction Accurate survival rates of children with neurologic disabilities is important in planning future medical expenses. This is of particular importance to pediatric skilled nursing facilities (SNF's) that depend on financial support from governmental sources. Eyman (1) published extremely pessimistic survival rates. This led to a review of our clinical results, which were published (2). Our results were much more optimistic than those reported by Eyman. Recently, several reports have appeared using Eyman's California database. This investigation compares our results with recently published ones.

Results Our survival rate data (2) is consistently better than recent reports. 5-year survival rates for the most disabled group 1 were: Eyman (1) 16%, Strauss (3) 43%, and Plioplys (2) 75%. In another report, 10-year survival rates for group 1 were: Eyman (1) 32%, Strauss (4) 45%, and Plioplys (2) 73%. In the most recent report, 8-year survival rates for group 1 were: Eyman (1) 38%, Strauss (5) 63%, and Plioplys (2) 73%.

Discussion The reasons for our better survival rates are multiple, including the fact that all of our patients were in SNF's where prompt medical care for acute illnesses was always provided (only 3% in California were in SNF's). Also, there were many methodologic and statistical errors contained in the California database and reports, which will be fully reviewed.

Conclusion Our survival rates continue to be much more optimistic and accurate than those using the California database.

1. Eyman et al. 1993. American Journal Diseases of Childhood, 147, 329-336
2. Plioplys et al. 1998. Southern Medical Journal, 91, 161-172
3. Strauss et al. 1997. Journal of Pediatrics, 131, 712-717
4. Strauss et al. 1998. Pediatric Neurology, 18, 143-149
5. Strauss et al. 2000. Pediatric Neurology, 23, 312-319

NEUROLOGIC DIAGNOSES (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 congenital syndromes	2.5%
Progressive CNS degenerative diseases	3.6%
Other conditions	5.8%
TOTAL	100%

FREQUENCY OF DISABILITIES

Cerebral palsy (onset before 28 days old)	83%
Profound mental retardation	93%
Epilepsy	64%
Gastrostomy tube feedings	52%
Nasogastric tube feedings	9%
Tracheostomy	11%
Immobile	79%
Unable to roll	69%
No hand or arm movement	45%
Incontinent	98%

CLINICAL GROUP DEFINITIONS

ALL WITH CEREBRAL PALSY, IMMOBILITY, PROFOUND MENTAL RETARDATION, INCONTINENCE

Group 1	tube fed not rolling no hand / arm use	Group2	tube fed not rolling hand / arm use present
Group 3	fed by others not rolling no hand / arm use	Group 4	fed by others not rolling hand / arm use present
Group 5	fed by others able to roll hand / arm use present	Group 6	tube fed able to roll hand / arm use present

Groups as defined by Eyman et al. 1993. Amer J Dis Child, 147, 329-336.

OTHER SIGNIFICANT MEDICAL DISEASES

Two or more bouts of pneumonia per year

Refractory seizures

Recurrent bowel obstructions

Severe asthma

Cardiac arrhythmia

Progressive cardiac failure

As defined by Plioplys et al.1998. Southern Medical Journal, 91, 161-172.

8-YEAR SURVIVAL RATES

Less than 1 year of age

Group number	Eyman (1993)	Plioplys(1998)	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

Between 1 and 15 years of age

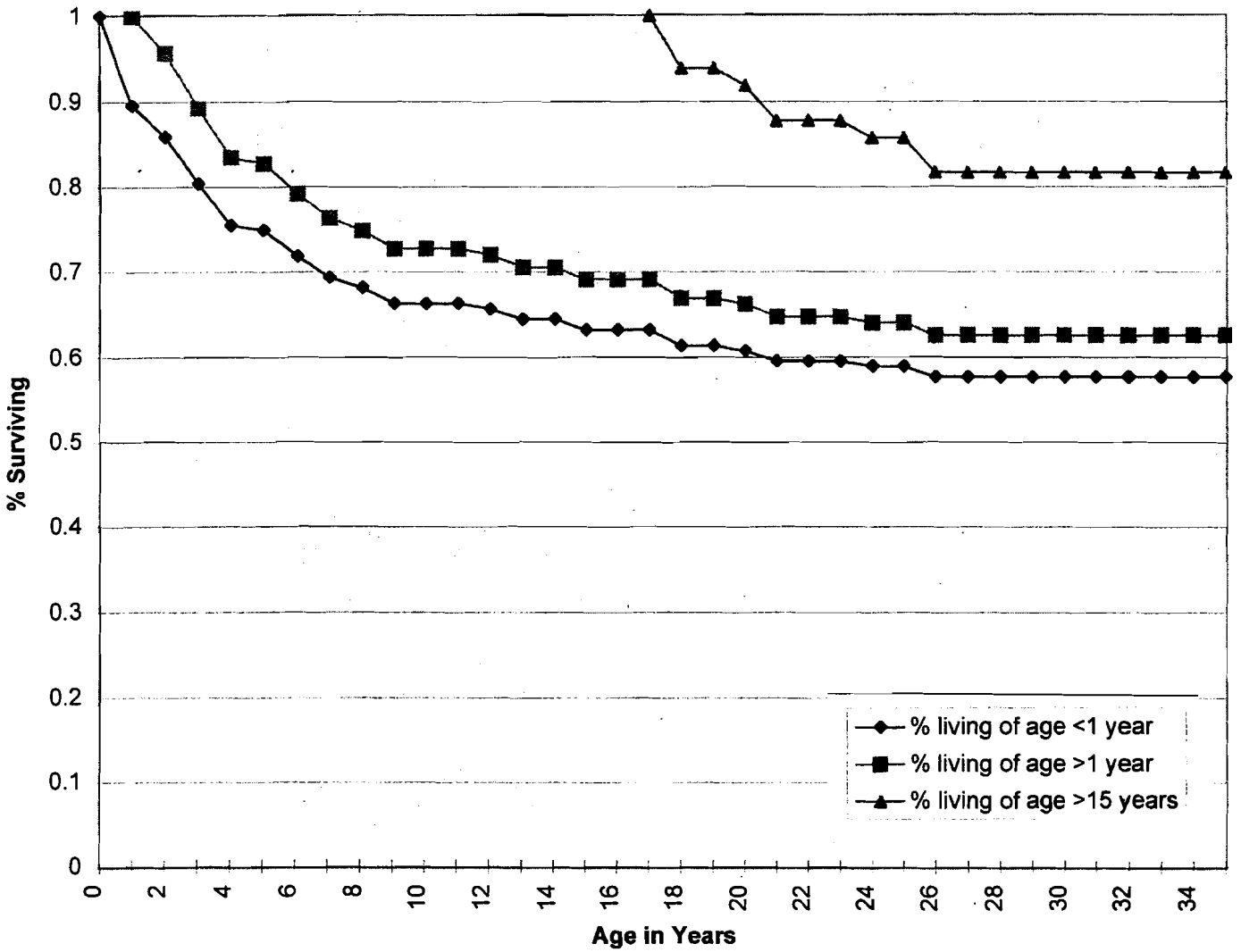
1	35%	73%	< 0.001
2	38%	95%	< 0.001
3	40%	94%	< 0.001
4	60%	91%	ns
5	80%	100%	ns
6	70%	100%	ns

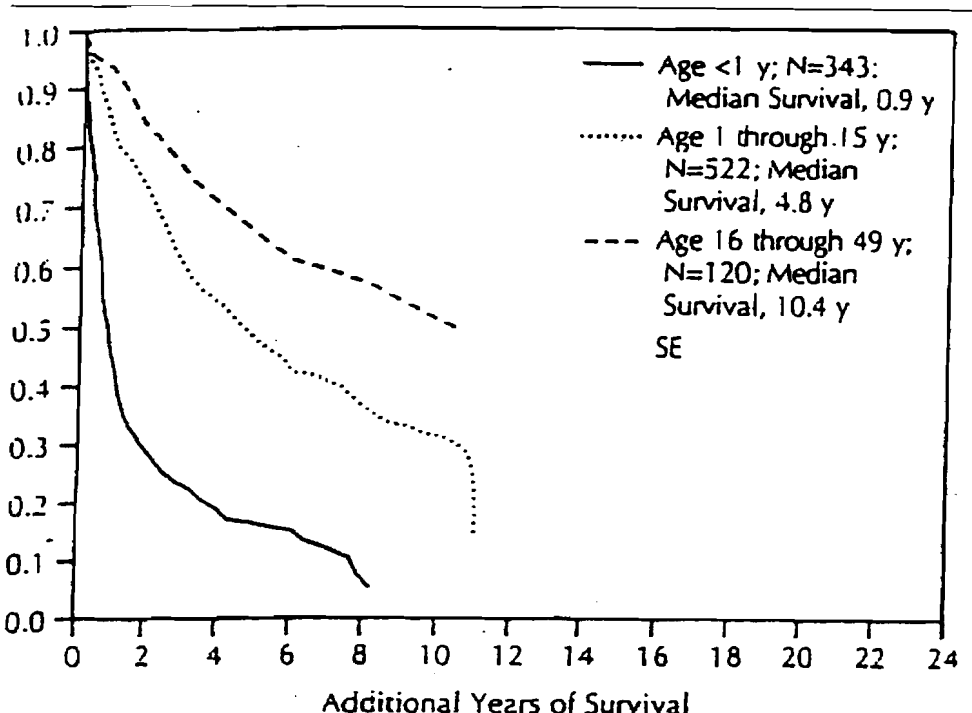
Older than 15 years of age

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

Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.
Plioplys et al.1998. Southern Medical Journal, 91, 161-172.

GROUP 1: SURVIVAL RATES



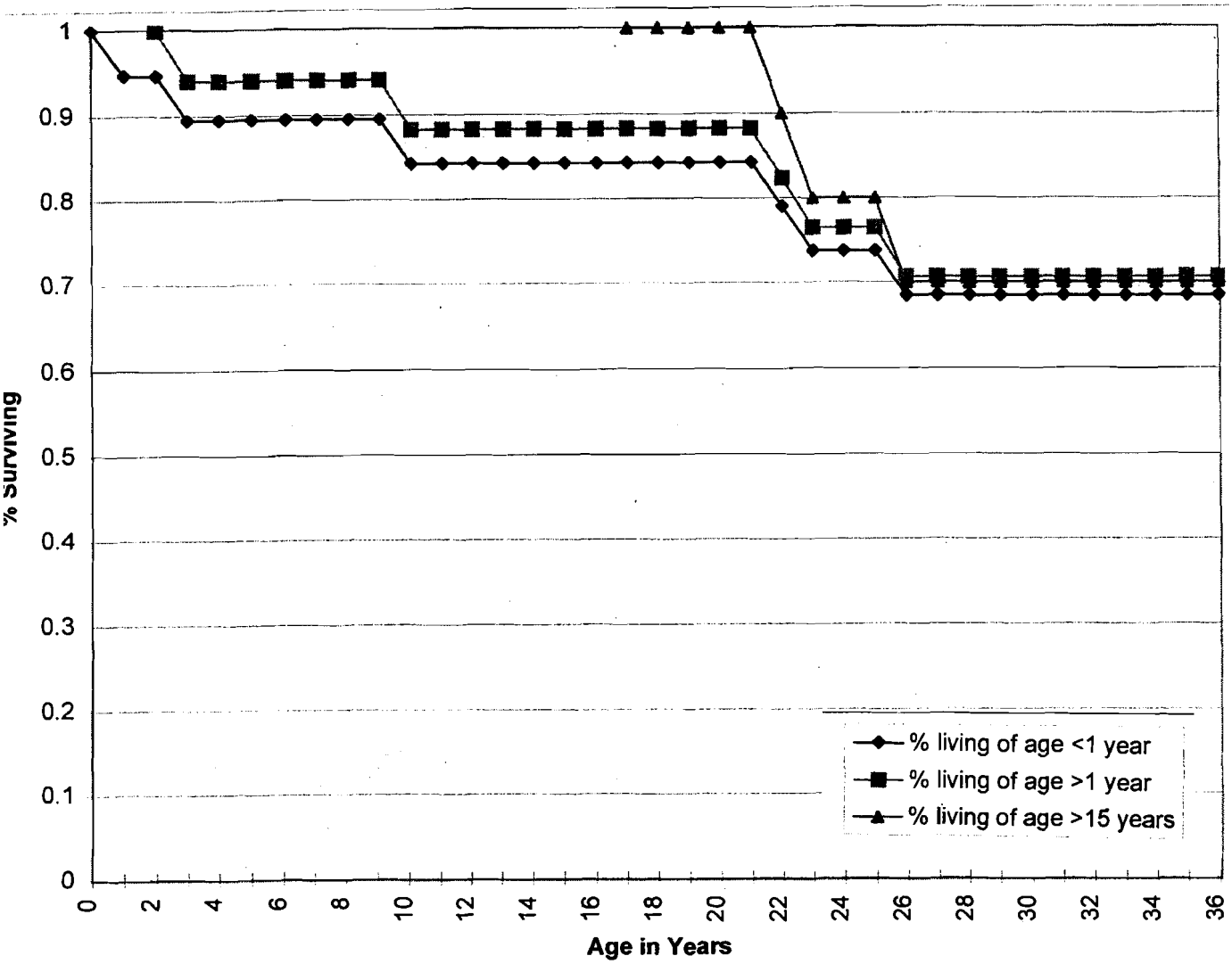


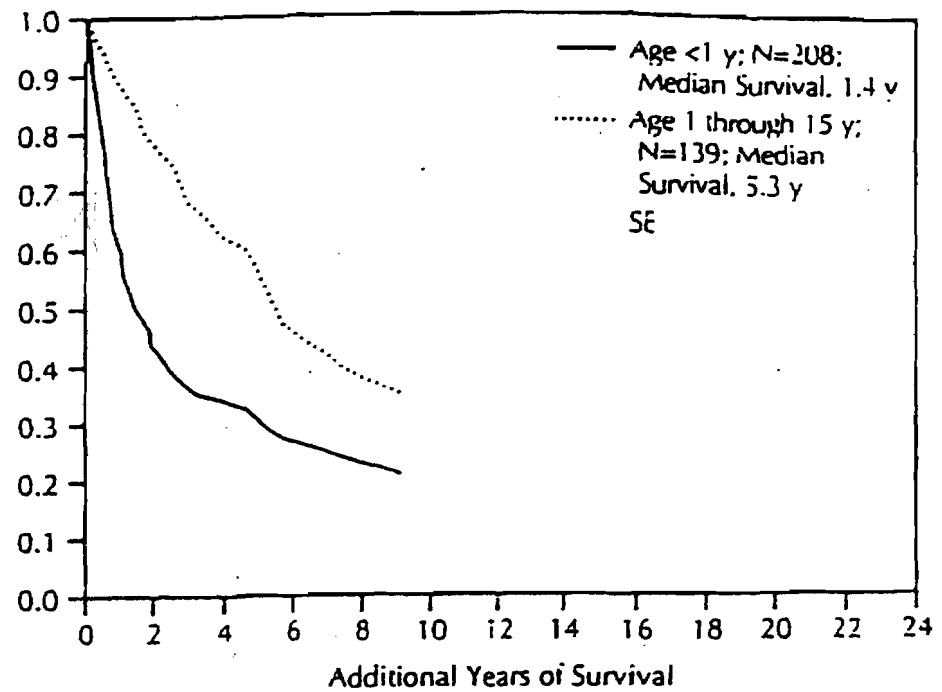
The graph above is from Plioplys et al. 1998. Southern Medical Journal, 91, 161-172.

The graph to the left is from Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.

Note that the X-axises of these two graphs overlap identically.

GROUP 2: SURVIVAL RATES





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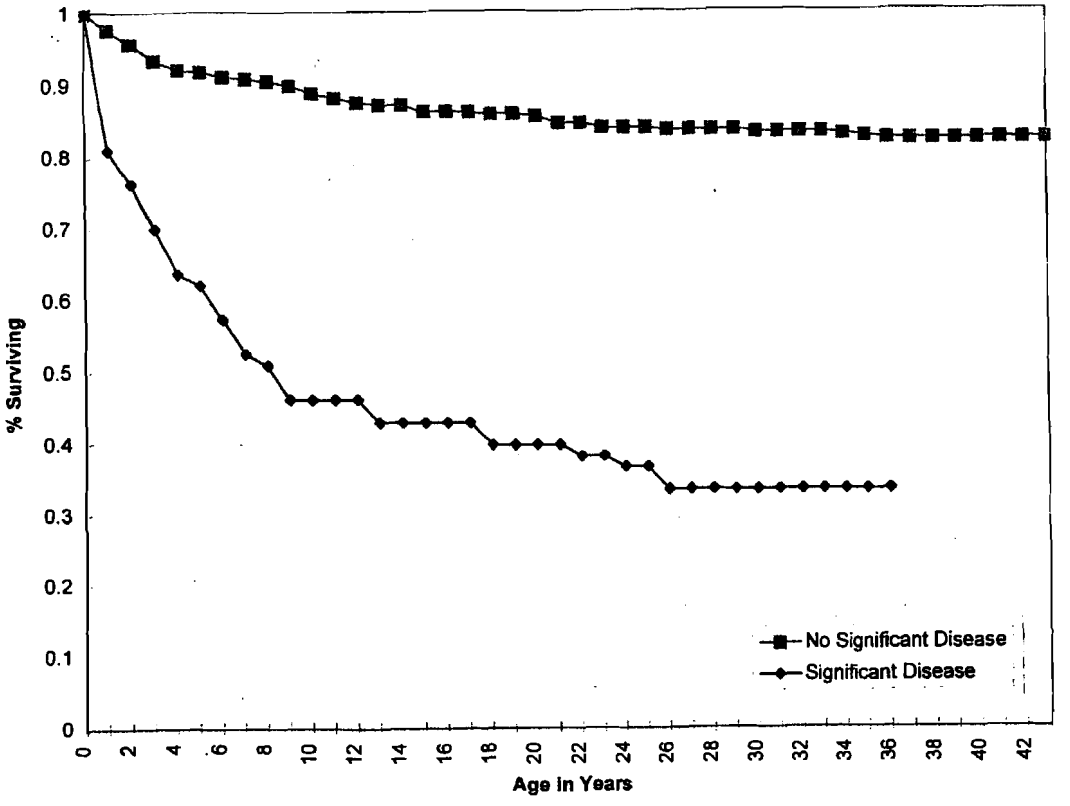
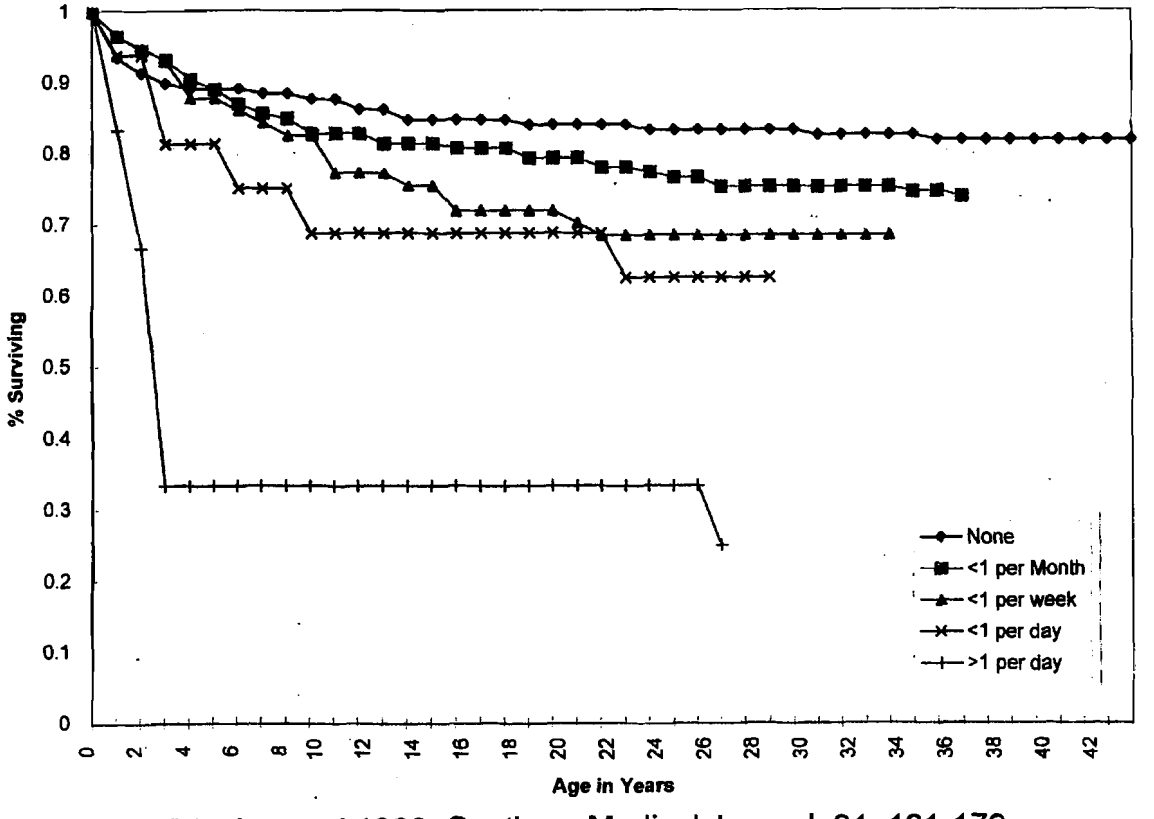
The graph to the left is from Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.

Note that the X-axises of these two graphs overlap identically.

SURVIVAL: OTHER SIGNIFICANT DISEASES



SURVIVAL: FREQUENCY OF SEIZURES



Plioplys et al.1998. Southern Medical Journal, 91, 161-172.

SURVIVAL FACTORS:

OTHER SIGNIFICANT MEDICAL ILLNESSES

18% had other significant diseases, of which

66% recurrent pneumonias

18% refractory seizures

7% cardiac failure

5% recurrent bowel obstruction

4% severe asthma.

10-year survival rates

90% if no other significant illness

45% if with other significant illness ($p < 0.0001$)

Our study (Plioplys et al. 1998) was the first to show that survival rates of disabled children is significantly reduced by a defined set of medical diseases. As a corollary, severely disabled children, who are otherwise healthy, have much better survival rates.

SURVIVAL FACTORS: MOBILITY

10-year survival rates

immobility: 76%;

any mobility: 96% ($p < 0.001$).

inablity to roll: 74%;

any rolling: 98% ($p < 0.0001$).

Decreased survival rates with immobility (Plioplys et al. 1998) is in keeping with many other published reports.

FACTORS: MENTAL RETARDATION

10-year survival rates

100% moderate mental retardation

95% severe mental retardation

81% profound mental retardation

Decreased survival rates with profound mental retardation (Plioplys et al. 1998) is in keeping with many published reports.

FACTORS: EPILEPSY

10-year survival rates

87% if no seizures

83% if <1 seizure / week

68% if between 1 seizure / day and 1 / week

33% if >1 seizure / day ($p < 0.001$)

Decreased survival rates with epilepsy (Plioplys et al. 1998) is in keeping with many published reports.

SURVIVAL FACTORS: FEEDING

10-year survival rates

95% for self-feeders

95% orally fed by others

78% G-tube fed ($p < 0.001$)

41% nasogastric tube fed ($p < 0.0001$)

10-year survival rates with no significant disease

86% for G-tube fed

56% for nasogastric tube fed ($p < 0.01$)

10-year rates for those who had other significant diseases

49% for G-tube

18% for nasogastric tube

Decreased survival rates with tube feeding
(Plioplys et al. 1998) is in keeping with other published reports.

FACTORS: TRACHEOSTOMY

10-year survival rates

83% no tracheostomy

75% with tracheostomy ($p = ns$)

No significant effect of tracheostomy on survival rates
(Plioplys et al. 1998) has been noted in one other study.

MORE RECENT COMPARISONS-1

5-YEAR SURVIVAL RATES

Less than 1 year of age

	Group 1	Group 2	Group 3
Eyman (1993)	16%	30%	25%
Strauss (1997)	43%	62%	77%
Plioplys (1998)	75%	89%	91%

Over 1 year of age

	Group 1	Group 2	Group 3
Eyman (1993)	50%	60%	55%
Strauss (1997)	55%	69%	69%
Plioplys (1998)	82%	94%	93%

Note that the groupings are approximate

Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.
Plioplys et al.1998. Southern Medical Journal, 91, 161-172.
Strauss et al. 1997. Journal of Pediatrics, 131, 712-7.

MORE RECENT COMPARISONS-2

10-YEAR SURVIVAL RATES

	Group 1	Group 2	Group 3
Eyman (1993)	32%	36%	37%
Strauss (1998)	45%	62%	70%
Plioplys (1998)	73%	85%	90%

	Group 4	Group 5	Group 6
Eyman (1993)	50%	78%	68%
Strauss (1998)	80%, 87%	85%	94%
Plioplys (1998)	91%	100%	100%

Note that the groupings are approximate.

Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.
Plioplys et al.1998. Southern Medical Journal, 91, 161-172.
Strauss et al. 1998. Pediatric Neurology, 18, 143-149.

8-YEAR SURVIVAL RATES

	Group 1	Group 2
Eyman (1993)	38%	40%
Strauss (2000)	63%, 65%	81%
Plioplys (1998)	73%	94%

Note that the groupings are approximate.

Eyman et al. 1993. American Journal of Diseases of Childhood, 147, 329-336.
Plioplys et al.1998. Southern Medical Journal, 91, 161-172.
Strauss et al. 2000. Pediatric Neurology, 23, 312-319.

COMPARISON OF SURVIVAL RATE RESULTS: INTERNATIONAL STUDIES

Our survival rate results (Plioplys et al. 1998) cannot be compared directly to most of the previous studies about the survival rates of children with cerebral palsy. 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 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 et al. (1993) was to define groupings of neurologic severity such that subsequent investigators, such as ourselves, could collect comparative data.

Survival rate studies:

Blair et al. 2001. Developmental Medicine and Child Neurology, 43, 508-515.

Crichton et al. 1995. Developmental Medicine and Child Neurology, 37, 567-576.

Evans et al. 1990. Archives of Diseases of Childhood, 65, 1329-1333.

Hutton et al. 1994. British Medical Journal, 309, 431-435.

Hutton et al. 2000. Archives of Diseases of Childhood, 83, 468-474.

Hutton et al. 2002. . Archives Diseases of Childhood, 86, 84-90.

Kudrjavcev et al. 1985. Neurology, 35, 900-903.

Patja et al. 2000. Journal of Intellectual Disability Research, 44, 591-599.

Roboz. 1972. Medical Journal of Australia, 1, 218-221.

Williams & Alberman. 1998. Developmental Medicine and Child Neurology, 40, 376-379.

WHY THE DIFFERENCE IN RESULTS?

1998 EXPLANATIONS

CHANGE IN PUBLIC ATTITUDE

Eyman's data: 1980-1991 decade

Our study: 1985-1996 decade

Medical advances from 1980 to 1985

More important: shift in public attitudes towards disabled individuals

Before 1985, G-tubes and tracheostomies were almost not in use

Analogous to Down syndrome 10 year survival:

1944-1955: 37%

1966-1975: 86%

UNIFORMITY OF MEDICAL CARE

Our study: Under uniform direction for 10 years

Eyman data: 21 regional centers across California, diverse medical care

ACCESS TO MEDICAL CARE

All of out patients in a skilled nursing facility (SNF)

Registered nurses available in-house, 24 hours per day

Acute illnesses addressed promptly

Eyman data: 65% at home; only 3.5% in a SNF

Home or group home placement, compared to state institutions (not SNF's) results in 25% greater mortality rate in all groups of severity of disability.

Strauss et al. 1996. Am J Pub Health, 86, 1422-1429.

WHY THE DIFFERENCE IN RESULTS?

MORE RECENT EXPLANATIONS

CRITIQUE OF EYMAN REPORTS: **DELETION OF DATA**

Strauss (1997) explained the statistical methods used by Eyman as: "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."

Subsequently, Strauss elaborated further (Strauss & Shavelle. 1998): "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 as a "methodologic mistake" (Strauss & Shavelle. 1998) 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." In the methods section of the 1993 report, Eyman wrote: "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."

Grossman & Eyman. 1998. Pediatric Neurology, 19, 243.

Strauss. 1997. Lancet, 349, 283-284.

Strauss. 2001. www.lifeexpectancy.com/eyman.html.

Strauss & Shavelle. 1998. Pediatric Neurology, 19, 243-244.

METHODOLOGIC DIFFICULTIES IN THE CALIFORNIA DATABASE

UNRELIABILITY OF DATA

Grossman and Eyman (1998) stated that the clinical information in the database about cause of disability and other 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". One example of unreliability is that the California data cannot distinguish between G-tube and nasogastric tube feedings.

In the 1993 Eyman report, the presence or absence of epilepsy could not be determined in **36%**. Epilepsy is an integral part of a person's neurologic disability, which should be easily ascertainable.

In a report of adults with developmental disabilities (Strauss & Shavelle, 1998), there was no basic neurologic diagnosis in **79%** of cases.

In a study of children with cerebral palsy (Strauss & Shavelle, 1998b), **82%** carried no identifiable neurologic diagnosis for their disability.

In a more recent report, dealing with severely neurologically disabled children from 1988 to 1997 (Strauss et al. 2000) **40%** had no identifiable neurologic diagnosis. Thus, the problem lies not with the time period from which the data was collected, but rather in the data collecting system itself.

Grossman & Eyman. 1998. Pediatric Neurology, 19, 243.
Srauss & Shavelle. 1998. Developmental Medicine and Child Neurology, 40, 369-375.
Strauss & Shavelle. 1998b. Pediatric Neurology, 19, 243-244.
Strauss et al. 2000. Pediatric Neurology, 23, 312-319.

METHODOLOGIC DIFFICULTIES IN THE CALIFORNIA DATABASE

OTHER SIGNIFICANT MEDICAL ILLNESSES

We found that other serious medical illnesses are important predictors of survival rates (Plioplys et al. 1998). Eyman (1990) also paid attention to this issue and in the client development evaluation report (CDER) defined a number of serious medical conditions as: "diabetes, heart disease, chronic respiratory infection, or hepatitis." Strauss 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, makes the use of this designation at best entirely unreliable, and at worst, completely capricious.

It is thus not surprising that in most reports from California serious medical conditions are specifically noted to have no impact on survival rates. These results are completely contrary to our findings (Plioplys et al. 1998) and to common clinical experience. A disabled child who has very frequent pneumonias, cannot have a life expectancy equivalent to that of a child who is otherwise healthy.

In refractory epilepsy, we found markedly decreased survival rates in those who had more than one seizure per day (Plioplys et al. 1998). Our results are in keeping with very many studies reporting decreased survival rates in patients with epilepsy, with or without cerebral palsy.

Given all of these previous results, it is surprising to repeatedly find that studies from California report that epilepsy and the frequency of seizures have no impact on survival rates. The only possible explanation is that the California data is incomplete and / or inaccurate.

COMPARISON OF CALIFORNIA DATA TO WESTERN AUSTRALIA

There is a recent claim that survival rate data from California (Shavelle et al. 2001) is similar to Western Australia (Blair et al. 2001). By Blair's definition of degrees of severity, the "severe" group simply was not able to ambulate. In Eyman's 1993 report, the studied number of cases corresponding to Blair's "severe" category, and with an IQ of less than 20, was 3,157. This was a minimum number of "severe" cases within the California database: many cases that did not fit into groups 1 to 6 would still have been "severe" by Blair.

Since 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, would be **over 6,000** cases. A California report about adults with cerebral palsy revealed **8,093** severe cases, and **6,939** severe cases with profound mental retardation (Strauss & Shavelle, 1998). The available numbers are large. However, the report claiming similar outcomes was based **only on 974** cases, suggesting that a highly selected set of cases from the California database was studied.

Our own survival rate results (Plioplys et al., 1998) vary greatly with the degree of disability. 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 cannot be accepted as presented. The best explanation lies in a careful preselection of cases compared.

Blair et al. 2001. Developmental Medicine and Child Neurology, 43, 508-515.

Shavelle et al. 2001. Developmental Medicine and Child Neurology, 43, 574.

Srauss, & Shavelle. 1998. Developmental Medicine and Child Neurology, 40, 369-375.

CONCLUSION

Although the number of cases in our study was relatively small (447; Plioplys et al. 1998), all of the cases were thoroughly examined, and their records reviewed by medical personelle with many years of experience taking care of neurologically disabled children (the Directors of Nursing of the SNF's, and a Board Certified Child Neurologist). 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 (1998) argue quite correctly that life 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 applicant may have, and the ascertainment of such illnesses is the role of the physician.

In conducting epidemiological studies and making life expectancy predictions for disabled children, accurate medical information is just as necessary. Unfortunately, the California database, that has generated so many studies and reports, incorporated neither accurate nor adequate medical information.

Plioplys et al. 1998. Southern Medical Journal, 91, 161-172.

Strauss & Shavelle. 1998. Expert Witness, 3, 11-13.