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Life Expectancy of Children With Cerebral Palsy

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Risk factors for mortality of young children with cerebral palsy were studied using a sample of 12,709 children aged 0.5-3.5 years with cerebral palsy who had received services from the State of California between 1980 and 1995. The most powerful prognostic factors for survival were simple functional items: mobility and feeding skills. Once these were known, factors such as severity of mental retardation and presence of quadriplegia contributed relatively little. Children with fair motor and eating skills had good survival prospects, with 90% or more reaching adulthood, but those without such skills had much poorer prospects. Among children who were unable to lift their heads, median survival time was 7 additional years for those who were tube fed (n = 557) and 14 years for those fed entirely by others (n = 997). Although a child's approximate survival chances can be assessed from such functional classifications, we indicate the manner in which additional information on the child's condition can be used to obtain more accurate survival data.

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Introduction

In the United States there is no system for monitoring the prevalence of cerebral palsy (CP), and little information on life expectancy is available [1]. Such data are valuable for counseling parents, planning for school, employment, and other needs, and for medicolegal purposes [2,3]. The prognosis is related to the child's level of functioning and other risk factors [2-7]. Good assessment of the role of the various risk factors, and good prognoses based on them, requires both a very large sample of children with CP and data on a wide variety of potential risk factors. To date, the largest study is that of Chrichton et al. [3], who studied 3,189 persons with CP with respect to some six risk factors.

The present study was based on 12,709 infants with CP and data on some 200 potential risk factors. The sample consisted of all such children who received any services from the State of California between 1980 and 1995. The primary goals of the study were as follows:

(1) To assess which factors were important to a child's prognosis for survival. For example, quadriplegia and severe mental retardation have been cited as factors associated with the most marked reduction of life expectancy [1]. On the basis of our previous work [8-10], however, we hypothesized that these factors, together with cognitive and etiologic data, are less important than simple functional skills such as mobility and eating.
(2) To use the key factors identified in goal (1) to classify the children into cohorts and to assess the survival prospects for each cohort.
(3) To demonstrate how more detailed information about a given child can be used to obtain improved survival data specific to that child.

Methods

Instruments. Our base population was the 182,263 persons with developmental disability who received services from the State of California between January 1980 and December 1995. Services may include medical treatment, occupational or physical therapy, and board and care. All such persons are evaluated approximately annually, using the Client Development Evaluation Report (CDER) [11]. This instrument contains some 200 psychologic, medical, functional, behavioral, and cognitive items. The reliability of the functional items has been assessed previously and was judged as being satisfactory [12-15]. Interrater reliabilities of the motoric and eating skill variables used in this study exceeded 0.85 [14].

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Table 1. Characteristics and associated hazard ratios of children with cerebral palsy (CP) (n = 12,709 children, 1,248 deaths)

	Number of Children	Number of Deaths	Hazard Ratio*
Age at first CP evaluation			
7-18 months	4,953	686	2.00†
19-30 months	3,635	311	1.18‡
31–42 months	4,131	251	1.00
Sex			
Male	7,149	683	0.95
Female	5,570	565	1.00
Rolling and sitting§			
Cannot lift head when lying on stomach	1,571	531	13.91†
Lifts head, not chest	1,521	295	7.05†
Partial rolling ability	1,921	211	3.67†
Full rolling; may be able to sit independently	7,706	211	1.00
Hand use‖			
No functional use	2,561	721	5.69†
Some functional use	10,158	527	1.00
Ambulation			
Does not walk	9,275	1,208	12.90†
Walks with support	1,368	24	1.83
Walks unsteadily alone at least 10 feet	944	6	0.71
Walks well alone at least 20 feet	1,132	10	1.00
Feeding			
Tube fed	1,060	394	23.65†
Fed by others, no feed tube	5,679	734	6.18†
Some self-feeding skill	5,980	120	1.00
Severity of CP			
Mild	1,975	60	1.00
Moderate	4,939	277	1.90†
Severe	3,637	630	5.84†
Unspecified	2,168	281	3.62†
Location of CP			
Monoplegia	193	13	0.88
Hemiplegia	1,700	42	0.25†
Diplegia	1,609	52	0.34†
Triplegia	151	6	0.38‡
Paraplegia	531	34	0.68‡
Quadriplegia	6,863	924	1.40†
Unspecified	1,672	177	1.00
Type of dysfunction			
Spasticity	6,708	646	1.00
Ataxia	1,054	102	0.86
Dyskinesis	467	43	0.79
Hypotonia	1,983	195	1.00
Other	2,507	262	1.00
Severity of mental retardation¶			
None, mild, moderate	7,760	453	1.00
Severe, profound, unspecified#	4,959	795	3.11†
Year of first CP evaluation			
1980-1985	4,767	633	1.00
1986-1990	4,301	444	1.08
1991-1995	3,651	171	0.90
Receptive nonverbal communication			
Does not appear to understand gestures/facial expressions	6,444	1,067	5.61†
Understands simple gestures ("yes," "no," pointing)	4,954	148	1.05
Understands complex gestures	1,321	33	1.00
Low birth weight/decreased gestational age			
Yes	2,512	154	0.80†
No	10,207	1,094	1.00

* Ratio of risk for each category relative to the referent category (which has a hazard ratio of 1.00 by convention). The ratio was based on a proportional hazards survival analysis (see Statistical Methods), with no other factors taken into account.

† Significantly different from 1.00 at $P < 0.01$.

‡ Significantly different from 1.00 at $P < 0.05$.

§ Condensed from the 9-point Client Development Evaluation Report scale, as explained in the text.

‖ Condensed from the 4-point Client Development Evaluation Report scale.

¶ Categories for severity of retardation are as in Grossman [17].

Unspecified is a California term for those whose IQ has not been formally assessed. Such children are generally severely impaired.

CP was assessed by an evaluation team contracted by the California Department of Developmental Services as part of initial evaluation for receiving services. We used three CDER items to identify subjects with CP. These were severity, type, and location of CP; definitions will be given shortly. We defined a CDER evaluation as being one of CP if all three items indicated presence of CP. All three interitem reliabilities exceeded 0.999.

Mortality information was obtained from annual computer tapes from the California Department of Health Services.

Subjects. We extracted data on all children aged older than 6 months and younger than 3 years 6 months with an evaluation of CP. CDER evaluations of children younger than 6 months were not used because functional data at such ages would be difficult to interpret. Older children, with a first evaluation of CP after age 3.5 years, will be addressed elsewhere. This resulted in a sample of 13,378 children. Coded ICD-9 etiologic information [16] was given in the CDER for some children, although its reliability is not known. Numbers of cases were as follows: infections (ICD-9 code 000-139; n = 116), circulatory disorders (390-459; n = 163), congenital anomalies (740-759; n = 1,158), ill-defined (799; n = 2,554), injuries/poisonings (800-999; n = 179), other etiologic codes (n = 100), and no code specified (n = 8,449).

Frequently it is the initial diagnosis of CP that brings the child into the state disability system. Consequently, many children were older at the date of first CDER evaluation than at the first diagnosis of CP. Undoubtedly, some children diagnosed with CP died before entering the system. This occurrence does not introduce a bias into the study, however, because we measured survival time only from the time of first CP evaluation. This required us to include age at first CP evaluation as a potential risk factor.

Because CP is generally considered a congenital or perinatal condition, we excluded the 659 children listed as having suffered a major accident, such as near-drowning or motor-vehicle related.[1] This left a sample of 12,709 children, of whom 1,248 died during the 1980-1995 study period.

Variables. Table 1 [17] shows some of the major variables considered in the study, and it also indicates the "simple" hazard ratios associated with each level (these can be interpreted as relative mortality rates, unadjusted for the effects of other factors). Infants with a CDER evaluation of CP at age 7-18 months had twice the hazard rate as those entering 1 or 2 years later. This finding was not surprising because these children were generally more seriously disabled. There was the expected excess of boys; the hazard rate did not vary with sex.

Mobility was strongly associated with risk, with children unable to lift their heads being at 14 times the risk of those who could roll or sit. Similarly, complete inability to walk was a major risk factor. The 1,060 children who were tube fed were at more than 23 times the risk of those with some self-feeding skills. By contrast, type, severity, and location of CP appeared to have a much smaller effect on survival. Table 1 also shows one of the many CDER cognitive items—receptive nonverbal communication. Children lacking such skill were at substantially higher risk.

Statistical Methods. The Cox proportional hazards survival model [18] was used for initial screening of risk factors and for combining categories. CDER functional items, which are scored on scales of 4 to 9 levels, were collapsed when adjacent levels were not associated with significantly different mortality risks. Survival time was measured from the date of first evaluation indicating CP. Tests for significant risk factors, their possible interactions, and the proportional hazards assumption were based on likelihood ratio tests [19] for nested factors and the Akaike Information Criterion [18] for all other cases. The large sample size enabled us to use a stringent 0.01 significance level for retaining risk factors.

The best predictive factors were used to classify the children into cohorts. Kaplan-Meier survival curves were constructed [18]. Median survival times were reported when these were less than the 16-year study period; for other, higher functioning cohorts, 5-, 10-, and 15-year survival probabilities were reported. Standard errors were based on the Greenwood formula [20]. All computations were carried out using SAS [21], with S-PLUS [22] used for graphics. Estimation of a survival curve for a child with a specific profile of risk factors was based on standard procedures for the Cox model [18].

Table 2. Proportional hazards model for mortality predictors (all effects significant at $P < 0.01$)

Variable	Hazard Ratio*	95% Confidence Interval	
		Lower	Upper
Cannot lift head†	7.95	4.54	13.92
Lifts head but not chest†	6.26	3.59	10.91
Partial rolling ability†	4.36	2.51	7.58
Rolls/sits, but cannot walk unaided†	2.40	1.41	4.09
Tube fed,‡ age 1	5.14	3.89	6.80
Tube fed,‡ ages 2 and 3	3.85	2.88	5.14
Fed by others, not tube fed‡	2.01	1.59	2.54
No functional hand use	1.52	1.31	1.76
Severe or profound mental retardation§	1.39	1.22	1.58
"Severe" cerebral palsy‖	1.32	1.15	1.52
Quadriplegia¶	1.27	1.04	1.53
Hypotonia#	1.26	1.07	1.47

* Mortality risk, compared with the referent group, with other factors held constant. For example, in any short time interval, other things being equal, children unable to lift their heads were 7.95 times more likely to die than those able to walk 10 feet unaided.

† Compared with referent group (children able to roll/sit and able to walk 10 feet or more unaided).

‡ Children with gastrostomy tubes; the referent group is composed of those with some self-feeding skills.

§ Referent group consists of those with mild or moderate mental retardation.

‖ Referent group consists of those whose condition is judged as being mild or moderate.

¶ Referent group consists of those without involvement of all four limbs.

\# Referent group consists of those with spasticity, ataxia, dyskinesis, and mixed forms.

Results

The proportional hazards model presented in Table 2 shows the factors contributing significantly to the prediction of mortality. The first four rows contrast four levels of reduced mobility with the referent group ("can roll and sit, and walk 10 feet without assistance"). The risk ratio of 7.95 in the first row indicates that, other things being equal, children unable to lift their heads when lying on their stomachs had about 8 times greater mortality risk than those in the referent group. For a short interval, such as 1 year, this means a roughly 8 times larger chance of dying [18].

The second row of Table 2 shows that the risk for children who can lift their heads but not their chests is

[1] Interestingly, such children proved to have a better survival than the others, even after adjustment for risk factors; we hope to report on this issue in detail elsewhere.

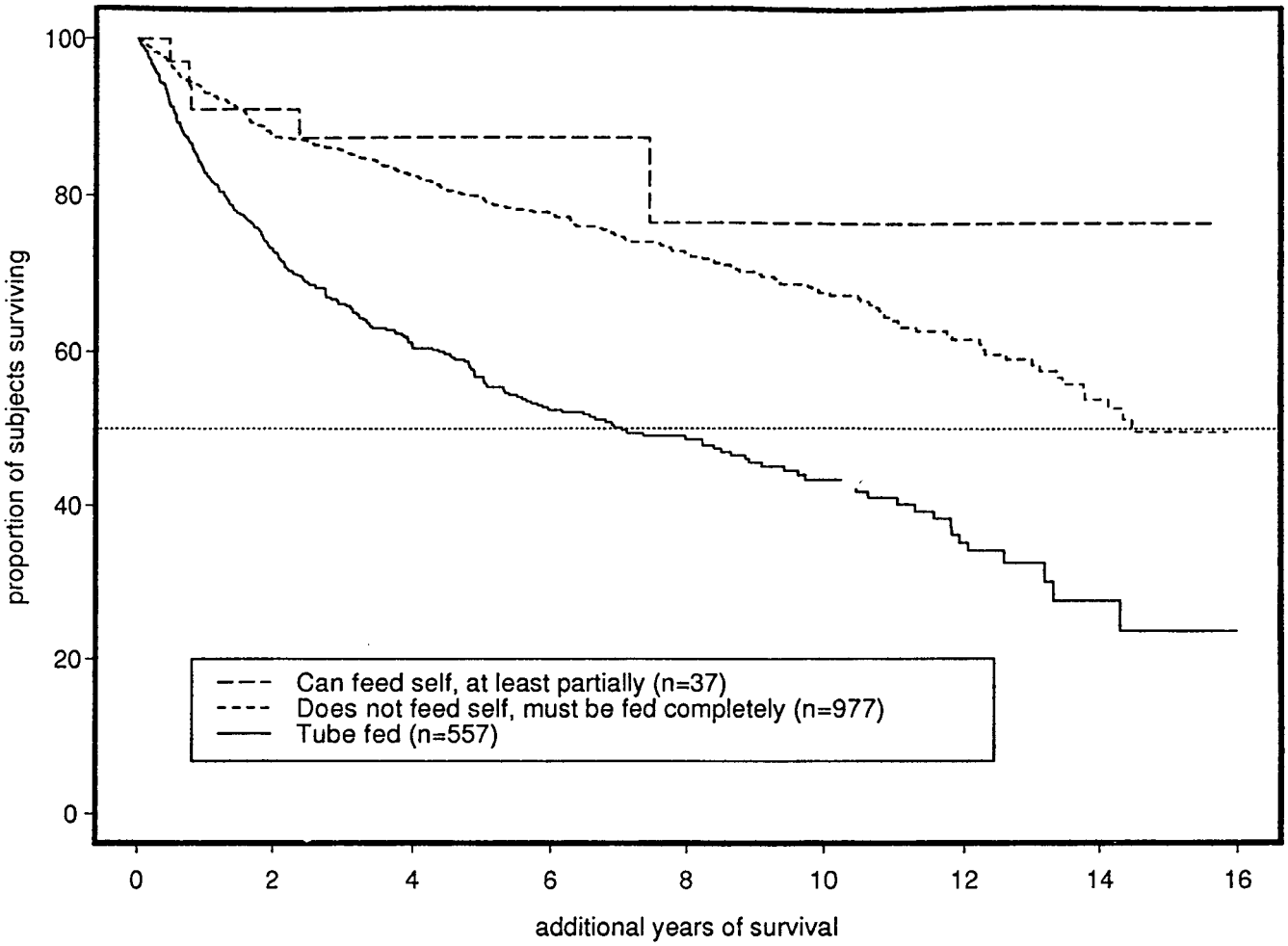

Figure 1. Additional survival time for children unable to lift their heads when lying on their stomachs.

6.36, again relative to the referent group. The risk ratios of 4.36 and 2.40 correspond, respectively, to those who have limited rolling ability and to those who can roll and sit but cannot walk unaided. These results demonstrate that immobility is a very powerful predictor of mortality in this population.

For the feeding variable, the referent group consists of those with at least some self-feeding ability, if only with fingers. The increased risk associated with tube feeding is 5.14 for children entering the system at age 1 year and 3.85 for those entering at ages 2 and 3. The larger risk for the younger children probably reflects the extreme neurologic impairment of the young children who require gastrostomy feeding. Being fed by others doubles the risk, regardless of age at entry. There were no other significant effects related to age.

As a measure of the predictive power [23] of the simple model with just motor and feeding items, this model (with 6 degrees of freedom) accounts for 92% of the likelihood ratio achieved by the model based on all the variables in Table 1 (37 degrees of freedom).

Other factors making a significant, albeit smaller, contribution to the prediction are as follows:

(1) Lack of hand use increases risk by 52% (compared with otherwise comparable children with some hand use).
(2) Presence versus absence of epilepsy accounts for a 46% increase.[2]
(3) Severe or profound mental retardation (compared with mild or moderate retardation) results in a 39% increase.
(4) Severe (compared with mild or moderate) CP accounts for a 32% increase.
(5) Quadriplegia (compared with monoplegia, diplegia, paraplegia, etc.) causes a 27% increase.
(6) Hypotonia (compared with spasticity, ataxia, dyskinesis, etc.) results in a 26% increase.

[2] Relevant data on epilepsy were available only after 1986. We therefore assessed its effect with a special analysis (not shown) of post-1986 data. For technical reasons, the epilepsy variable has not been included in the main analysis of Table 2. This proved not to have a noticeable effect on the other risk factors.

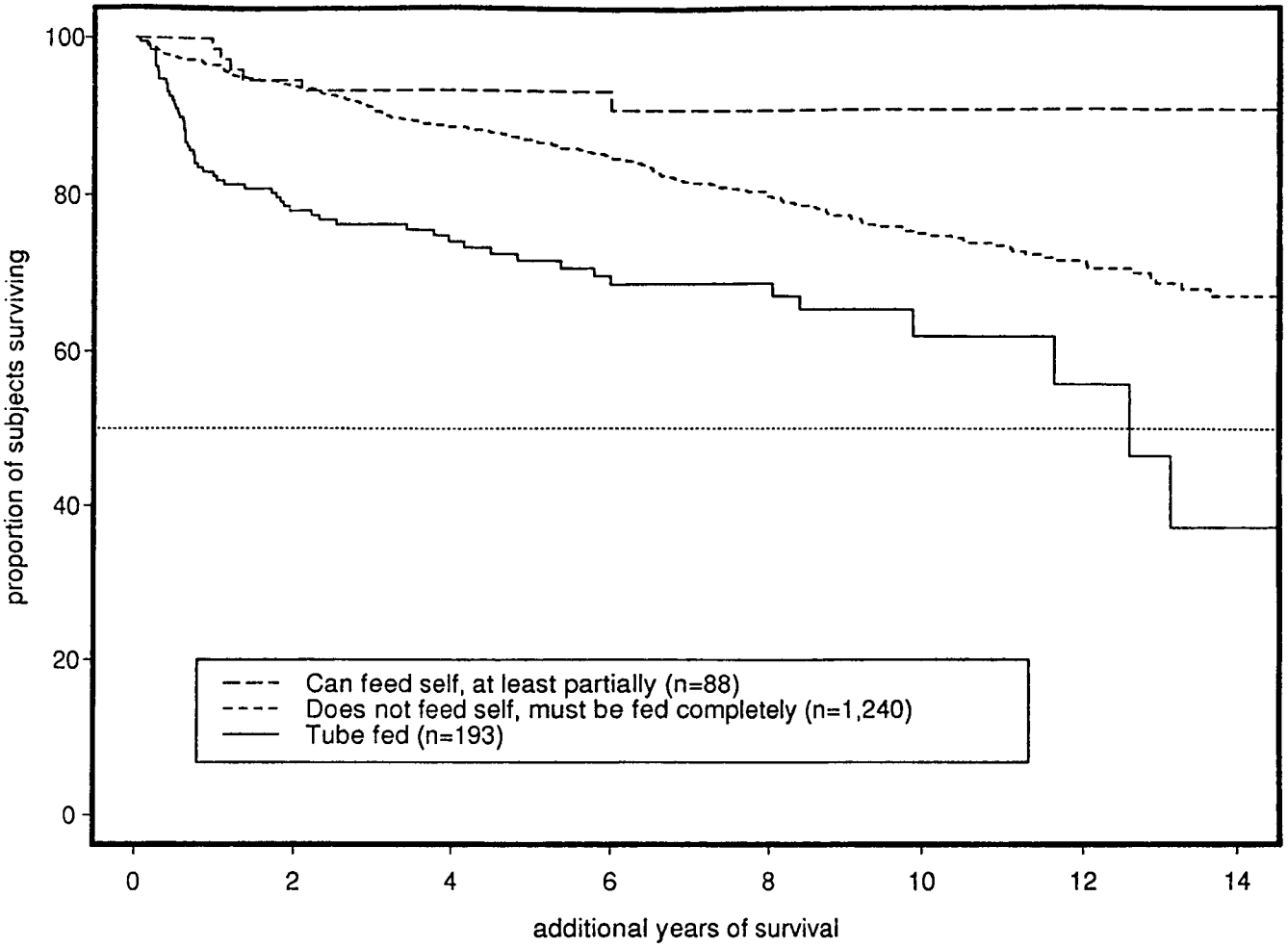

Figure 2. Additional survival time for children able to lift their heads but not their chests when lying on their stomachs.

Several variables that seemed from Table 1 to be mortality predictors are conspicuously absent from Table 2. Apart from presence or absence of quadriplegia or hypotonia, further data on type and location of CP failed to contribute to a prognosis. Other factors not contributing significantly were cognitive skills, sex, low birth weight, teen pregnancy, maternal age older than 35, and age at entry into the system.

Children entering later in the study period seemed to be at slightly lower risk than those entering earlier, but this secular trend was not statistically significant. Etiologic group also had no significant effect on mortality after the above factors were taken into account.

Stratifying by motor and feeding skills, we obtained 15 cohorts of children (5 motor × 3 feeding skills). Figure 1 shows subsequent survival for children unable to lift their heads at first CP evaluation, stratified according to feeding skill. Median survival time for the tube-fed children (n = 557) was 7 additional years, and it was 14 years for those without gastrostomies but who were fed completely by others (n = 997). Figure 2, for children able to lift their heads but not their chests, shows a more optimistic prognosis, but even so only 50% of those in the tube-fed subgroup survived beyond 12 years.

Children with more mobility (Table 3) had better prospects: the majority of children in any of these groups survive to adulthood. In the higher functioning groups, more than 90% live at least 15 more years.

The information in Figures 1 and 2 or Table 3 is only useful for a preliminary approximation of the survival chances. A more accurate prognosis for a child with a given profile of the variables in Table 2 is obtained from a survival curve specific to that profile. This may be obtained directly from a Cox model. The procedure uses more information about the child's condition than Figures 1 and 2 or Table 3. As an example, one of the cohorts in Figure 1 consists of children fed by others and unable to lift their heads when lying on their stomachs. According to the survival curve (not shown here) based on the Cox model, the median survival time is 14.9 additional years. If in addition it is known that the child is quadriplegic, lacks hand use, and has severe CP and mental retardation, however, the new survival curve indicates a median survival time of only 11.8 years.

Table 3. Survival probabilities and their associated 95% confidence intervals (expressed as percentages) for cohorts with at least some mobility

	n	Survival Probability		
		5 Years	10 Years	15 Years
Lifts head and chest, partial rolling				
Tube fed	136	73% (65,81)	62 (51,72)	62 (51,72)
Fed by others	1,403	93% (91,94)	87 (84,89)	79 (75,83)
Some self-feeding	382	98% (97,99)	95 (92,98)	91 (84,97)
Full rolling, does not walk unaided				
Tube fed	156	89% (84,94)	85 (78,92)	80 (69,91)
Fed by others	1,936	97% (96,97)	94 (92,95)	90 (88,93)
Some self-feeding	3,553	98% (98,99)	97 (96,98)	95 (94,97)
Walks 10 feet unaided				
Tube fed	18	89% (74,100)	—* (—,—)*	—* (—,—)*
Fed by others	123	98% (94,100)	98 (94,100)	91 (78,100)
Some self-feeding	1,920	100% (99,100)	99 (98,100)	99 (98,100)

* Sample size too small to compute.

Discussion

This study of survival of children with CP was based on a much larger sample and examined more potential risk factors than earlier ones. This enabled us to identify several new findings.

The most debilitated children in this study, with greatly reduced mobility and need for gastrostomy feeding, have a poorer prognosis than has been reported [2,3,6], the lowest functioning group having a median survival of only 7 additional years. The higher functioning groups, however, have a much more favorable prognosis.

It has been noted that functional variables are the best predictors of survival [2,6,24,25]. We have found that simple, easily measured motor and feeding skills convey most of the information. It had been suggested [26] that cognitive skills (e.g., receptive nonverbal communication), which are important in establishing bonds with parents, might play an appreciable role in determining the survival of these severely compromised children, even after mobility and other factors are taken into account. This hypothesis is not supported by the present data. Furthermore, etiology, low birth weight and gestational age, sex, cognitive skills, and type and location of CP also carry little additional information. Even the presence of quadriplegia increases the risk only slightly.

Concerning low birth weight and gestation, Hutton et al. [2] found as we did (Table 1) that children with this history had better survival rates than those without it. We found, however, that such children had on average slightly better mobility. Because low birth weight had no predictive effect on survival when other factors were taken into account, the association may have reflected confounding with the more important motor skill variables.

Comparison with the earlier studies by Eyman [7,25] on the same California database may be of interest. First, their studies combined persons with CP with accident victims, persons with autism, and other groups. Second, as discussed elsewhere, these studies also excluded persons whose condition improved [27]. Unless one is sure that the child's condition will not change, this approach is unsuitable for estimation of life expectancies. Finally, the present work was based on 5 more years of monitoring and used multivariate methods to generate a more refined grouping of the subjects into cohorts.[3]

Two limitations on the use of the cohort analyses of Figures 1 and 2 and Table 3 should be noted; these remarks apply equally to the results of other studies [2,3,6,7,10,25]. First, the analyses are based on only a small number of characteristics (mobility and feeding) and should be used only as a preliminary approximation. More reliable prognoses result if one also uses data on variables contributing to the multivariate model (Table 2). The methodology, a standard feature of proportional hazards analysis, was illustrated in the previous section. Second, the cohort analyses should not be used to obtain, for example, the prognosis of a tube-fed, immobile child aged 7 years. The reason is that, of the cohort of children in this condition at entry to the study, many of those surviving to age 7 will no longer be in the same condition. Again, it is preferable to fit a new proportional hazards model, in this case specifically for 7 year olds.

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[3] As pointed out by Hutton et al. [2], the Eyman studies (and the present one) use a state database that includes virtually all the most severely impaired persons but excludes many persons whose disability is mild enough that they have not sought state services. If such persons had been included in the study, the sizes of the less involved cohorts (Table 3) would have increased markedly. It appears unlikely, however, that the more severely impaired cohorts would have been materially affected.

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