Causes of excess mortality in cerebral palsy

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It is well known that individuals with cerebral palsy (CP) are subject to higher mortality than the general population but the causes of this have not been systematically analysed. This study investigates mortality in a Californian population of 45 292 individuals with CP, 4028 of whom died during the 1986 to 1995 study period. The aims of this study were to identify diseases that may be causally linked with CP, and diseases whose diagnosis and/or treatment need improvement. Overall, the standardized mortality ratio (SMR) was 8.4. Mortality from breast cancer was three times that of the general population, suggesting poorer detection and/or treatment. The dramatic elevation of mortality due to brain cancer, especially in children (SMR=24), raises the possibility of a link between this and CP. Cause of death was non-specific in some individuals. Therefore, these SMRs are conservative. As expected, SMRs due to respiratory diseases were very high but, contrary to anecdotal reports, such diseases did not account for most deaths. High SMRs were also found for diseases of the circulatory and digestive systems. Finally, a marked elevation of deaths was due to external causes, including drowning and being hit by motor vehicles.

Cerebral palsy (CP) has been described as '... a term of convenience applied to a group of motor disorders of central origin... As generally understood there must be motor impairment, and this impairment must stem from a malfunction of the brain (rather than spinal cord or muscles) .... [the brain malfunction] must be non-progressive and it must be manifest early in life' (p 520) (Badawi et al. 1998). There is substantial literature on mortality and life expectancy of individuals with CP (see Evans et al. 1990, Hutton et al. 1994, Chrichton et al. 1995, Strauss 1997, Strauss and Shavelle 1998a, Strauss et al. 1998a). Life expectancy is shorter than in the general population, especially when the CP is accompanied by severe disabilities. Respiratory diseases are often considered the main cause of death in CP, but there have been few studies of cause-specific mortality.

In a recent investigation of individuals with developmental disability in London, Hollins and coworkers (1998) reported that 52% of the deaths were due to respiratory disease. Plioplys and coworkers (1998) found 77% of the deaths to be due to pneumonia in their population (mostly children) with very severe neurological disabilities. Jancar and Speller (1994) observed high death rates due to intestinal obstruction, especially volvulus. A study in California, USA of externally caused deaths among individuals with developmental disabilities found elevated mortality due to drowning and pedestrian versus motor-vehicle accidents (Strauss et al. 1998b).

The present study was undertaken to compare mortality rates in CP with those in the general population. The large database available (45 292 subjects, 4028 of whom died, with information on the cause of death on a computerized database) permitted detailed cause-specific comparisons. The aims were as follows: to identify diseases for further investigation that may be causally linked with CP; to quantify cause-specific mortality rates so that the effect on life expectancy of eliminating certain avoidable causes may be assessed; and to identify causes of death whose elevated rates relative to the general population may reflect the need for better diagnosis and/or treatment.

Our approach was epidemiological rather than clinical, and the work should be viewed as a stimulus and guide to more in-depth investigations.

Method

SUBJECTS

Our base population comprised 182 263 individuals with developmental disability who received services from the State of California, USA between January 1980 and December 1995. Services include medical treatment, occupational or physical therapy, and accommodation and care. All such individuals are evaluated approximately annually, using the Client Development Evaluation Report (CDER; California Department of Developmental Services 1986). This instrument contains some 200 psychological, medical, functional, behavioral, and cognitive items. The reliability of the functional items has been assessed previously and judged satisfactory (Harris et al. 1982, Arias et al. 1983, Widaman 1984, Widaman et al. 1985, Citygate 1998). Interrater reliabilities (Agresti 1990) of the motoric and feeding variables described in the present study exceeded 0.85.

The study period was the 10-year interval 1986 to 1995. For the purposes of this study, individuals with CP were considered at risk from the date of the first CDER evaluation or 1 January 1986, whichever came later, until the earliest of the

following: the date of death, the end of the study period (31 December 1995), or 3 years after the date of the subject's last CDER evaluation. This last condition was included to minimize the potential bias due to subjects who left California. Deaths of such individuals would not be in our records, but because of the last condition these subjects would also not be counted as being at risk for more than a fairly short period. Migration from California is generally believed to be uncommon because California is the only US state providing services to individuals with developmental disability as an entitlement.

CP was assessed by an evaluation team contracted by the California Department of Developmental Services as part of the initial work-up for receiving services. We used three CDER items to identify subjects with CP: severity, type, and location of CP. Definitions are given below. A CDER evaluation was defined as being one of CP if all three items indicated presence of CP. All three interitem reliabilities exceeded 0.99. We identified 45 292 individuals with CP.

It seems likely that only a small proportion of individuals in the study acquired their disability in the postneonatal period. For example, of the subjects who were born after 1980, so that they could have had an evaluation of CP in infancy, 84% did in fact have such an evaluation before the age of 3 years, and presumably almost all of these children had congenital or neonatal defects. Of the others, some origins are doubtless congenital but their families moved into California later, perhaps to take advantage of the state's entitlement to services for CP, and some entered the California system only after attending school, when their condition became a disability. Nevertheless, some children undoubtedly had insults after the neonatal condition, and their recorded cause of death is sometimes also the etiology of the dysfunction. Unfortunately, we can only identify the etiology (near-drowning, motor-vehicle accidents and so on) for a very small proportion of the subjects.

Mortality information was obtained from annual computer tapes from the California Department of Health Services, and matched against the subjects with CP on the basis of name, date of birth, and social-security number when available. We identified 4028 subjects who died during the study period. Causes of death are given on the computer tapes in the form of ICD-9 codes (Context Software Systems, Inc., 1995).

Data are available only on individuals receiving services from the state, therefore, the sample is weighted towards the more severely involved subjects. There is, for example, a higher proportion of individuals with quadriplegia and a lower proportion with hemiplegia than in many studies. This underscores the need to take account of functional level when making comparisons.

For each cause of death we computed the age- and sex-specific mortality rates (deaths per 100 000 person-years of exposure) in the general Californian population, using the mortality tapes. We then applied these rates to the age- and sex-specific exposure times for individuals with CP. This yielded an expected number of deaths due to each cause, on the basis of the general population. Each ratio of observed to expected numbers of deaths gives the standardized mortality ratio (SMR) (Kahn and Sempos 1989). By using the same source of mortality information when comparing the group with CP with the general Californian population we attempted to minimize various sources of reporting bias. Further, by using the midpoint of the study period (the year 1990) for calculating the age-, sex-, and cause-specific mortality rates in the general population, our comparisons relate to approximately the same calendar years. This should minimize bias due to various cause-specific secular trends.

Results were subsequently stratified into five age groups: <5 years, 5 to 14 years, 15 to 34 years, 35 to 54 years, and ≥ 55 years. After some preliminary analysis we also chose to stratify subjects into two groups on the basis of the severity of impact of CP: severe and not severe. Severe CP is defined as a 'condition so substantial that it is exceedingly difficult to find an appropriate placement for the client and/or constant care/supervision is required' (CDER 1986). Our results on cause-specific mortality are broken down into 10 (5 × 2) categories. In the tables that follow, the results of significance tests of the SMRs are reported (against the null hypothesis that the death rate is the same as in the general population). It was considered inappropriate to carry out such tests separately for each age group. For brevity, age groups were combined when numbers were judged to be too small to be informative.

Results

Table I shows the demographics and functional characteristics of the 45 292 subjects with CP. The impact of CP was reported as 'severe' for 14 655 subjects (32%). The other categories (mild, 22%; moderate, 33%; unspecified, 13%) were combined into a 'not severe' group.

Table I shows the usual excess of males. Regarding age distribution, it should be noted that many subjects enter the study long after they are identified as having CP. For example, subjects born before 1986, the beginning of the study period, could not contribute to the study until at least 1986. Not surprisingly, most (77%) of the subjects with severe CP impact had quadriplegia. The corresponding figure for those not severely impacted was 34%.

The instrument (CDER 1986) also includes functional items, some of which are shown in Table I. Each item is scaled from low to high level of skill, and the subjects' highest level is reported. The table shows, for example, that only 24% of the severe group, compared with 70% of those not severely affected, could assume and maintain a sitting position independently. A similar pattern may be seen in the other functional items.

Table II presents the aggregated mortality statistics (i.e. not broken down by cause of death). As expected, the SMRs for the severe group are much larger than those for the not severe group in every age category. The decrease in SMRs with age reflects both a healthy survivor effect and the increased mortality risk with age in the general population. Even in the oldest group, however, mortality was more than double that of the general population.

Table III shows the causes of death due to various cancers. In each age group the total number of cancer deaths is substantially higher than expected in the general population, as indicated by the significantly large SMRs. Cancers of the digestive system are elevated in both groups. The very small SMR (0.2; $P<0.001$) for lung cancer is noteworthy; this presumably reflects the low rate of smoking among individuals with CP, and thus represents one more piece of evidence on the effect of smoking. By contrast, breast cancer deaths were three times higher than expected.

Eighteen children aged under 15 years died from brain cancer, compared with 0.68 expected in the general population (SMR=26). These 18 deaths account for 44% of all cancer deaths in children under the age of 15 years in this study. The

Table I: Demographic and functional characteristics of the 45 292 Californian subjects with cerebral palsy, 1986–1995

	Impact of CP	
	Not severe	*Severe*
Sample size	30 637	14 655
Sex (%)		
Female	44.5	45.
Male	55.5	54.1
Ethnicity (%)		
White	51.7	51.3
Hispanic	22.7	24.2
Black	9.1	8.5
Other	16.5	16.0
Age at first evaluation (y) (%)		
<1	7.8	5.7
1–2	18.9	16.8
3–4	11.7	11.3
5–9	10.3	15.1
10–14	8.5	11.6
15–19	10.8	11.8
20–24	10.2	8.5
25–29	6.9	5.8
30–39	8.7	7.3
40–49	3.4	3.1
≥ 50	2.8	2.9
Type of CP (%)		
Spastic	33.4	40.2
Ataxic	31.5	32.4
Dyskinetic	10.3	12.8
Hypotonic	8.2	5.2
Other/unspecified	16.6	9.4
Location of CP (%)		
Monoplegia	2.8	0.5
Hemiplegia	21.6	4.9
Diplegia	13.0	5.9
Triplegia	1.3	1.9
Paraplegia	6.4	4.8
Quadriplegia	34.4	77.4
Other/unspecified	20.5	4.6
Rolling and sitting (%)		
Cannot lift head	4.8	18.8
Lifts head not chest	4.7	13.5
Lifts head and chest	1.9	4.6
Rolls	9.2	20.3
Partial sitting	9.8	18.9
Assumes and maintains sitting position independently	69.7	24.0
Hand use (%)		
No functional use	8.3	29.1
Raking motion or grasps	20.4	37.0
Uses thumb and fingers	18.0	14.7
Uses fingers independently	53.3	19.3
Ambulation (%)		
Does not walk	32.7	80.3
Walk with support	11.8	12.0
Walks alone, 10 ft	15.2	4.1
Walks alone, 20 ft, balances well	40.3	3.6
Tube fed (%)		
No	96.9	89.0
Yes	3.1	11.0
Self-feeding (%)		
Does not feed self	21.1	54.9
Attempts to finger feed	5.5	8.5
Finger feeds	7.9	5.5
Uses spoon, spillage	18.6	13.7
Spoon and fork, spillage	19.4	9.3
Uses utensils, no spillage	27.5	8.2
Self-dressing (%)		
Does not put on clothes	31.7	64.4
Cooperates	20.4	20.3
Puts on some clothes	10.8	5.9
Puts on all clothes	15.9	4.9
Dresses completely, including buttons	21.1	4.5
Receptive language (%)		
Does not understand speech	19.3	36.5
Understands simple words	17.8	20.9
Simple phrases	25.1	16.5
Simple conversations	24.8	13.6
Understands story plots and complex conversations	13.0	12.6
Expressive language (%)		
Makes no sound	10.7	24.2
Babbles, no words	27.9	39.2
Simple words	14.6	10.9
Two-word sentences	6.9	3.7
Sentences of three or more words	11.8	5.7
Basic conversation	18.3	8.8
Complex conversation	9.9	7.5
Mental retardation level* (%)		
None/unspecified	33.4	27.4
Mild	24.2	8.9
Moderate	15.8	8.4
Severe	11.8	16.3
Profound	14.9	39.1

* UK usage – learning disabilities.

20 brain-cancer deaths in adults is also more than expected on the basis of the general population, although the corresponding SMR (6.1) is much lower than for the children.

Data on respiratory diseases are shown in Table IV. The very large SMRs for these causes, especially in the group with severe CP impact, is as anticipated. Nevertheless, the overall proportions of deaths due to respiratory causes, 11%, 11%, and 15% in the respective age groups 0 to 4 years, 5 to 14 years, and ≥ 15 years, were much lower than is generally thought.

Table V summarizes data on diseases of circulation. Not surprisingly, most deaths occurred in the oldest groups, although the 53 deaths in the 0 to 14 years group is significantly greater than that expected in the general population (SMR=60). There was substantial excess mortality due to most kinds of circulatory disease, including ischemic heart disease and cerebrovascular disease.

Individuals with CP are also at substantially greater mortality risk from diseases of the digestive system (Table VI). The increased risks due to intestinal obstruction and to esophageal diseases are well known, though they have not been previously quantified. Interestingly, the nine adult deaths due to chronic liver disease and cirrhosis, code 571, was less than the expected number in the general population (12.6), presumably reflecting the low alcohol consumption of individuals with CP. There were only two adult deaths due to hepatitis (code 573) and none due to liver cancer (code 155), which can be a sequela of hepatitis. This does not support the common view that individuals with CP, and especially those living in large congregate settings, are at increased risk of contracting hepatitis.

Deaths due to external causes, such as accidents and injuries, are of particular interest because they are generally preventable. There were 334 such deaths. The largest subgroup (103 deaths) were coded as 'late effects of accidental injuries'. Of these, 63 occurred in the 5 to 14 age group. In some cases these accidents may actually have been the cause of the CP, though as noted in the introduction, CP is generally understood to be a disease that manifests itself early in life. We were not able to clarify this issue further.

Drowning accounted for 40 deaths, of which 12 occurred in infants aged <5 years (SMR = 11), 15 to children aged 5 to 14 years (SMR=25), and 13 aged ≥ 15 (SMR=5). Of the cases where the circumstances were indicated, most accidents occurred in swimming pools or quenching tanks (jacuzzi for one person), and the remainder in a bath tub.

Homicide accounted for 29 deaths, a rate comparable to the general population (SMR = 0.9). Four were due to 'child battering and other maltreatment by unspecified person'. The largest subgroup (14 deaths) corresponded to 'late effects of injury purposely inflicted by other person', and again it is possible that some of these incidents were the cause of CP.

Pedestrians hit by motor vehicles accounted for 17 deaths (SMR=2.8, P<0.01). All but two of the victims were in the 'not severe' group; this is not surprising because that group tends to be more ambulatory.*

Table II: All causes of mortality in the 45 292 Californians with CP, 1986–1995

Age group (y)	*Impact of CP*					
	Not severe			*Severe*		
	Obs[a]	*Exp*[b]	*SMR*[c]	*Obs*[a]	*Exp*[b]	*SMR*[c]
0–4	299	7.5	39.8	340	3.5	97.1
5–14	333	10.1	33.0	640	5.9	108.5
15–34	451	94.1	4.8	877	53.3	16.4
35–54	337	91.8	3.7	333	44.7	7.4
≥ 55	234	105.2	2.2	183	64.2	2.9
All ages	1655	308.8	5.4	2373	171.7	13.8

[a] Observed number of deaths.
[b] Expected number in general Californian population for same distribution by age and sex.
[c] Ratio of observed to expected. All are significantly >1.0 (P<0.001).

Table III: Cancer mortality (ICD-9 codes 140–239)

Age (y)/disease group	*Impact of CP*					
	Not severe			*Severe*		
	Obs[a]	*Exp*[b]	*SMR*[c]	*Obs*[a]	*Exp*[b]	*SMR*[c]
All cancer[d]						
0–4	9	0.6	16.4	7	0.3	26.9
5–14	14	1.5	9.5	11	0.9	12.6
15–34	30	5.8	5.1	20	3.3	6.1
35–54	44	18.2	2.4	28	9.1	3.1
≥ 55	24	31.0	0.8	15	18.3	0.8
Breast cancer, female (174) age ≥ 15 only[e]						
15–34 y	2	0.4	4.7	1	0.2	4.3
35–54 y	8	2.8	2.6	6	1.5	4.1
55+ y	5	2.4	2.1	5	1.6	3.2
Digestive organs (ICD-9 codes 150–159) age ≥ 15 only[e]						
Esophagus (150)	4	1.0	3.9	5	0.6	9.1
Colon (153)	6	3.7	1.6	6	2.1	2.9
Other (151,152, 154–159)	9	6.8	1.3	7	3.8	1.9
Trachea, bronchus, lung (162)[f]						
Age ≥ 15 only	2	13.6	0.2	2	7.6	0.3
Genitourinary (179–189)[e]						
Age ≥ 15 only	15	6.6	2.3	6	3.7	1.6
Brain cancer (191)[e,g]						
0–4	5	0.1	38.2	3	0.1	50.3
5–14	4	0.3	13.0	6	0.2	33.3
15–34	10	0.6	17.4	5	0.3	15.4
35–54	1	0.9	1.1	4	0.4	9.2
≥ 55	0	0.6	0.0	0	0.5	0.0

[a] Observed number of deaths due to indicated cause.
[b] Expected number in general California population the same distribution by age and sex.
[c] Ratio of observed to expected.
[d] SMRs for all causes of cancer SMRs were 2.5 for severe group, 2.1 for not-severe group (P< 0.001 in both cases).
[e] SMR for this disease (ages ≥ 15 pooled) is significantly >1 (P<0.01).
[f] SMR for this disease (ages ≥ 15 pooled) is significantly <1 (P<0.01).
[g] Most cases were classified as 'brain, unspecified' (code 191.9).

Table IV: Diseases of respiratory system (ICD-9 codes 460–519)

Age (y)/disease group	*Impact of CP*					
	Not severe			*Severe*		
	Obs[a]	*Exp*[b]	*SMR*[c]	*Obs*[a]	*Exp*[b]	*SMR*[c]
Pneumonia, organism unspecified (ICD-9 code 486)						
0–4	9	0.1	115.5	10	0.0	279.5
5–14	10	0.1	139.6	29	0.0	688.6
15–34	30	0.3	90.8	49	0.2	266.0
35–54	20	0.6	33.9	35	0.3	120.7
≥ 55	19	2.2	8.7	16	1.5	10.7
Other pneumonia and influenza (480–485, 487)						
0–4	11	0.1	91.7	10	0.1	200.0
5–14	5	0.1	83.3	13	0.0	325.0
15–34	15	0.4	13.5	38	0.2	54.3
35–54	11	0.4	27.5	8	0.3	32.0
≥ 55	5	0.5	10.6	5	0.2	23.8
Chronic obstructive pulmonary disease (490–496)						
0–4	1	0.0	100.0	4	0.0	400.0
5–14	1	0.1	7.1	11	0.1	137.5
15–34	4	0.4	10.8	19	0.2	79.2
35–54	3	1.0	3.0	13	0.5	27.1
≥ 55	7	5.2	1.3	9	3.2	2.8
All ages	16	6.9	2.3	56	4.1	13.8
Pneumonitis due to solids and liquids (507)						
All ages	14	0.2	58.3	26	0.1	185.7
All other respiratory diseases						
0–4	9	0.2	60.0	11	0.1	157.1
5–14	6	0.1	60.0	22	0.1	440.0
≥ 15	16	1.5	10.5	28	0.9	32.9

[a] Observed number of deaths due to indicated cause.
[b] Expected number in general California population for same distribution by age and sex.
[c] Ratio of observed to expected. All 10 disease-severity groupings gave SMRs significantly >1 (P<0.01).

* A computer printout cross-classifying all 4028 deaths by age group, severity of CP, and the full ICD-9 cause-of-death code (up to 5 digits) is available on request from the first author.

Discussion

This is the first study to quantify the cause-specific mortality risks associated with CP. Overall there is a marked increase in mortality compared with the general population. A notable exception is lung cancer, the SMR being only 0.2; this no doubt reflects the benefits of not smoking.

The generally elevated cancer mortality is striking. For example, death from breast cancer is three times more likely than in a group of comparable age and sex distribution in the general population. Unless there is a link between CP and breast cancer – which seems unlikely but which should not be dismissed – the increase suggests that diagnosis and/or treatment of the disease is worse in this group than in the general population. It may be that part of the increased risk of breast cancer reflects the nulliparity of most women with CP. This is unlikely to be the full explanation, however, because nulliparity has been estimated to increase the risk by no more than about 60% (Hsieh et al. 1994, Modan et al. 1998, Weiss et al. 1998). In a population with developmental disability and frequent difficulty in communication, delay in diagnosis of cancer may be common. The fact that the SMRs for all causes of cancer are higher in the 'severe' than in the 'not severe' groups for all five age classes is consistent with this. In-depth research on the issue would be valuable. For example, it would be helpful to compare individuals with CP and the general population with respect to the stage of cancer at diagnosis, subsequent type of treatment, and survival rates.

The high prevalence of brain cancer and the typically young age at death are striking. It is unlikely that these findings can be explained on the basis of differences in treatment. It is possible that some infantile brain tumors are misdiagnosed as CP, although this explanation is unlikely to account for higher mortality due to brain tumors in the ≥ 15 age group; as noted, CP is generally understood to be a disease that becomes manifest early in life. As indicated in Table III, most brain cancers were classified as 'brain, unspecified' (code 191.9). We are not aware that a causal link between brain cancer and CP has been suggested in the literature. We believe, however, that the possibility cannot be ruled out at this time, and that the statistical association between the two conditions requires further investigation.

Mortality due to diseases of the circulatory system were also greatly elevated, especially in the younger age groups. As noted by a reviewer, this may be at least partly explained by the reduced physical activity of individuals with CP.

Respiratory disease is known to be a leading cause of death among individuals with CP, and so the high mortality rates observed here are not unexpected. Indeed, the number of respiratory deaths reported here is perhaps lower than anticipated. Specifically, among individuals aged over 14 years there were 2416 deaths, of which only 367 (15%) were attributed to diseases of the respiratory system (ICD-9 codes 460 to 519). An additional 724 (30%) had ICD-9 codes that were uninformative for our purposes (for example, congenital anomalies, infantile CP, or ill-defined). An unknown proportion of the latter would be due to respiratory disease. We would, therefore, estimate that the proportion of adult deaths due to this cause to be in the range of 15% to 45%. This is noteworthy because respiratory infection has often been stated as the cause of death in most individuals with CP.

Table V: Diseases of circulatory system (ICD-9 codes 390–459)

Age (y)/disease group	*Impact of CP*					
	Not severe			*Severe*		
	Obs[a]	*Exp*[b]	*SMR*[c]	*Obs*[a]	*Exp*[b]	*SMR*[c]
Ischemic heart disease (410–414)						
0–34	5	0.9	5.5	5	0.4	13.5
35–54	16	7.3	2.2	16	3.6	3.6
≥ 55	45	20.6	2.2	23	12.8	1.8
Other heart disease (390–405, 415–429)						
0–34	35	2.6	13.5	31	1.4	21.8
35–54	26	4.6	5.7	16	2.3	7.1
≥ 55	24	8.2	2.9	15	5.0	3.3
Cerebrovascular disease (430–438)						
0–34	11	0.9	11.7	20	0.5	38.5
35–54	8	2.6	3.1	3	1.3	2.3
≥ 55	12	5.6	2.1	15	3.7	4.1
Other (440–459)						
all ages	12	2.9	4.1	10	1.7	5.8

[a] Observed number of deaths due to indicated cause.
[b] Expected number in the general Californian population for the same distribution by age and sex.
[c] Ratio of observed to expected. All eight disease-severity groupings gave SMRs significantly >1 (P<0.01).

Table VI: Diseases of the digestive system (ICD-9 codes 520–579); all ages combined

Age/disease group	*Impact of CP*					
	Not severe			*Severe*		
	Obs[a]	*Exp*[b]	*SMR*[c]	*Obs*[a]	*Exp*[b]	*SMR*[c]
Diseases of the esophagus (530)	10	0.2	55.6	29	0.1	290.0
Intestinal obstruction (560)	21	0.3	77.8	30	0.2	187.5
Other digestive system diseases	44	12.3	3.6	64	6.5	9.9

[a] Observed number of deaths due to indicated cause.
[b] Expected number in the general Californian population for the same distribution by age and sex.
[c] Ratio of observed to expected. All six disease-severity groupings gave SMRs significantly >1 (P<0.01).

Table VII: Underlying and contributory causes of death for 401 Californians aged 18 or older with CP[a]

Cause of death	*% where the cause is*	
	Underlying	*Underlying or contributory*
Cancer (ICD code 140–239)	15	18
Circulatory System (390–459)	46	75
Respiratory (460–519)	7	36
Digestive (520–579)	3	10
External (800–999)	2	8
Other	27[b]	100
	100	

[a] Source: National Health Interview Surveys, 1986–1994 (NHIS 1997).
[b] Includes 7% for whom CP is cited as the underlying cause.

We also found excess mortality due to certain external causes, such as pedestrian accidents and drowning. Similar results have been reported for other individuals with developmental disabilities (Strauss et al. 1998b). In a supposedly protected group of vulnerable individuals, such incidents ought be a rarity, and parents/caregivers should be aware of the elevated risks.

The results reported here can be used to adjust life expectancy estimates to reflect reductions in cause-specific mortality rates. Such reductions may be appropriate for a patient with exceptionally good medical care. As an example, we show the effect of halving the mortality risk due to respiratory disease for a 15-year-old male with CP and quadriplegia. Before reduction the life expectancy is 34.3 additional years, according to an actuarial analysis (Strauss and Shavelle 1998b). If we assume that 45% of all mortality in such patients is respiratory-related we have a 22.5% reduction in overall mortality. This leads to a revised life expectancy of 38.1 additional years, an increase of 3.8 years.

The SMRs reported here are actually likely to be conservative (i.e. underestimates). This is because a substantial number of deaths with uninformative codes, such as 'congenital anomalies', should really be ascribed to specific causes. Unfortunately we do not have access to multiple-cause-of-death data which can be linked to our own. Multiple-cause-of-death data without personal identifiers were, however, available from the 1986 to 1994 National Health Interview Surveys (NHIS 1997).

Using the operational definition of CP of Boyle and colleagues (1994), namely ICD-9 codes 333.2, 333.7, and 342 to 344, we identified the 401 individuals whose death certificate included a reference to CP. Table VII shows the prevalence of some of the principal diseases on the death certificates, and the frequency with which they were listed as the underlying cause. The table confirms, for example, that respiratory infection is by no means the most common cause in this population. The most common cause is disease of the circulatory system, just as in the general population. The data here, however, were derived only from adults; presumably the contribution of respiratory disease to infant mortality is greater.

Necessarily, large epidemiological studies such as this deal only with gross patterns, and leave many questions unanswered. They complement in-depth clinical observation rather than serve as a substitute for it. It is hoped that the findings will stimulate more focused research on the reasons for excess mortality in CP.

Accepted for publication 31st March 1999.

Acknowledgement

Provision of data from the California Departments of Developmental Services and Health Services is gratefully acknowledged. The views expressed are those of the authors, and do not necessarily reflect those of the State of California or its agencies. The authors also thank Dr Eva Alberman and an anonymous reviewer for helpful suggestions.

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